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NEUROPATHIC PAIN - A CASE-BASED DISCUSSION ON DIAGNOSIS & MANAGEMENT IN A CANCER PATIENT

RN Aslam, S Sivaranjan, R Al-Qurainy



Abstract

Neuropathic pain is a common symptom that can be both distressing and disabling for patients. Accurate diagnosis and proper management can dramatically improve the quality of patient lives. Here we discuss how to effectively identify and manage palliative care patients with neuropathic pain.

Case History

A 40-year-old man with recurrent, un-resectable, metastatic chondrosarcoma of the right thigh has been referred to the palliative care team for pain control. His cancer has started to infiltrate his muscles and he is complaining of pain that shoots down the back of his thigh to the foot. He describes the pain as 'sharp, shooting and sometimes burning' in nature. What do you think is causing the patient's pain and how would you manage this problem?

What is neuropathic pain?

The patient in this case scenario is most likely suffering from neuropathic pain caused by tumour infiltration of the sciatic nerve. Neuropathic pain caused by advanced cancer can be difficult to manage and often doesn't respond well to common analgesics. As pain is one of the most feared consequences of a terminal diagnosis, being able to manage it effectively is essential in palliative care.

Neuropathic pain can be considered as 'pain initiated or caused by a primary lesion or dysfunction of the nervous system' (1). Historically, pain has been classified into 'nociceptive' and 'neuropathic'. In nociceptive pain, noxious stimuli activate peripheral nociceptors which then relay warning messages to the higher brain centres. However in neuropathic pain, a dysfunction or damage to the peripheral or central nervous system results in the abnormal activation of these pain pathways. For some patients this pain may resolve as the damaged nervous system heals whereas in several cases the pain may persist, despite tissue recovery. The pain is considered chronic when it lasts for longer than three months (2).

Neuropathic Pain - A Case-Based Discussion On Diagnosis & Management In A Cancer Patient Patient Management

Pathophysiology

The sensation of pain is normally generated through activation of C- (unmyelinated) and A delta (thinly myelinated) primary afferent neurones. These neurones become increasingly sensitive and develop abnormal activity following a peripheral nerve injury. This is accompanied by various molecular and cellular changes, leading to the pathological mechanisms that result in neuropathic pain. These changes include up-regulation of voltage-gated sodium channels and the release of growth factors from degenerating nerve fibres. This hyperactivity leads to changes in the dorsal horn of the spinal cord and the brain (3).

Taking a good pain history

Taking a good pain history is extremely important as it can help narrow down the cause, allowing targeted treatment. A thorough general medical history and examination should also be carried out. The 'SOCRATES' acronym (Figure 1) has been used widely to assist health professionals in evaluating the nature of a patient's pain. It provides a good structure when taking a pain history.

- **Site:** Where is the pain? Can it be localised, or is it more diffuse and difficult to pin point?
- **Onset:** When did the pain start and was it sudden or gradual?
- **Character:** How can the pain be described (e.g. sharp, throbbing etc...)?
- **Radiation:** Does the pain radiate to anywhere?
- **Associated symptoms:** Are there any other symptoms or signs that are associated with the pain (e.g. chest pain and shortness of breath)?
- **Time course:** Does the pain follow a particular pattern over time?
- **Exacerbating & relieving factors:** What, if anything, makes the pain better or worse?
- **Severity:** How severe is the pain? (can use a numerical scale between 0-10)

Figure 1: The SOCRATES acronym for Pain

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Clues pointing towards neuropathic pain are if the pain is described as being 'shooting', 'stabbing' or 'burning'. Patients may also complain of 'numbness' or a 'prickling' sensation. Neuropathic pain can often be characterised by allodynia (when a stimulus that normally shouldn't provoke pain causes pain) and hyperalgesia (a heightened sense of pain from a stimulus that usually provokes pain). The pain can be intermittent or constant and patients may have a longstanding history of these symptoms. Although the underlying pathological cause can be complex and varied, common conditions that are often associated with neuropathic pain and worth screening for include diabetic neuropathy, multiple sclerosis, stroke and spinal cord damage (4).

Several other history taking tools have been designed to specifically help distinguish neuropathic pain. Of note, 'The Leeds Assessment of Neuropathic Symptoms and Signs' (LANNS) has been shown to be effective at assessing neuropathic pain in various different clinical contexts (5). The LANNS pain scale is a straight forward bedside test that consists of a patient completed questionnaire and a short examination by the clinician. The 7 step questionnaire & examination is shown in Figure 2.

1. Does your pain feel like strange, unpleasant sensations in your skin? Words like pricking, tingling, pins and needles might describe these sensations.

- a. NO – My pain doesn't really feel like this (0)
- b. YES – I get these sensations quite a lot (5)

2. Does your pain make the skin in the painful area look different from normal? Words like mottled or looking more red or pink might describe the appearance.

- a. NO – My pain doesn't affect the colour of my skin (0)
- b. YES – I've noticed that the pain does make my skin look different from normal (5)

3. Does your pain make the affected skin abnormally sensitive to touch? Getting unpleasant sensations when lightly stroking the skin, or getting pain when wearing tight clothes might describe the abnormal sensitivity.

- a. NO – My pain doesn't make my skin abnormally sensitive in that area (0)
- b. YES – My skin seems abnormally sensitive to touch in that area (3)

4. Does your pain come on suddenly and in bursts for no apparent reason when you're still? Words like electric shocks, jumping and bursting describe these sensations.

- a. NO – My pain doesn't really feel like this (0)
- b. YES – I get these sensations quite a lot (2)

5. Does your pain feel as if the skin temperature in the painful area has changed abnormally? Words like hot and burning describe these sensations.

- a) NO – I don't really get these sensations (0)
- b) YES – I get these sensations quite a lot (1)

The examination component

1. Does stroking the affected area of skin with a piece of cotton wool produce an unpleasant painful sensation? – Yes (5) / No (0)
2. Does touching the affected area of skin with a sharp needle feel sharper or duller when compared to an area of normal skin? – Yes (3) / No (0)

A score of less than 12 means that it is unlikely that the underlying pathology is neuropathic in nature. A score of 12 or more points suggests that a neuropathic mechanism is highly likely. (8)

Management

The complexity of neuropathic pain makes it challenging to manage. The diagnosis and management of the underlying cause, where possible, is of primary importance. For example, radiotherapy is often used to alleviate neuropathic pain caused by a bony metastasis. Following this, management may be pharmacological, non-pharmacological or interventional; optimal management may be a combination of these approaches.

The management of neuropathic pain will vary in the specialist and non-specialist setting. The NICE guidelines on neuropathic pain management are a useful tool for the non-specialist (7). Here we will discuss the management of neuropathic pain in the palliative care setting.

Pharmacological management

Combination pharmacotherapy with two drugs acting synergistically or providing additive benefit is likely to be required for many patients. However, evidence to show that combination pharmacotherapy is safer and more efficacious than for single drugs is currently lacking (8).

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Figure 3 illustrates a stepwise approach for the management of many patients with neuropathic pain (7,9,10). However, certain neuropathic pain conditions have alternative first-line management. For patients with trigeminal neuralgia, offer carbamazepine first-line; if this is ineffective, not tolerated, or contraindicated – seek specialist advice. For painful diabetic neuropathy, offer duloxetine first-line or amitriptyline if contraindicated. For all drugs, provide an adequate trial; two to eight weeks is recommended, with at least one to two weeks at the maximum tolerated dose (9).

Table 1 provides the mechanism of action and dosing information for the main neuropathic agents. Consult the BNF for full prescribing information and consider: comorbidities, safety considerations, contra-indications, patient preference, lifestyle factors, any history of mental health problems, and current medications (11).

When prescribing, give patients clear advice regarding dosage both verbally and in writing. It is important to review patients soon after starting or changing treatment. At reviews discuss: pain-control, side-effects, effect on activities of daily living, mood, sleep and overall improvement (9).

Opioid analgesics—such as tramadol, morphine and oxycodone—form a valuable second-line approach to neuropathic pain. With all opioids, it is safety concerns that limit their use to second-line therapy. However, in palliative care, opioids are often started early to treat neuropathic pain with a multifactorial aetiology (9). Tapentadol—an oral mu-opioid receptor agonist and a noradrenaline reuptake inhibitor—is a good consideration when other pharmacotherapy and intervention have failed, though evidence comparing its safety and efficacy to other neuropathic agents is lacking (12).

Patients non-responsive to first- and second-line drugs, may benefit from other pain-relieving drugs; however the evidence-base for other drugs is limited. Currently, there is little evidence supporting the use of non-steroidal anti-inflammatory drugs (NSAIDs) for neuropathic pain (9).

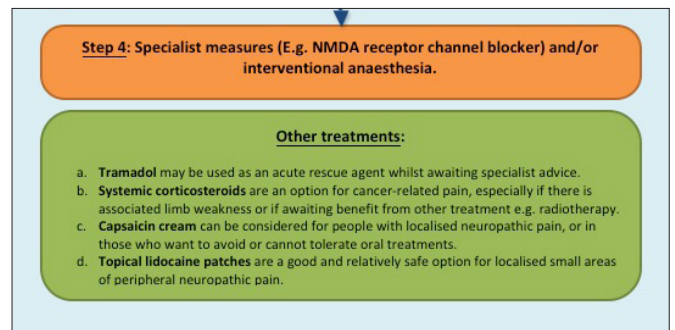
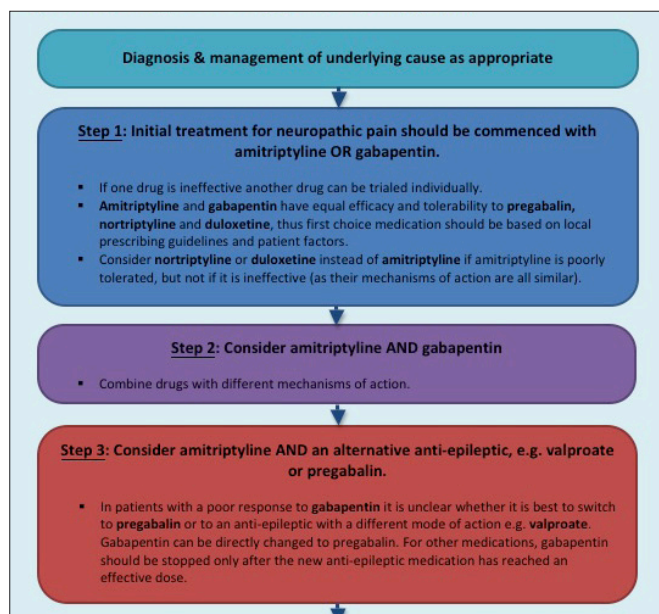


Figure 3: A stepwise approach to managing neuropathic pain (7, 9, 10, 13)

	Drug Name (Mechanism of action)	Dosing information
A N T I D E P R E S S A N T S	Amitriptyline (Serotonin and noradrenaline re-uptake inhibitor (SNRI) and a muscarinic, 5HT _{2A} , 5HT _{2C} , H ₁ , and α ₁ -adrenergic receptor antagonist)	<ul style="list-style-type: none"> Start with 10mg PO at bedtime If tolerated, increase to 25mg after 3-7 days If needed, increase by 25mg every 1-2weeks Maximum dose: Up to 150mg under specialist supervision
	Nortriptyline (Noradrenaline re-uptake inhibitor (NRI) and 5HT _{2A} , 5HT _{2C} , H ₁ , and α ₁ -adrenergic receptor antagonist)	<ul style="list-style-type: none"> Start with 10-25mg PO at bedtime If tolerated, increase by 10mg/day every 3-5 days up to 50mg, or double dose from 25 to 50mg after 2 weeks. Increase up to a max. of 150mg daily if successive increases are tolerated and add benefit. Maximum dose: 150mg daily
	Duloxetine (SNRI)	<ul style="list-style-type: none"> Start with 60mg PO once daily. If needed, increase to 60mg PO BD. Maximum dose: 120mg daily in divided doses.
A N T I - E P I L E P T I C S	Gabapentin (Pre-synaptic calcium channel blocker)	<ul style="list-style-type: none"> Start with 300mg PO at night. If needed, increase by 300mg/24h every 2-3days. For example: Day 3 300mg BD → Day 5 300mg TDS → Day 8 300mg, 300mg, 600mg → Day 11 600mg, 300mg, 600mg → Day 14 600mg TDS (typical dose for neuropathic pain) Maximum dose: 1200mg TDS. Note: The starting and max. doses of gabapentin should be reduced in adults with renal impairment & those on haemodialysis. Smaller starting doses and slower titration is recommended for elderly and frail patients.
	Pregabalin (Pre-synaptic calcium channel blocker)	<ul style="list-style-type: none"> Start with 50 - 75mg PO BD. If needed, at intervals of 3-7 days, increase up to 150mg BD → 225mg BD → 300mg BD. Maximum dose: 300mg BD. Note: In debilitated patients, start with 25-50mg BD & increase cautiously if needed. In renal impairment, reduced doses are required.
	Carbamazepine (Sodium channel blocker)	<ul style="list-style-type: none"> Start at 50-100mg PO BD If needed, increase by 50-100mg every 1-2weeks Maximum dose: 2g daily. (Undesirable effects are reduced by starting at a low dose & titrating upwards slowly.)

Table 1: Key drugs for neuropathic pain (10, 11)

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Non-Pharmacological management

There is benefit to considering non-pharmacological options, as the risk of adverse effects is low. However, for patients at the end-of-life, some modalities may not have time to be effective. Consider referral to physiotherapy (for mobilisation, exercise, TENS & acupuncture) or occupational therapy. The following treatment options may be of benefit (7):

1. Psychological techniques:

CBT has shown benefit in chronic pain treatment.

2. Electrical stimulation:

a. Transcutaneous electrical nerve stimulation (TENS) performs consistently well compared with placebo.

b. Percutaneous electrical nerve stimulation (PENS) has shown evidence of short-term benefit for refractory neuropathic pain.

3. Spinal Cord stimulation

NICE recommends this for patients with chronic pain lasting over six months (measuring 50 mm or more on a 0-100 mm visual analogue scale) despite conventional medical management and provided a prior trial of stimulation has proved to be effective.

4. Acupuncture

This has a limited evidence-base; however, it is popular among patients and is relatively harmless.

Interventional therapies

Interventional treatments for neuropathic pain should be offered to patients whose pain is not adequately managed with pharmacology, or for patients who suffer side-effects from drugs (14).

1. Steroid injection

Steroid injections can be given at a peripheral site and they are commonly given at the site of nerve injury following trauma or surgery. However, more central steroid injections can also be performed into the dorsal root ganglia or the epidural space.

2. Neuromodulation

This encompasses a range of treatments that alter pain perception by stimulating or inhibiting neural pathways.

3. Spinal cord stimulation

One or two epidural electrodes are connected to a power source which provides stimulation; this is a recognised treatment for refractory neuropathic pain.

4. Pulsed radiofrequency

An insulated needle introduced to the site of nerve damage or the dorsal root ganglion generates heat creating a temporary lesion in the nerve tissue. Beneficial effects generally last a few months.

5. Sympathetic nervous system blockade

Sympathetic nerve fibres (cervical, thoracic or lumbar) are targeted using local anaesthesia, thermal radiofrequency or chemical nerve destruction. Duration of the effects depend on the mode of treatment.

Summary

The patient presented here had sciatica subsequent to tumor infiltration. It had resulted in reduced mobility and patient anxiety. The diagnosis of neuropathic pain was made through effective history-taking, making use of the SOCRATES acronym and LANNIS questionnaire. Management was initiated using established guidelines (detailed above) and the patient was initially started on Gabapentin which was titrated to an effective dose. A referral was also made to his oncology team regarding palliative chemotherapy.

As this case exemplifies, optimal management of patients with neuropathic pain requires a multidisciplinary and multimodal approach. Effective management of neuropathic pain can result in a significant improvement in the quality of life of patients in palliative care.

Questions

1. Neuropathic pain is:

- Due to damage to the central nervous system only*
- Solely a result of increased cytokine production*
- Mainly associated with peripheral mononeuropathies*
- Due to damage or dysfunction of the peripheral or central nervous system*
- Is mainly only seen in patients with metastatic cancer*

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2. A patient comes in with unilateral severe, intense and sharp pain facial pain. Which of the following treatments should be first line?

- a) Paracetamol and NSAIDs
- b) Oramorph
- c) Gabapentin
- d) Amitriptyline
- e) Carbamazepine

3. A known diabetic patient complains of numbness and tingling in the feet which is getting worse over the past six months. Which of the following treatments would you consider initiating?

- a) Oxycodone
- b) Duloxetine
- c) Sertraline
- d) Pregabalin
- e) TENS

4. A patient has recently been commenced on gabapentin for neuropathic pain. He has been on 300mg nocte for 2 weeks. His pain remains poorly controlled. How would you manage this patient?

- a) Change gabapentin to pregabalin
- b) Stop gabapentin and consider non-pharmacological treatment
- c) Increase the gabapentin dose to 600mg nocte
- d) Increase the gabapentin dose to 450mg nocte
- e) Increase gabapentin to 300mg BD

5. A patient suffering from radicular pain secondary to lumbar disc herniation has not responded to pharmacological treatment. What intervention may be appropriate?

- a) Acupuncture
- b) CBT
- c) Epidural injection of local anaesthetic
- d) Neuromodulation
- e) TENS

Answers

1. d

Neuropathic pain can be caused by damage or dysfunction of the central or peripheral nervous system. A disruption of these pathways can result in various molecular changes that play a role in the pathophysiology of neuropathic pain.

2. e

This patient has trigeminal neuralgia. First-line treatment for this is Carbamazepine; if this is ineffective, poorly tolerated or contraindicated, specialist advice should be sought.

3. b

This patient has diabetic peripheral neuropathy. First-line treatment is Duloxetine, or Amitriptyline if contraindicated.

4. e

Gabapentin is normally started at 300mg nocte and increased in 300mg increments every 2-3 days. E.g. 300mg PO at night $\bar{\bar{D}}$ 300mg BD $\bar{\bar{D}}$ 300mg TDS $\bar{\bar{D}}$ 300mg, 300mg, 600mg $\bar{\bar{D}}$ 600mg, 300mg, 600mg $\bar{\bar{D}}$ 600mg TDS. Maximum dose is 1200mg TDS.

5. c

Epidural injections of local anaesthetics have been shown to provide symptom relief in pain secondary to spinal pathology. This is likely to be the most beneficial treatment in this patient.

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ENDOSCOPIC RETROGRADE CHOLANGIO-PANCREATOGRAPHY & ITS ROLE IN PALLIATION OF PATIENTS WITH PANCREATIC & BILIARY MALIGNANCY

L Hoggett, A Bulcock, N Amtul, J Varghese



Endoscopic Retrograde Cholangio-Pancreatography & Its Role In Palliation Of Patients With Pancreatic & Biliary Malignancy Teaching & Training

Introduction

Endoscopic retrograde cholangiopancreatography (ERCP) is a radiologically based treatment used within Gastroenterology and General Surgery for the diagnosis and management of malignancy in the pancreas and biliary system. Although the role of ERCP in diagnosis of pancreatic and biliary malignancy has been largely supplanted by the introduction and increased access to MRCP (1), there remains a role due to the ability to directly visualise and biopsy the tumour. It's most important use however, and the focus of this article, is on therapeutic stenting for obstructive jaundice for relief of associated symptoms.

Case study

A 76 year old gentleman with known pancreatic cancer presented to hospital generally unwell with lethargy, pruritus and worsening jaundice. On examination he was clinically jaundiced and appeared dehydrated. He had a slightly distended and soft abdomen. There were no masses.

His admission bloods demonstrated deranged liver function (ALT 89, ALP 443, Bilirubin 544, Prothrombin Time 17.1). He underwent Computed Tomography of his abdomen which showed a pancreatic mass with liver metastasis and associated biliary obstruction.

The patient was listed for urgent ERCP in order to facilitate relief of his obstructive jaundice.

Indications

Since its first description in the late 1960s as a diagnostic technique (2), ERCP has evolved into an almost exclusively therapeutic procedure. Over 1.3 million ERCPs are performed each year globally, with 54,000 being performed within the UK (3). The vast majority are performed in the management of choledocholithiasis due to the much higher incidence of cases compared to malignant biliary obstruction.

In patients with malignant biliary obstruction who are unable to have surgical resection with a curative intent, treatment becomes palliative and is undertaken to relieve symptoms. This can be done either surgically, endoscopically or with chemo/radiotherapy with an aim to reduce the degree of biliary obstruction and alleviate: jaundice, pruritus and fatigue. Trials have been conducted comparing surgical bypass and endoscopic stenting for palliation and do not favour one technique for all patients (4).

ERCP is advantageous as it is associated with fewer complications and a shorter treatment time. However, in patients expected to survive longer than six months, surgical palliation is preferable due to the risk of stent blockage (5).

ERCP is therefore best used in patients with advanced disease and comorbid features that leave them unable to have surgical resection.

Contraindications

Absolute contraindications for ERCP include patient refusal to undergo the procedure, severe comorbidity and perforation of the bowel. Existing structural abnormalities in the stomach, oesophagus and small intestine are relative contraindications. However, gastric outlet obstruction secondary to the malignancy may be amenable to stenting at the time of procedure (6). Finally, coagulopathy (international normalized ratio [INR] >1.5 or platelet count < 50,000/ μ L) is a relative contraindication due to increased risk of haemorrhage.

Complications

Mild pain should be expected by most patients following the procedure due to the traumatic nature of the intervention. Another common complication is mild bleeding, which for the vast majority of patients can be managed conservatively. If bleeding is more sustained, then any prescribed anticoagulants should be omitted (7) and endoscopic therapeutic intervention considered.

Infection can occur following ERCP and most patients can be adequately managed with antibiotics. However, 0.2% will suffer from cholangitis or cholecystitis, with E.Coli being the most common causative organism. Following stent insertion occlusion can occur due to precipitation of biliary sludge. This occurs predominantly with plastic stents and is less problematic with the larger metal stents (8).

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Figure 2: An interventional suite used for ERCP.

Patients are positioned prone on the ERCP table. Midazolam is titrated for sedation and Fentanyl is used for pain relief throughout the procedure. Hyoscine Bromide is admitted during the procedure for smooth muscle relaxation. Peripheral oxygen saturations are taken and oxygen can be given via nasal cannula if required.

4. The procedure itself

The side viewing endoscope is passed through the mouth into the oesophagus, stomach and through the pylorus into the duodenum. As the endoscope is side viewing any resistance should be approached with caution and a normal (forward view) endoscope passed to identify any problems. Once in the duodenum, the ampulla is identified and cannulated with a guidewire. Radio luminescent contrast is then injected via the guidewire and fluoroscopic images taken.



Figure 3: Fluoroscopic image showing the endoscope and guidewire passing into a dilated cystic duct.

Once the site of stenosis has been identified, the guidewire is passed beyond the area to facilitate stent placement. The stent is then passed endoscopically, over the guidewire into the narrowed segment and expanded. Positioning is confirmed fluoroscopically.



Figure 4: Fluoroscopic image showing a metal stent in situ.

Once the procedure is completed the endoscope is withdrawn.

5. Post procedure

Once the procedure is completed, the patient is moved to a recovery area for around 30 minutes for monitoring of observations. If any problems arise patients can be seen promptly by a member of the medical team. Once stable, patients are transferred back to the ward. Findings from the ERCP are documented by the endoscopist along with any complications or intervention performed. If any complications have arisen responsibility lies with the endoscopist to initiate appropriate measures to correct them.

Common complications and their treatments are highlighted under complications earlier in this article. Following stent insertion patients are typically referred to an oncologist for consideration of chemo or radiotherapy. It is important that follow up support is put in place for patients and their families and this is typically facilitated by: the medical team, specialist nurses, palliative care teams and Macmillan support workers.

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Questions

1) A 59 year old man presents with painless jaundice. He has no documented history of gallstones and is otherwise well. O/E: A mass is palpable in the RUQ. His blood profile shows an obstructive picture. Diagnosis?

- A. Biliary stricture
- B. Cholecystitis
- C. Head of pancreas tumour
- D. Choledocholithiasis
- E. CBD calculus

2) A 54 year old man presents with painless jaundice (bilirubin > 200 micromol/L). CT features suggest a pancreatic head mass with no lymphadenopathy. Further staging is awaited and will take over 2 weeks to arrange. Next appropriate step:

- A. Endoscopic stenting of CBD
- B. Best supportive care
- C. Referral for chemoradiation
- D. Whipple's procedure
- E. Gastrojejunostomy

3) Carcinoma of the pancreatic head:

- A. Typically presents with obstructive jaundice and non palpable gallbladder
- B. Is highly radiosensitive
- C. Is accurately staged with USS
- D. Causes a rise in serum CA19-9
- E. Is associated with a good prognosis

4) What type of stent is commonly used in unresectable disease if the patient is expected to live for another six months?

- A. Plastic stent
- B. Glass stent
- C. Metal stent
- D. Pigtail stent
- E. JJ stent

5) Absolute contraindications to ERCP include all of the following EXCEPT:

- A. Existing bowel perforation
- B. Existing esophageal perforation
- C. Patient refusal
- D. Gastric outlet obstruction
- E. Unstable cardiovascular status

Answers

Question 1: Courvoisier's Law

In the presence of an enlarged non-tender gallbladder accompanied with jaundice, the cause is unlikely to be gallstones. Likely to result from carcinoma of the head of the pancreas. Gallstones usually cause scarring of the gallbladder from infection making it fibrotic and it does not distend.

Question 2

The young patient may have resectable disease but until full staging info is complete, it would be preferable to relieve his jaundice. Should use a plastic stent as metal stents may render the disease more difficult to resect in the future. If surgery is to be delayed by more than 10 days, it is recommended that drainage is achieved and surgery delayed to allow the jaundice to resolve.

Question 3

CA19-9 (carbohydrate antigen 19-9) is a tumour marker that is used primarily in the management of pancreatic cancer. If the tumour is secreting it, then the levels should fall when the tumour is treated, and they may rise again if the disease recurs. No symptoms usually occur in the early stages that are specific enough to suspect pancreatic cancer. By the time of diagnosis, it has often spread to other parts of the body. Diagnosed by a combination of imaging ie. USS, CT.

It has a very poor prognosis. 1 year survival 25%; 5 year survival 5%.

Question 4

Metal stents are preferable in patients who have a live expectancy of around 6 months. This is due to the fact that metal stents are much less likely to become obstructed than the narrower gauge plastic stents.

Question 5

Gastric outlet obstruction is a relative contraindication and may also be amenable to stenting at the time of the procedure. This has the added benefit of symptomatic relief of the symptoms associated with gastric outlet obstruction. All the other options are absolute contraindications to ERCP.

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MALIGNANT BOWEL OBSTRUCTION

L Karsera, F Shakir, E Boland

Malignant Bowel Obstruction Patient Management

Abstract

In this article we present a patient with malignant bowel obstruction secondary to peritoneal carcinomatosis. The patient was not suitable for surgical treatment but had a radiological insertion of a venting gastrostomy, medical management of the symptoms and received artificial hydration. The patient was not suitable for surgical treatment but had a radiological insertion of a venting gastrostomy and received artificial hydration and symptomatic medical management. This case highlights the importance of palliating the symptoms of nausea and vomiting in malignant bowel obstruction and discusses non-surgical management in a patient with limited life expectancy. It also raises issues around artificial nutrition and hydration at the end of life.

Introduction

Malignant bowel obstruction (MBO) is a key differential diagnosis when a patient with advanced cancer presents with nausea and vomiting. Bowel obstruction is defined as any mechanical or functional obstruction of the intestine that prevents physiological transit and digestion. It is a common clinical complication in patients with advanced abdominal or pelvic malignancy, such as colonic, ovarian, or gastric cancer (1). Extra-abdominal cancers such as lung, breast, and melanoma can also spread to the abdomen, causing bowel obstruction. Obstruction can be due to direct tumour infiltration, intraluminal obstruction or external obstruction and may occur due to tumour growth, adhesions, carcinomatosis, faecal impaction and neuropathy (2).

Nausea and vomiting are common symptoms in patients with cancer and can be multifactorial. Table 1 includes the different aetiologies. Some of these causes are easily reversible but in others, management is more complex.

We present a patient with MBO who was not suitable for surgical treatment. This lady received palliation of her symptoms, artificial hydration and insertion of a venting gastrostomy. This case highlights the difficulties in palliating the symptoms of nausea and vomiting in MBO and discusses conservative management in a patient with a poor prognosis.



Gastrointestinal	Gastritis Gastroparesis (e.g. due to opioids, or mechanical factors) Hepatomegaly Ascites Bowel obstruction Localised tumour effect, Radiation colitis
Cranial	Raised intracranial pressure (e.g. due to brain metastasis or haemorrhage, meningeal disease or primary tumour)
Drugs	Cytotoxic chemotherapy Opioids
Metabolic	Hypercalcaemia Uraemia
Cortical	Anxiety Pain Anticipatory nausea

Table 1: Aetiology of nausea and vomiting in malignancy.

Case History

A 52 year old female with a history of advanced bilateral ovarian carcinoma presented with vomiting and constipation. She was diagnosed with ovarian cancer 3 years previously and was initially treated with chemotherapy and surgery. Unfortunately her disease progressed despite multiple lines of chemotherapy and had been complicated previously by small bowel obstruction, which had been managed conservatively.

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On admission, the patient reported intractable vomiting exacerbated by eating and drinking. She had not opened her bowels for 4 days and was not passing flatus. She was made nil by mouth, and commenced on intravenous (IV) fluids, laxative (movicol) and anti-emetics (cyclizine 50mg TDS subcutaneously and levomepromazine). Abdominal x-ray was unremarkable and initial bloods were within normal limits except for sodium 127mmol/L and urea 14.5mmol/L. A nasogastric (NG) tube was inserted and left on free drainage. Abdominal CT scan revealed dilated loops of small bowel (image 1) and extensive peritoneal disease (image 2). The surgical team felt that palliative surgery was not an option.



Image 1: A coronal CT scan showing some dilated loops of bowel.



Image 2: An axial CT that shows small bowel obstruction with peritoneal disease.

She continued to vomit large volumes on a daily basis with occasional nausea. She was commenced on continuous subcutaneous infusion over 24 hours of levomepromazine 6.25mg, increased to 12.5mg after 48hours, octreotide 500micrograms, later increased to 1000micrograms, and cyclizine was stopped. She only managed to take small sips of fluid and was dependent on IV fluids for hydration. She was reviewed by a dietician, and a multidisciplinary team decided that since the prognosis was less than 3 months she would not be a suitable candidate for total parenteral nutrition; small amounts of oral fluids and food should continue as tolerated. After her first NG tube was dislodged, the patient refused placement of another as she found this procedure distressing.

This lady was determined that she wanted to attend her son's wedding in 4 weeks' time and her preference was to be at home for as long as possible.

The possibility of a palliative venting gastrostomy for symptom relief was discussed with the radiologist especially as this lady felt that eating was important to her and did not want to have a NG tube re-inserted long-term. After discussion with the patient, the venting gastrostomy was inserted (image 3) and she managed to aspirate the gastrostomy effectively when she felt the need to vomit and thus was able to enjoy meals with her family.

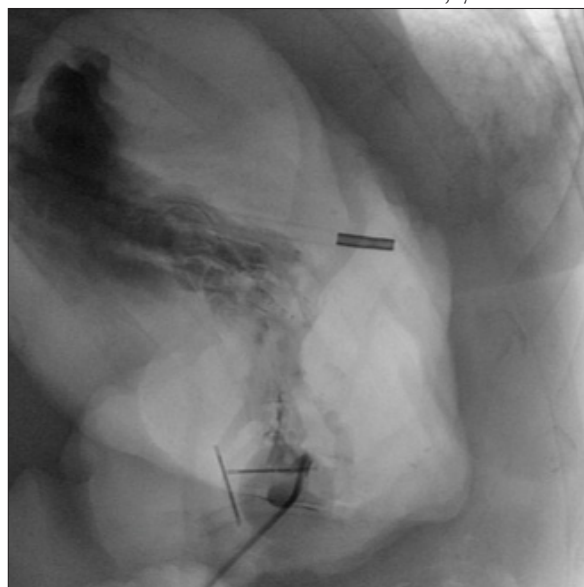


Image 3: Fluoroscopy – gastrostomy insertion with air in stomach.

She was still dependent on IV fluids;; therefore in an attempt to enable the patient to go home, a trial of subcutaneous fluids was tried. Unfortunately pooling of the subcutaneous fluid occurred and she had to return to having IV fluids, which could only be administered at hospital. A plan was implemented with the patient returning to the day unit three times per week for IV fluids. The patient was discharged home where she wanted to be; however, she continued to deteriorate and died peacefully at home a few days prior to the family wedding.

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Discussion

In this case, the lady had peritoneal carcinomatosis and symptoms of bowel obstruction. Investigations include biochemical profile to exclude hypokalemia, which may cause ileus, and hypercalcaemia, which may cause pseudo-obstruction due to constipation. A clinical and radiological assessment is vital; a CT scan of the abdomen is the recommended investigation for bowel obstruction.

Treatment requires individualised evaluation. Although surgery should be the primary treatment for selected cases, patients with poor nutritional and performance status, ascites, extensive peritoneal disease with multi-level obstruction and previous abdominal or pelvic radiotherapy have poor prognostic factors (3). In this case, the patient was not a candidate for surgery due to advanced peritoneal disease, poor performance status and poor nutritional state. This situation carries a poor prognosis and patients are at risk of recurrent obstruction even if the initial episode resolves.

When surgery is not an option, palliative management aimed at reducing symptoms and optimising quality of life becomes the focus of care. If intractable vomiting and/or gastric distention is present; NG suction or drainage would help with these symptoms. Long term NG drainage is not recommended due to nose and throat pain, abscess formation, erosion of nasal cartilage and social isolation. If the patient is draining more than 1L/day in the NG tube, then removal of this would lead to vomiting unless symptoms can be managed with pharmacological agents.

Self-expanding metallic stent placement is an effective non-invasive procedure to open stenosed lumen. It provides good palliation for unresectable advanced tumour causing obstruction at single level (4).

Figure 3 is a flow chart discussing the management of MBO.

Venting gastrostomy

Gastrostomy is the procedure of choice for long-term gastric decompression and intractable vomiting as an alternative to a NG tube. Intermittent venting of the gastrostomy provides patients the satisfaction of resuming oral intake of some foods, giving them significant psychological benefit, without the inconvenience of a NG tube. Venting gastrostomy can be performed under local anaesthesia either radiologically or endoscopically and may facilitate discharge of patient to their preferred place of care.

Medical symptom management

To control pain, nausea and vomiting in bowel obstruction, pharmacologic therapy involves anti-emetics, anti-secretory and analgesic agents. Pain might be either a constant dull ache and/or colicky abdominal pain.

As oral administration is unreliable due to nausea, vomiting and malabsorption, the parental route is mandatory; medication should generally be given by subcutaneous injection or a continuous subcutaneous infusion (CSCI) that allows constant drug infusion with minimal discomfort to the patient.

Antiemetics

Metoclopramide, a prokinetic, can be used in patients with functional or incomplete obstruction without colicky pain. Cyclizine is useful for managing vagally-mediated nausea and vomiting. Haloperidol acts on the vomiting centre. Levomepromazine is a broad-spectrum anti-emetic used 2nd or 3rd line, as it can cause sedation and hypotension. The 5-HT₃ receptor antagonists such as ondansetron (for chemotherapy and/or radiotherapy-induced nausea and vomiting) can be tried in obstruction as serotonin can be released from the intestine.

Antisecretory drugs

Anticholinergic drugs such as hyoscine butylbromide can reduce vomiting by decreasing secretions and also have antispasmodic properties. This should not be co-prescribed with metoclopramide.

Gastric antisecretory drugs, such as proton-pump inhibitors inhibit the secretion of hydrochloric acid and prevent bile reflux which can help in MBO.

Somatostatin Analogues

Octreotide inhibits vasoactive intestinal polypeptide activity in the gut, reducing gastric and pancreatic secretions and water and electrolyte excretion in the lumen. It also reduces splanchnic blood flow, indirectly decreasing gut wall oedema, peristalsis and bile excretion.

Steroids

Short course parenteral steroids (5 to 10 days) can reduce peri-tumour oedema and decrease the stenosis.

Hydration and Nutrition

The use of artificial hydration and nutrition in patients with limited prognosis such as in this case is a challenging. By law artificial hydration and nutrition are medical treatments. Good quality evidence is lacking (5), therefore decisions are made on an individual basis and daily assessment of the beneficial and detrimental effect of the fluids is needed, with clear documentation and communication with the patient and their families (6). In this case, IV fluids was continued on an intermittent outpatient basis as subcutaneous administration of fluids was not feasible. The driving factor for this was that the patient was getting symptomatic dehydration and to try and support her wishes to achieve her goal of surviving for her son's wedding.

Total parenteral nutrition (TPN) was also considered. The objective of TPN is to prevent malnutrition and maintain a good quality of life, with the aim to be able to deliver further anti-cancer treatment and improved survival in selected patients. TPN is an invasive technique that requires specific training for use and frequent monitoring of electrolytes and hydration. It also predisposes patients to infection (due to central venous access), thrombosis, diarrhoea, liver dysfunction, and hyperglycemia. It is used in patients with reasonable expectations of survival and a good performance status (7). In this case, the patient was not fit for further chemotherapy and the patient's prognosis was short, thus TPN was deemed not to be suitable.

MALIGNANT BOWEL OBSTRUCTION

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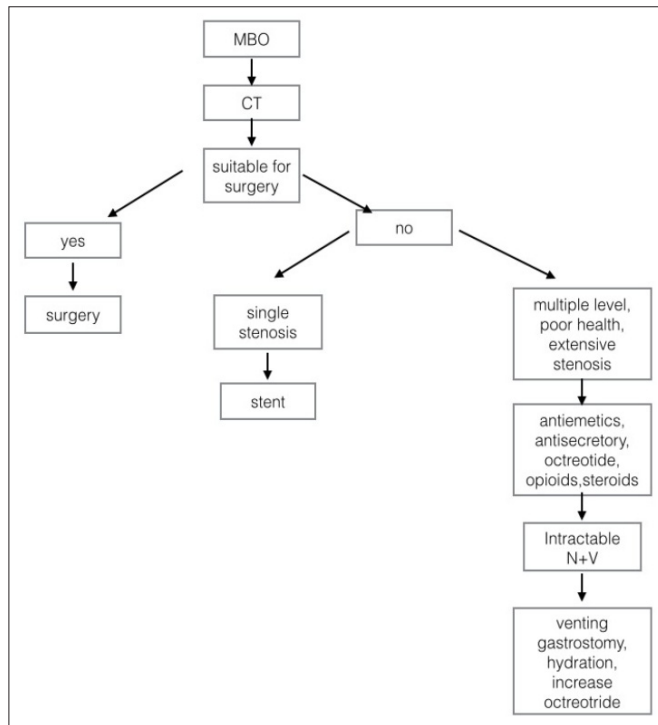


Figure 3: Flow chart for treatment of malignant bowel obstruction.

Bowel obstruction is a common yet very complex problem in patients with advanced abdominal and pelvic malignancies. Comprehensive care is achieved by a multi-disciplinary approach ensuring careful assessment, implementing an informed plan for controlling symptoms, and providing psychosocial support.

Note: Louise Karsera and Fahima Shakir contributed equally to this article and are joint 1st authors.

Questions

1. Which antiemetic should be avoided in complete intestinal obstruction?

- Cyclizine
- Levomopromazine
- Ondansetron
- Metoclopramide
- None of the above

2. Complete mechanical bowel obstruction can cause dehydration by:

- Interfering with oral intake of water.
- Inducing vomiting.
- Decreasing intestinal absorption of water
- Causing secretion of water into the intestinal lumen.
- All of the above

3. What are the signs and symptoms of dehydration?

- Thirst
- Diminished skin turgor
- Decreased urine output
- Confusion
- All of the above

4. With regards to venting gastrostomy, which of the following statement is false:

- Permits long term gastric drainage and decompression
- Requires an NG tube to remain in place
- Contraindication for gastrostomy tube is presence of significant ascites.
- Can alleviate intractable vomiting.
- Well tolerated and can go home with the tube.

5. Which of these is least likely to occur as a complication of total parenteral nutrition?

- Infection
- Electrolyte imbalance
- Renal failure
- Thrombosis
- Liver dysfunction

Answers

1. d.

Metoclopramide should be avoided in complete obstruction as it may worsen colicky pain and can lead to bowel perforation due to the drug's prokinetics effects but has been used successfully in patients with functional or incomplete obstruction.

2. e.

One of the most important events during mechanical bowel obstruction, loss of water and electrolytes from the body, is caused mainly by intestinal distention. Distention may produce reflex vomiting, intestinal secretion and causes decreased absorption of water.

3. e.

The signs and symptoms of dehydration include Thirst, Fatigue, Dizziness, Headache, Diminished skin turgor, Dry oral mucous membranes, Sedation, Agitation, Confusion, Delirium, Hallucinations, Myoclonus and Decreased urine output.

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4. b.

Whilst an NG tube may be required temporarily to guide the placement of a venting gastrostomy, this can be removed immediately after placement. The venting gastrostomy provides long term gastric drainage and decompression while avoiding the discomfort of a nasogastric tube which causes inconvenience and at worst contributes to complications of the upper and lower respiratory tract.

5. c.

The other options occur more frequently in association with total parenteral nutrition. Due to the risk of such complications the decision to commence TPN should be made as part of multidisciplinary team discussions.

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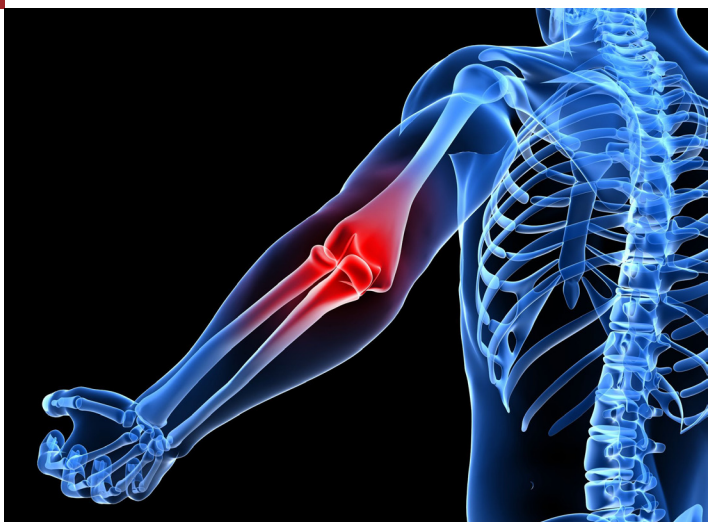
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MANAGING BONE PAIN IN METASTATIC DISEASE

A Ling, J Hayes



Managing Bone Pain In Metastatic Disease Patient Management

Case Report

A 75-year-old patient with metastatic renal cell carcinoma was admitted under the palliative care team for symptom control of pain. He had undergone a radical nephrectomy many years previously but had subsequently developed multiple bony metastases [see figures 1-2]. In addition to his initial surgery, he received chemotherapy and radiotherapy to both hips and pelvis. He then received prophylactic intramedullary limb nailing procedures in both femurs as metastatic disease had resulted in impending fractures.

On examination, his observations were stable but he appeared visibly in pain. He described a constant dull aching sensation across his right thigh, which he estimated at 3/10 in severity. This pain became sharper in nature and intolerable on movement, up to 8/10 in severity. He was unable to weight bear and was bedbound, with minimal activity (such as using a bedpan) causing severe discomfort. Leg movements were limited by pain but he was able to complete anti-gravity movements bilaterally. His left leg, although affected by metastatic disease, did not cause significant discomfort. No other abnormality on systemic examination was identified. Bloods, including a bone profile, were within normal limits.

A combination of non-opioids and opioids were trialled. Unfortunately he developed an acute kidney injury secondary to ibuprofen and experienced intolerable tremor when trialling gabapentin. An opioid-based continuous subcutaneous infusion was started with good reduction in background pain, but almost no improvement in movement-induced pain. However, at higher doses he became increasingly drowsy and confused, in keeping with opioid toxicity.

Despite opioid rotation, changing from diamorphine to oxycodone to alfentanil, pain and side effects remained problematic. The patient had an epidural catheter inserted for continuous infusion of local anaesthetic and opioid, reducing his perceived pain severity from 8/10 to 2/10. He remained bedbound but was able to turn and sit forward without pain. In addition to medical intervention, the patient also benefited from psychological support, physiotherapy and occupational therapy as part of a holistic approach.

Abstract

This case discusses the management of painful bone lesions in metastatic disease, the most common cause of cancer pain. Whilst working as a foundation doctor in a palliative care unit is unusual, foundation doctors will undoubtedly be involved with the management of palliative patients whilst working in other disciplines. Initiating appropriate and effective analgesia before requesting senior or specialist advice is a common and important responsibility of a foundation doctor. Using the example of a patient with complex pain from metastatic renal cell carcinoma, the available pharmacological and non-pharmacological treatments are explored.

Declaration

This paper has not been submitted elsewhere for publication. There are no conflicting interests at the time of writing but Marie Curie Cancer Care may pay for the publication fee of the article. This has yet to be confirmed. No ethical approval was required to write this article.

Confidentiality

Unfortunately the patient is now deceased. Attempts were made to contact next of kin to ask for consent on behalf of the patient, but these were unsuccessful. The images used are not identifiable to the patient as all demographic information has been removed. Some demographic aspects of the case have been modified to anonymise without affecting the message of the case.

The article was written by Dr Andrea Ling and supervised by Dr Joanne Hayes. The patient's consultant was Dr Joanne Hayes.

MANAGING BONE PAIN IN METASTATIC DISEASE

A Ling, J Hayes



Figure 1: CT scan (coronal slice) showing a right-sided renal mass, prior to radical nephrectomy.



Figure 2: X-Ray right femur. Metastatic lesion in the distal femur prior to fixation.

Discussion

The most common cause of chronic pain in cancer patients is due to bone metastases, although 25% of patients with known metastatic lesions do not complain of bone pain (1). The most common cancers to metastasise to bone include breast, prostate, lung and renal but any cancer type may be involved (2).

Patients may describe a dull, aching pain that is present most of the time as 'background pain'. They may complain of spontaneous increases of pain at rest and movement-related incident pain (2). The pathophysiology behind bone pain is multifactorial and includes both inflammatory and neuropathic elements. Patients who describe neuropathic features (e.g. shooting, burning) of their pain tend to rate their overall pain as more severe (3).

Disruption of bone metabolism is a significant factor. There is increased expression of RANK-ligand (a membrane protein secreted by many cells, including osteoblasts) in the presence of cancer cells and this stimulates increased osteoclast activity and thus bone destruction (4). Neurons within bone may be structurally damaged by metastases and become sensitised by the acidic environment caused by cytokine release, worsening neuropathic pain (4,5).

Analgesics

The World Health Organisation analgesic ladder is a useful tool for management in any type of cancer pain (6)[see figure 3]. However, in severe pain other pharmacological and non-pharmacological approaches may be needed. In the case described, an escalation to 'Step 3' of the analgesic ladder proved to be insufficient and caused significant opioid-related side effects.

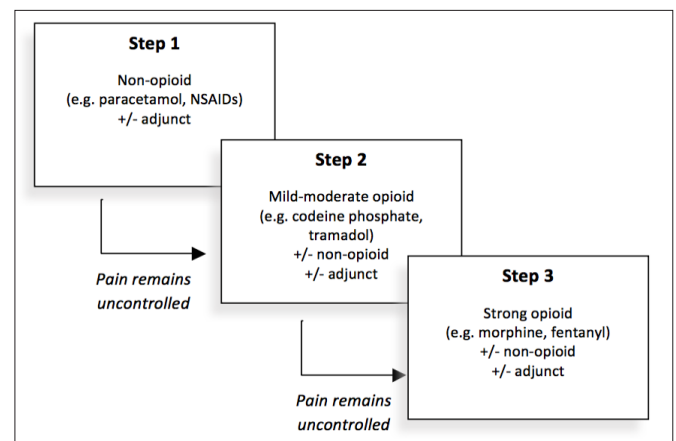


Figure 3: The World Health Organisation analgesic ladder (6,7).

Non steroidal anti-inflammatory drugs (NSAIDs) such as ibuprofen are often used as single agents or in combination with opioid based medication. There is evidence that COX2 inhibition, which forms the basis of action of NSAIDs, has anti-tumour and antiangiogenesis properties (8,9). A 2004 systematic review concluded that there is no benefit of any NSAID over another and that short term courses tend to be the most effective (10). Monitoring renal function and prescribing gut mucosal protection (e.g. a proton pump inhibitor) is important in these patients.

MALIGNANT BOWEL OBSTRUCTION

L Karsera, F Shakir, E Boland

Opioids are important and effective in preventing and controlling bone pain. These drugs work on four receptors; μ (mu), κ (kappa), δ (delta) and a newly recognised receptor, nociceptin orphanin FQ peptide (11). Of these, μ is clinically the most important. The μ opioid receptor inhibits nociceptive pathways and is used by all exogenous opioids (12). The activity at these receptors is also responsible for opioid side effects, including respiratory depression, reduced gut motility, sedation and tolerance (11). Patients may have a modified-release preparation (e.g. sustained-release morphine, fentanyl transdermal patches), with a breakthrough normal-release preparation (e.g. oral morphine, immediate release fentanyl preparations). The breakthrough choices may depend on the predictability of the breakthrough pain and the tolerability of side effects.

Adjuncts used for neuropathic pain (e.g. gabapentin, pregabalin, amitriptyline) have a role in the management of bone pain. The method of action of gabapentin and pregabalin is similar but not well described. The drugs are structurally similar to the neurotransmitter GABA, but do not work within GABA pathways, acting upon other neurotransmitters such as voltage-gated calcium channels (13). Tricyclic antidepressants such as amitriptyline typically achieve pain relief at lower doses than those prescribed for depression. The mode of action relies on NMDA, sodium and calcium channel receptors and therefore in part have actions similar to local anaesthetic (14).

Bisphosphonates

Osteoclast inhibitors such as bisphosphonates have multiple uses in the management of bone metastases. This includes treating hypercalcaemia, preventing fractures and reducing the number of bone metastases (15). Bisphosphonates can offer pain relief, although this treatment may take months to offer benefit. Short term effects are modest and evidence is based only on a small number of studies. It is possible the short term effects are due to the drug reducing local tumour growth factors, whilst the long term benefits are due to the reduction in bone resorption and healing (2).

Radiotherapy

External beam radiotherapy (RT) directed locally is the most commonly used method of using radiation to treat bone pain (2). A Cochrane review concluded that RT was an effective intervention with around 25% of patients achieving total pain control one month post treatment and 41% of patients having a 50% reduction in pain (2,16). Radiation doses given in palliative RT are high enough to kill tumour cells, but this fails to explain why the treatment continues to be effective at low doses. The destruction of inflammatory cells around the affected bone lesion are likely involved in reducing pain and evidence suggests RT inhibits osteoclastic activity, providing benefits in a similar way to bisphosphonates (17).

The patient had received palliative RT to both femurs in the year prior to fixation surgery. This had achieved good effect at the time with improved mobility. RT may not be suitable for all patients, as travel to a centre offering the treatment may be unsuitable and side effects such as nausea and fatigue must be considered (18).

Orthopaedic procedures

Bone metastasis in the long bones cause fractures in about 25% of cases, although this is significantly more likely if metastases affect the proximal femur. In the case of fracture, fixation is used to help unite the bone (2). In the case of bone pain with a significant risk of fracture, patients may be offered fixation surgery; Mirel's scoring system has been used to classify the risk of pathological fracture in bone lesions and offers a guideline for orthopaedic surgeons (19). Risk factors for impending fracture in this patient included the site of the metastasis (lower limb), severity of pain and the lytic nature of the lesion. He was therefore offered an intramedullary fixation (Figures 3 & 4).



Figures 3 & 4: These images show the placement of the intramedullary nail in both the proximal femur (left image) and distal femur (right image). The lytic lesion can be seen in the distal femur proximal to the fixation screws.

Interventional pain techniques

In patients where usual methods have failed to control pain or where side effects of increasing opioid load are intolerable, the advice of a specialist pain team should be sought. Interventions including nerve blocks, epidural devices or intrathecal devices can be used. Intrathecal pain relief can be administered either by a catheter with external attachment, or by a pump which is placed under the subcutaneous fat to offer a long-term option (e.g. months to years) (20).

Compared with epidural interventions, intrathecal devices offer a more reliable longer term option, use smaller devices and require less frequent refilling (21). However, the risk of serious infection increases as the dura is breached. Similarly to RT as discussed earlier, these options may not be appropriate if patients are too unstable for transfer to a site that offers these treatments. The post-procedural care is also significant and may mean a patient requires care in a specialist facility with staff trained to use these devices.

MALIGNANT BOWEL OBSTRUCTION

L Karsera, F Shakir, E Boland

In the case described, intervention with an epidural device was warranted due to the complexity of the patient's pain and the inability to obtain adequate pain control with usual methods. However, the challenge of arranging the procedure and ensuring subsequent aftercare was considerable. The procedure was successful and provided the patient with adequate pain relief over a longer-term basis.

Holistic approach



A specialist palliative care facility will provide additional means of support for patients with pain. Before being admitted, the patient had received a wheelchair and shower aids to facilitate independence at home through the occupational therapy team. He was seen throughout admission by the physiotherapists, who provided exercises to maintain strength where appropriate and also advised other staff on moving and handling techniques. Counselling and reflexology were also offered.



Conclusion

Bone pain from metastatic disease is common in the cancer population and there are many medical and surgical interventions that can assist patients in managing this. The WHO analgesic ladder is an appropriate starting point in a non-specialised setting, but advice should be sought from oncology, palliative care and / or an interventional pain team if pain is not easily managed.

Learning Points

1. Pain from metastatic bone disease is multifactorial. An accurate pain history will guide which opioids or adjuncts are most likely to be helpful.
2. The WHO analgesic ladder provides a basis for non-specialists to trial analgesics before referring to specialist services.
3. Surgical intervention such as intramedullary fixation should be considered for patients at high risk of fracture.

MCQs - Managing bone pain in metastatic disease

1. Which of these opioids is safest in the context of renal failure?

- a) Codeine phosphate
- b) Diamorphine
- c) Fentanyl
- d) Morphine
- e) Oxycodone

2. A patient is receiving 45mg sustained-release morphine BD for pain management. You are asked to prescribe a breakthrough dose of oral morphine. What dose would be most appropriate?

- a) 4.5mg
- b) 7.5mg
- c) 15mg
- d) 18mg
- e) 30mg

MALIGNANT BOWEL OBSTRUCTION

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3. Small doses of oral morphine may be used routinely in managing which common symptom (other than pain) in palliative care?

- a) Anxiety
- b) Breathlessness
- c) Diarrhoea
- d) Hallucinations
- e) Hiccups

4. What intervention should be treated with caution in patients taking bisphosphonates?

- a) Central line insertion
- b) Dental extraction
- c) Nasogastric tube placement
- d) Intravenous rehydration
- e) Radiotherapy

5. An inpatient with metastatic lung cancer has been titrated up to 80mg sustained-release morphine BD with 25mg oral morphine as a breakthrough dose. Her pain has been well controlled. When you see her, she is drowsy, confused and has myoclonic jerking of both arms. Her respiratory rate is 10/min and renal function is normal. You suspect opioid toxicity. You start some IV rehydration. Bloods are unremarkable. What should you change in relation to her opioids?

- a) Change sustained-release morphine to oxycodone
- b) Reduce the dose of breakthrough oral morphine
- c) Reduce the sustained-release morphine dose by one half
- d) Reduce the sustained-release morphine dose by one third
- e) Stop sustained-release morphine and use regular oral morphine

Answers

1. Answer: C

Fentanyl. Fentanyl is the most suitable option here. The other opioids have active or toxic metabolites excreted by the kidney.

2. Answer: D

15mg. One sixth to one tenth of the total daily dose is appropriate breakthrough analgesia. This patient receives 90mg sustained-release morphine daily, so the breakthrough dose would be around 15mg.

3. Answer: B

Breathlessness. Oral morphine is frequently used for patients experiencing breathlessness on an as required basis. The mechanism of action is not well described. Other interventions should be trialled first in patients with diarrhoea. Opioids could worsen anxiety and/or hallucinations in some patients. Opioids are not recommended for treatment of hiccups.

4. Answer: B

Dental extraction. Having dental procedures such as extraction increases the risk of osteonecrosis of the jaw, a rare but important side effect of bisphosphonates.

5. Answer: D

Reduce the sustained-release morphine by one third. If a patient becomes opioid toxic but has adequate pain control, the total daily dose should be reduced by one third and the patient should be kept well hydrated. If this is unsuccessful, rotation to another opioid (e.g. oxycodone) could be considered.

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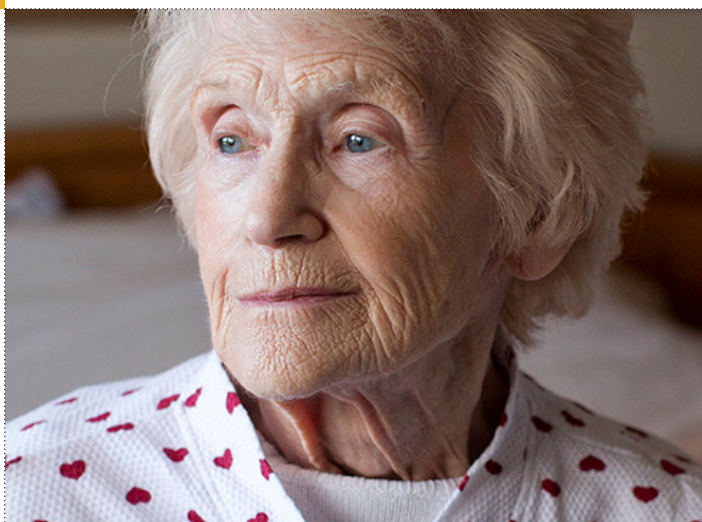
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PALLIATIVE CARE EMERGENCIES FOR THE JUNIOR DOCTOR

K Georgiade, E McKenna



Palliative Care Emergencies For The Junior Doctor Good Clinical Care

Abstract

This is an article regarding the clinical management of palliative emergencies. The aim of this review article is to increase the confidence of front line doctors in diagnosing and managing emergencies in palliative care. Through a literature review, the discussion wishes to raise awareness in the management of the palliative patients but also explore some of the ethical dilemmas related to their treatment.

Palliative care provides a multidisciplinary approach to people suffering from all life-limiting illnesses. We normally associate the symptom relief with this speciality rather than the management of acute clinical situations. Over the years though, there has been an increased understanding of the importance of recognising emergency scenarios and the need for an urgent response. The most commonly encountered conditions and their management are listed below.

Hypercalcemia of malignancy

Metabolic disturbance that is characterised by raised levels of calcium, can be classified as mild (values 2.60-3.0 mmol/l), moderate (3.0-3.4 mmol/l) or severe (above than 3.0mmol/l). The value taken into consideration is that of corrected calcium. This is of particular importance to the population seen by palliative care services in view of the high percentage of cachexia.

Hypercalcemia can be seen in about 3-30% of the total population of cancer patients(1). Lung (more specifically squamous cell carcinoma), breast, head, neck and haematological malignancies can be complicated by hypercalcemia with increased frequency(2). The main mechanisms involved are the production of a parathyroid-like protein (PTH-rP) by the primary tumour and the production of cytokines causing bone resorption typically by secondary bone tumours(3). Hypercalcemia is a sign of advanced disease and poor prognosis, and therefore it is not surprising that it is frequently encountered in the population seen by palliative care services.

The symptoms associated with hypercalcemia are closely related to the severity of metabolic disturbance and are similar irrespective of the primary cause of high calcium. Polyuria/polydipsia, nausea/vomiting, constipation, tiredness/reduced level of consciousness, electrocardiographic changes, can be observed depending on the level of calcium(4). Notably, levels above 3.0 are deemed to be life-threatening and can lead to a terminal event. The decision to treat will need to be made on the grounds of the patient's functional level and their personal wishes.

The first step in treating hypercalcemia is the correction of dehydration with parenteral fluids(5). Biphosphonates are the mainstay of treatment. The ones that are used with increased frequency are Zoledronic acid and Pamidronate, with research favouring the use of Zoledronic(6). In the palliative setting the above medications have the additional advantage of improving the pain related to bone disease. Denosumab, a human monoclonal antibody, has been successfully used for biphosphonate-refractory hypercalcemia but evidence is still limited(7),(8). The use of Furosemide, despite being widespread, is not supported by strong evidence(9).

Opioid toxicity

Opioid use is widespread in the setting of palliative care. Toxicity in different degrees of severity can therefore be seen quite frequently in this patients' population. Toxicity can be seen after intentional or accidental overdoses but also with the use of the patient's regular doses of medication.

Opioids are generally metabolised in the liver and excreted by the kidneys. The accumulation of the opioids' metabolites results in a variety of side-effects such as shadows at the peripheries of visual fields, visual and auditory hallucinations, pin-point pupils, myoclonus, sedation, confusion and reduced consciousness, as well as the feared respiratory depression. The majority of opioids are renally excreted and therefore toxicity is frequently seen in relation to dehydration or renal impairment(10).

PALLIATIVE CARE EMERGENCIES FOR THE JUNIOR DOCTOR

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The management of opioid toxicity depends on the severity of symptoms. For most patients a dose reduction in their routine opioid and subsequent careful monitoring will suffice. For more severe symptoms, opioid reduction and rotation is required. It is good practice to check the patient's renal function(11) and eliminate other potential factors which could be altering the metabolism of their routine opioid, such as sepsis. In suspected renal impairment, Alfentanil is a well tolerated opioid as it is metabolised in the liver(12). If there are signs of dehydration or the patient has a generally reduced oral intake, the administration of fluids via the intravenous or subcutaneous route can also be considered. It is rarely necessary to withdraw opioids completely. Complete withdrawal should only be considered in severe or life-threatening symptoms. Close monitoring of patients following a dose reduction or an opioid rotation will be necessary until the symptoms improve. The well-known antidote of opioids, Naloxone, is also rarely used. In palliative medicine it is used only when the respiratory effort is compromised (respiratory rate of less than 8). Under these circumstances, the Naloxone is diluted with Normal Saline (400 micrograms of Naloxone in 10 ml of Normal Saline) and a bolus of 20 micrograms (0.5 ml of the dilution) is administered intravenously. Further boluses of 20 micrograms are given in two-minute intervals until the respiratory rate is above 8(11). As the half life of opioids is much longer than that of Naloxone, repeated doses may be required, and a Naloxone infusion might need to be instituted.

Spinal Cord Compression

Spinal cord compression is generally seen in about 5% of the patients with spinal disease. Lung, breast, prostate cancer as well as lymphoma tend to metastasise to the spine with increased frequency(13),(14)(table 1/table 2) and the spread can be singular or multifocal. The development of spinal cord compression is generally a sign of poor prognosis but about one third of these patients will survive for over a year.

Site of compression	Percentage (from total number of SCC)
Thoracic	77%
Lumbar	29%
Cervical	12%
Sacral	7%

Primary site	Percentage (from total number of SCC)	Mean survival after the diagnosis of SCC
Breast	37%	14 months
Prostate	28%	12 months
Lung	18%	3 months
Other solid tumours	17%	variable

Table 1/Table 2: Demographics of spinal cord compression/The spinal cord compression as a poor prognostic factor(13),(14)

The importance of early recognition of the spinal cord compression is that early treatment will minimise the long-term effects of the syndrome as the damage to the spinal cord is irreversible. The main clinical symptoms are of severe back pain, peripheral weakness and sensory loss as well as bowel and bladder disturbances. The clinical examination will reveal upper motor neuron signs with a typical sensory level encountered in about 1 in 5 patients.

The investigation of choice is an MRI and an urgent referral should be made. In view of the risk of multifocal metastases and spinal cord compression on different levels an MRI of the whole spine is recommended. In many hospitals multi-disciplinary teams work closely in investigating, diagnosing and managing spinal cord compression and referral to those teams is highly recommended.

Some research suggests that administration of steroids is of benefit only in patients with motor symptoms(15). However if there is clinical suspicion of spinal cord compression steroids should be initiated immediately while awaiting further investigations. UK NICE guidelines recommend a daily dose of 16 mg of Dexamethasone which can be given orally or intravenously.

If the MRI confirms the presence of spinal cord compression a decision on the management is taken based on the patient's wishes and the long-term prognosis of the patient. Short courses of radiotherapy are preferred over more prolonged courses(16) , especially for patients with a prognosis of less than 6 months. Surgical interventions are only considered for patients with longer prognosis, a good performance status and a single metastasis.

Major haemorrhage

Bleeding can be seen in about 10% of cancer patients although the prediction of patients who might suffer a catastrophic bleed is very difficult. It is one of the most distressing emergencies. Head and neck tumours are more prone to bleed, but bleeding can occur in any malignancy and it can manifest as haematemesis or melaena, vaginal bleeding or major haemoptysis.

Different mechanisms, seen in patients with terminal disease can be involved, such as thrombocytopenia, disseminated intravascular coagulation or direct erosion of a major vessel by the growing tumour(17). The risk of a major haemorrhage can be heralded by the presence of previous episodes of minor bleeding and this should initiate discussions around the possibility of a catastrophic event with patients and their families.

In a hospital setting it is important to identify the patients at risk of a major haemorrhage at an early stage and rationalise their medication accordingly to minimise the risk, for example by withholding antiplatelets, anticoagulants and high-molecular heparins. Oral tranexamic can be used in patients able to swallow if episodes of minor bleeds are observed.

PALLIATIVE CARE EMERGENCIES FOR THE JUNIOR DOCTOR

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Correction of coagulation disturbances can also be considered if appropriate for the patient's condition, as well as local measures, such as local adrenaline(17). More importantly, the risk of a major haemorrhage needs to be communicated to the members of the team as well as the patient and their family and preparations for such a terminal event should take place.

It is good practice for dark towels to be kept close to the patient, to minimise visual distress to both the patient and the family, due to the potentially large volume of blood. Reassuring the patient and the family is paramount in reducing the anxiety levels. High doses of Midazolam can be used in cases of catastrophic bleeding, usually subcutaneously. An initial dose of 10 mg of Midazolam is advised, with subsequent doses as required. Opioids can also be used if pain is an associated symptom(18).

Good communication throughout, reassurance of the relatives and gentle approach after the event should also take place.

Seizures

Seizures are not uncommon towards the end of life. They are seen far more often in patients with primary or secondary brain disease with the incidence reported to be as high as 45% in patients with high-grade glioma approaching end of life (19), but they can also be seen in relation to metabolic disorders and in people with longstanding epilepsy. As the oral intake is reduced and oral medications are with-held due to severe dysphagia, the discontinuation of regular antiepileptic medication adds to the increased risk of seizures. In patients at earlier stages of their disease, the management of seizures differs to the management of dying patients.

During a seizure, common supportive measures, such as oxygen administration, are taken. Midazolam 5 mg subcutaneously is generally preferred in the palliative setting, but Lorazepam 4 mg intravenously or Diazepam 10 mg rectally are suitable alternatives. If the patient is in the early stages of the disease, introduction of antiepileptic medication is then considered, as well as the introduction of steroids if there is known brain disease. In patients approaching the end of life, an infusion of Midazolam at a dose of 20mg-30mg via a syringe driver is instituted. Appropriate alternatives are Phenytoin or Phenobarbital(20).

Discussion/Conclusion

We have described above a rough guide approach to palliative care emergencies but palliative medicine runs beyond any strict guidelines. We have excluded discussion of superior vena cava obstruction as this is covered elsewhere within this journal edition. Patients going through the palliative phase of their disease have complex needs, both physical and emotional more than in any other specialty.

A careful balance between the patient's wishes the stage of the disease and appropriateness of any intervention needs to be considered. The importance of good communication between teams involved in the patients' care as well as with the individual patient and their families cannot be emphasised enough. It is the key to a successful palliative care approach and management.

Questions

1. A 46 year old-lady on active treatment for metastatic breast cancer presents with confusion, nausea and vomiting. Blood tests reveal: Urea: 23.4, eGFR: 35, Albumin: 26, Calcium: 2.6. What is the most appropriate investigation/treatment?

- A. Rehydrate and proceed with chemotherapy.
- B. Urgent abdominal US.
- C. IV fluids.
- D. IV fluids and full dose of Biphosphonates.
- E. IV fluids with reduced dose of Biphosphonates

2. An 82 year old man with known prostate cancer and metastatic bone disease, calls the palliative advice telephone line late at night, complaining of severe back pain. He has a background of chronic back pain but his pain is more severe than usual, and radiates down his legs. On further questioning he also mentions difficulty in passing urine during the hours prior to his phone-call. The advice should be:

- A. Reassure the patient. His pain is chronic and probably unrelated to his cancer.
- B. Advise over the phone regarding analgesia and follow-up call if necessary.
- C. Advise him to seek urgent medical review that same night.
- D. Advise for a GP review the next morning to exclude a urinary infection.
- E. Advise for oral steroids and medical review the next day.

3. Mrs X, 68 year old lady with ovarian cancer, is an inpatient in the hospice for symptom control. She is on Oxycontin 80 mg twice a day. On the evening ward round she is administered Oxycontin 140 mg, the dose of lady S who is on the bed next to her. What are the priorities in her management?

- A. No need for monitoring; insignificant increase in the dose
- B. Stat dose of Naloxone followed by Naloxone infusion
- C. Immediate transfer to the acute hospital
- D. Monitor patient's symptoms. Her management will depend on the severity of symptoms.
- E. IV fluids

PALLIATIVE CARE EMERGENCIES FOR THE JUNIOR DOCTOR

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4. A 77 year old man with a recent diagnosis of locally advanced stomach cancer and multiple co-morbidities, is an inpatient in the acute hospital waiting for a package of care prior to his discharge. He has declined any active treatment and has clearly stated that he wants to concentrate on quality of life, as his prognosis is very poor. Suddenly, he deteriorates and soon after he suffers several episodes of large-volume haematemesis. The on-call team is called. Things to consider:

- A. Urgent referral to the gastroenterologists
- B. Urgent blood tests, correction of abnormalities of the coagulation, urgent transfusion
- C. Supportive measures for the haemodynamic instability
- D. Reassure the patient, communicate the news to the family and manage symptoms as appropriate
- E. Intravenous PPI

5. A 58 year old lady with glioblastoma is on prophylactic Phenytoin in view of recurrent seizures, the more recent leading to a hospital admission. Despite high doses of steroids her condition has started deteriorating and there are increasing concerns that she is aspirating when taking her tablets. Her prescription will need to be altered to:

- A. Discontinuation of Phenytoin and introduction of a syringe driver with 20 mg Midazolam/24 hours.
- B. Liquid Phenytoin.
- C. Continuation of her current tablets but change the timing so that she is awake enough when taking her medication.
- D. Discontinuation of anti-epileptic medication with close monitoring of symptoms. Benzodiazepines can be prescribed for PRN use.
- E. Switch to a different oral antiepileptic.

Answers

Question 1: Correct Answer E

The diagnosis is of hypercalcemia. On patients with low albumin the adjusted calcium needs to be calculated. The equation used is: $\text{total calcium} + (40 - \text{Albumin}) \times 0.02$. The corrected calcium for this patient, is above the normal limit, even if the total calcium is within normal limits. The presenting symptoms are typical for hypercalcemia.

The first step on her management is intensive IV rehydration, followed by the administration of biphosphonates, preferably of Zoledronic Acid. Biphosphonates' dose needs to be reduced in renal dysfunction. The deterioration in her renal function may reflect dehydration secondary to the hypercalcemia and may improve with rehydration. An US would not be a priority in this case. Hypercalcemia is a marker of poor prognosis and can reflect advanced disease or disease-unresponsive to treatment.

Question 2: Correct answer C

It is important to exclude spinal cord compression on this patient. Although his pain is chronic, its' character has now changed. The dysuria could be unrelated but in the first instance should raise the suspicion of spinal compression. He warrants urgent medical review to exclude signs of upper motor neuron compression.

Oral steroids need to be started immediately if there are clinical signs suggestive of spinal cord compression but it is important not to mask any signs prior to an urgent medical review. An urgent MRI would either exclude or confirm the diagnosis and determine further management, most likely urgent radiotherapy.

Question 3: Correct answer D

The management will depend on the symptoms. If there is no evidence of opioid toxicity, close monitoring might suffice. Fluids, either intravenously or subcutaneously can be administered if the patient experiences side effects such as increased drowsiness, hallucinations or confusion. Her respiratory rate will also need close monitoring, initially in 10-minute intervals. Naloxone will be needed if this rate was to drop below 8.

If she had signs of toxicity her next dose will need reducing, or even withdrawing if respiratory depression is observed. It is essential to consult senior support to help review and confirm the ongoing opioid regimen and to construct a firm management plan. An incident form should also be completed so that the drug error can be investigated.

This is a very serious incident and will need to be managed as such. The patient and their family will need to be informed and apologies given. It is vital to recognise that the involved staff will need to be supported throughout and following the incident.

Question 4: Correct answer D

The patient has already chosen not to have any active treatment. Although this is an unexpected deterioration, in view of the severity of the situation, the medical team need to recognise the irreversibility of the situation and act in accordance to the patient's documented wishes, as there are no concerns regarding his capacity.

Major haemorrhage is not infrequently a terminal event, and support of the patient and the family are the more important things to consider. Sedation of the patient may be required if the patient appeared to be distressed, usually with high doses of Midazolam. For patients discharged home this risk needs to be communicated to the family and to the community team to facilitate the early implementation of anticipatory measures.

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Question 5: Correct answer A

There is no need to consider a change in her regular antiepileptic to a different form of oral medication, as her swallowing will continue to deteriorate and her risk of aspiration will outweigh the benefit of treatment. It would be reasonable to start a driver with Midazolam to prevent any events. The starting dose can be low with an appropriate adjustment if appropriate. Prescription of Benzodiazepines on the PRN section needs also to be considered.

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REVIEW ARTICLE: FACILITATING RAPID DISCHARGE HOME FOR DYING PATIENTS

A Gorringe, D Feuer

Review Article: Facilitating Rapid Discharge Home for Dying Patients Teaching & Training

Introduction

Most patients would prefer not to die in hospital, yet over 50% deaths in the UK occur in hospital(1). The reasons for this are complex and multifactorial, but successful discharge from hospital for dying patients whose preferred place of death is home can often be achieved. It requires clinical assessment and identification of dying patients, clear communication with the patient and family, and close working with the multidisciplinary team both within the hospital and the community.

As a Foundation Year doctor you will be responsible for facilitating rapid discharge home for dying patients under your team's care. This article aims to provide practical guidance and a framework to help you achieve safe, successful discharges home for patients at the end of life.

Identification of the Dying Patient

Identifying when a patient may be starting to die is essential in order to formulate an appropriate care plan which ensures that the patient's wishes are met. The terminal phase is defined as the period when day to day deterioration, particularly of strength, appetite and awareness, is occurring(2). Should such symptoms develop suddenly over a matter of days instead of the usual weeks, it is important to exclude a reversible cause of deterioration such as infection, hypercalcaemia, or medication changes.

If potentially reversible problems are identified, it may or may not be appropriate to treat them – sometimes if a patient is clearly dying it will not be appropriate to treat a high calcium or infection. It can be difficult to diagnose dying, so it is important that decisions are taken by the multidisciplinary team, having reviewed the patient's longer-term disease trajectory, and involving the patient and family as far as possible. The clearest signs of approaching death are picked up by the day by day assessment of deterioration:

• Profound Weakness

- Bedbound
- Needs assistance with all care

• Diminished intake of food and fluids



• Drowsy or reduced cognition

- May be disorientated in time and place
- Difficulty concentrating
- Scarcely able to co-operate with carers

• Gaunt appearance

• Difficulty swallowing medicine

• Altered circulation

- Peripheries cold and/or mottled

Communication

For patients recognised to be in the last days or short weeks or life, good communication is essential to ensure that the person's needs and wishes are ascertained and met. The Leadership Alliance for Care of Dying People has set out 5 priorities for care, when it is thought that a person may die within the next few days or hours(3):

1. This possibility is recognised and communicated clearly, decisions made and actions taken in accordance with the person's needs and wishes, and these are regularly reviewed and decisions revised accordingly.
2. Sensitive communication takes place between staff and the dying person, and those identified as important to them.
3. The dying person, and those identified as important to them, are involved in decisions about treatment and care to the extent that the dying person wants.

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4. The needs of families and others identified as important to the dying person are actively explored, respected and met as far as possible.

5. An individual plan of care, which includes food and drink, symptom control and psychological, social and spiritual support, is agreed, co-ordinated and delivered with compassion.

A senior clinician should speak with the patient and family to explain that the patient is now dying and when and how death might be expected to occur. The patient, relatives and carers should have the opportunity to ask questions. Clear, contemporaneous records must be kept to facilitate ongoing discussions.

Care Planning and Community Services

Patients who have been unwell for some time may have made advanced care plans or stated preferred priorities of care, such as where they would like to be looked after, and you should ask specifically about these. They may have appointed a Lasting Power of Attorney or made an Advance Decision to Refuse Treatment. If so, a copy of these should be placed in the medical notes. In agreeing a care plan with the patient, relatives and/or carers, an important consideration is the patient's preferred place of care.

Options to discuss include hospital, home (including nursing home or care home) and hospice. It is important to have gathered information about the services available in the community so that you can discuss these with the patient and relatives. Your hospital palliative care team should be able to advise you of the services available locally and will support you in facilitating rapid discharge home to die. Typically services will include:

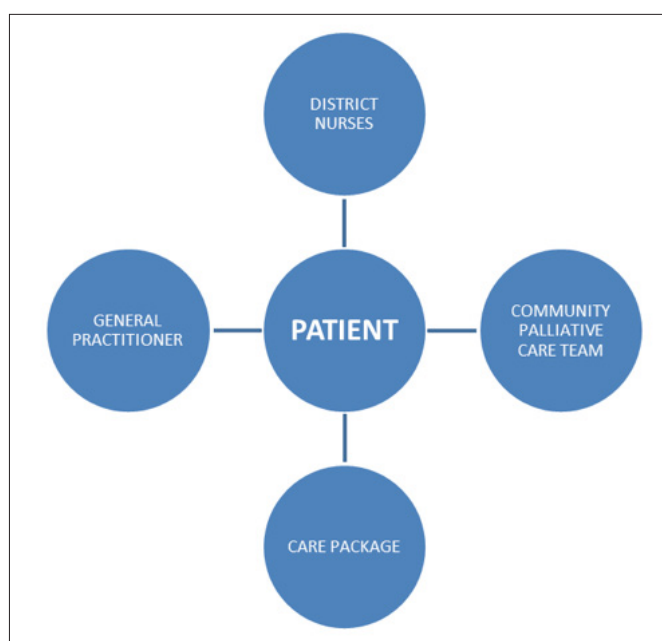


Figure 1: Services available to patients in the community.

Care package: carers usually able to visit a maximum of 4 times a day, though some regions have access to 24hr care. For patients with a short prognosis and medical need for care this will be funded by NHS Continuing Care, rather than social care, and can be applied for by completing the Fast Track Continuing Care Tool.

District Nurses: refer for administration of medication (including syringe drivers and subcutaneous injections), equipment ordering (for example hospital bed and mattress, commode, sliding sheets), daily symptom review, wound care, end of life care.

Community Palliative Care Team: provide a visiting multidisciplinary team including Clinical Nurse Specialists and Palliative Care Doctors who can advise on symptom control, provide support for patient and family, and act as a source of specialist knowledge for GPs and District Nurses. The community palliative care team do not typically provide hands-on nursing care at home. Some teams have a "hospice at home" service which can provide 24hr support from specially trained carers and/or nurses for patients who wish to die at home.

GP: the General Practitioner remains the clinician with overall responsibility for the patient's care in the community. They are also often the first port of call for bereaved relatives.

Patients whose preferred place of death is home

For patients whose preferred place of death is home, there are several clinical and practical steps that must be taken to facilitate rapid discharge. Good multidisciplinary working is essential to ensure a smooth discharge and maintain the patient's and relatives' confidence. The following checklist should help you:

Rapid Discharge Home Checklist

A member of the MDT to complete check:

- Ensure patient, supported by family, wishes for end of life care at home/nursing home. Facilitate discussions regarding prognosis, symptom management and services available in the community.
- Complete Fast Track Continuing Care Tool and Care Plan. Fax to relevant provider service (liaise with discharge team).
- Set up Care Package. Once Fast Track approved liaise with Continuing Care and District Nurses to organise care package.

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- Order Equipment. Ensure relevant equipment has been ordered and delivered.
- Refer to Community Palliative Care Team. If patient is already known to team, ensure update is communicated. Liaise with Palliative Care Team regarding plans for the syringe driver if continuous subcutaneous infusion in situ.
- Inform GP of discharge. Request a home visit as soon as possible on discharge to assess patient. The GP will be responsible for completing a death certificate so will usually need to have seen the patient within the last 2 weeks of life. Inform out of hours community services provider of patient discharge.
- Complete Electronic Palliative Care Co-ordination System (EPaCCs) if applicable. EPaCCs enable the recording and sharing of people's care preferences and key details about their care at the end of life. The records can be accessed electronically by community care providers including GPs and paramedics. There are a number of different models in use across the country, such as Coordinate My Care in London(4).

Doctor to complete check:

- Prescribe Medication for Discharge to include end of life care injectable medication and community authorisation charts. Community authorisation charts allow District Nurses to administer injectable symptom control medication. Refer to local guidelines and palliative care team for what to prescribe, but be sure to include anticipatory ('as required') oral/subcutaneous medication for:
 - Pain e.g. *diamorphine*
 - Agitation e.g. *midazolam*
 - Respiratory secretions e.g. *glycopyrronium*
 - Nausea/vomiting e.g. *metoclopramide*
- Assess for home oxygen requirements. Medication such as opioids and benzodiazepines can contribute to management of breathlessness for patients in the last few days of life. Assessment using pulse oximetry and providing oxygen to patients who did not previously require it may not be appropriate.
- Agree care plan with family and carers, and ensure they are aware of how to access support at home to prevent unnecessary readmission to hospital.

Nurses to complete check:

- Book transport. Consider if the patient requires a stretcher, oxygen, any difficulties transferring within the home (e.g. steps) and a DNA CPR order for the journey.
- Refer to District Nurses. Refer for administration of medication (including syringe drivers and subcutaneous injections), equipment ordering (for example hospital bed and mattress, commode, sliding sheets), daily symptom review, wound care, end of life care.

One day prior to discharge:

- Ascertain if discharge is still appropriate and still wanted by patient and family.
- Confirm equipment in place.
- Confirm Fast Track care package in place.
- Ensure District Nurses aware of patient being discharged.
- Ensure Community Palliative Care Team aware of patient being discharged.
- Ensure GP aware of patient being discharged.
- Confirm all discharge medications are correct.
- Confirm authorisation form for end of life medication is completed and faxed to District Nurses.
- Ensure syringe driver (if being used) is available prior to discharge.
- Ensure valid community DNA CPR form goes with the patient.
- Provide supply of needles, syringes, water for injection, dressings, pads, urine bottles if applicable.
- Confirm stretcher ambulance is booked.
- Provide patient/relatives with useful contact numbers including District Nurses, GP and Community Palliative Care Team.
- Ensure relatives are aware of what to do when the patient dies.

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Speaking to Family Members about What to Expect

With relatively few deaths occurring at home these days, many people haven't experienced seeing someone die and may not know what to expect. Your Trust may have leaflets to provide to families, but if not, you can refer them to the booklet "End of Life: A Guide", produced by Marie Curie(5). Here is some advice you can give relatives who are caring for a family member dying at home. Not everyone will display all of the signs, but this is often how the body prepares for the final stages of life(6):

Signs of approaching death	Advice
Sleeps more and difficult to wake at times.	Plan conversation times for when the person seems more alert.
Loses appetite and may "forget" to swallow.	Offer small servings of favourite foods or drink without forcing. At this time the body has minimal needs.
Becomes confused about time or may not recognise familiar faces.	Speak calmly. Remind the person of the day, time and who is in the room.
Becomes restless, pulls at bedclothes, has visions of people or things that aren't really there. They may develop a fixed stare.	Leave a soft light on in the room. Provide reassurance and avoid physical restrictions where possible. Even if the person cannot respond, don't assume they can't hear you - hearing is the last sense to be lost.
Loses control of bowels or bladder.	This does not usually occur until death is close. The amount of urine will decrease or stop as death nears. The district nurse can advise on how this can be managed.
Secretions collect at the back of the throat and sound like a rattle.	This is because the person cannot swallow saliva but does not mean they are uncomfortable. Turn the person on their side or raise the head of the bed. Sometimes medication can be given to help - ask the district nurse or your community nurse.
Arms and legs cool as the circulation slows down. Sometimes one side of the body will be warm and the other cold. Face becomes pale and feet and legs adopt a purple-blue appearance	Use just enough coverings to keep the person comfortable.
Breathing becomes irregular and even stops for short periods. The pulse becomes fast and irregular.	There is no need to become alarmed about this. It causes no distress to the patient.
When death occurs <ul style="list-style-type: none"> • Breathing stops • Heartbeat and pulse stop • Eyes may be open or closed • A 'last sigh' or gurgling sound may be heard • There is no sign of life 	
What to do if you think death has occurred: Try not to call 999 for an ambulance. You should call your GP's surgery and a doctor will come to certify the death and support you. They will also remove any medical equipment from the patient. (If the patient has not seen a doctor in the 2 weeks prior to death, the police may also come.) If you are uncertain what to do, call your District Nurse, GP or Community Palliative Care Team.	

Table 1: Advice for relatives caring for a dying patient at home

Care After Death

For patients who die at home, the GP will be responsible for completing a death certificate so will usually need to have seen the patient within the last 2 weeks of life. If possible, telephone the patient's GP before discharge so that they can prioritise the visit. Bear in mind that some communities require burial or cremation of the deceased within 24 hours.

This can sometimes be difficult to achieve, particularly if death occurs at a weekend as this can delay registering the death. Also remember that some deaths require discussion with the Coroner before a death certificate can be issued, such as those related to industrial diseases e.g. mesothelioma(7). It is helpful to speak with families in advance and prepare them for any potential delays.

Conclusion

Foundation doctors play a key role in facilitating rapid discharge home from hospital for dying patients. Achieving a patient's preferred place of care can be extremely important not just for the patient but also the relatives' experience of bereavement. Safe, successful discharges require careful planning and excellent communication with the patient, family and multidisciplinary teams, both in the acute and community settings.

MCQs for Review Article: Facilitating Rapid Discharge Home for Dying Patients

1. What percentage of deaths in the UK occur in hospital?

- Under 10%
- Under 30%
- Over 50%
- Over 60%
- Over 80%

2. When asked in advance, where do most people identify as their preferred place of death?

- Hospital
- Home
- Nursing Home
- Hospice
- No preference

3. The clinician with overall responsibility for a patient's care in the community is:

- General Practitioner
- Palliative Care Consultant
- Hospital team Consultant
- District Nurse
- Palliative Care Clinical Nurse Specialist

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4. Which of the following symptoms does not commonly occur in the dying phase?

- a. Pain
- b. Agitation
- c. Seizures
- d. Respiratory secretions
- e. Nausea and vomiting

5. For the purposes of issuing a death certificate, the certifying doctor should have seen the deceased within how long before death?

- a. 1 day
- b. 5 days
- c. 1 week
- d. 2 weeks
- e. 1 month

Answers

1. Answer: (c)

2. Answer: (b)

3. Answer: (a)

4. Answer: (c)

New onset seizures are uncommon in the terminal phase. The other 4 symptoms are common and anticipatory medication should be routinely prescribed for all dying patients.

5. Answer: (d)

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TIME TO TALK: DISCUSSING DO NOT ATTEMPT CARDIO-PULMONARY RESUSCITATION (DNACPR), COMMUNICATING DECISIONS & NEW GUIDANCE

J Lee, J Brady



Time to talk: Discussing do not attempt cardio-pulmonary resuscitation (DNACPR), communicating decisions & new guidance

Teaching & Training

Introduction

Cardio-pulmonary resuscitation (CPR) at its conception in the 1960s was intended to restart the heart following a sudden cardiac arrest due to a cardiac arrhythmia in the setting of myocardial infarction. Through the application of forceful chest compressions, high voltage defibrillation and intubation, a dramatic intervention could be made in preventing premature death in those with hearts 'too young to die.'

However, since its introduction, undergoing CPR appears to be becoming more common for those at the end of life. This is despite an increasingly aging co-morbid population, and knowledge that the chance of surviving to discharge and complete physiological recovery following an in hospital cardiac arrest has been shown to be fewer than 20% of adult patients. (4) Avoiding this requires a 'do not attempt cardio-pulmonary resuscitation' (DNACPR) decision to be made by physicians who have acknowledged that this violent intervention would be futile.

NCEPOD 2012 Report

In 2012 the National Confidential Enquiry into Patient Outcome and Death (NCEPOD) published their report reviewing in-hospital cardiac arrests, Time to intervene? (1) They concluded the following:

- *A high proportion of in-hospital deaths involved CPR attempts even when the patient's underlying condition and general health made it unlikely to be successful.*
- *Even in cases where cardiac arrest and death are likely, poor documentation of CPR status decisions may result in patients undergoing futile attempts at CPR during their dying process.*

Abstract

As a foundation doctor, it is inevitable that as you progress through your career, you will encounter patients who, despite our best efforts and treatment, will be at the terminal stages of life. Often the approach to dying people and do not attempt cardio-pulmonary resuscitation (DNACPR) decisions can feel inherently difficult, causing trepidation to physicians at all levels. Only by confronting these situations directly can dying people be treated appropriately and allowed to die with dignity.

Palliative care and discussions regarding DNACPR and its communication have been topical issues in healthcare and the media lately. Guidance has evolved as a result of in-hospital reviews, guidance following the cessation of the Liverpool Care Pathway, and related court case judgements. Notably, there are three particular important publications that focus on this issue: The NCEPOD 2012 report, (1) the Tracey judgement (2) and the 5 Priorities established by the Leadership Alliance for the Care of the Dying People. (3)

The objectives of this article are to explain why DNACPR discussions are important in the in-hospital setting, outline current guidance and reviews of practice, and to present a framework for approaching the DNACPR discussion.

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The NCEPOD identified a need to identify at an early stage those patients for whom CPR cannot prolong life but merely prolong the dying process, thereby allowing patients to avoid suffering and an undignified death. They reiterated that DNACPR should not negate patients receiving active treatment.

Consequently, they produced five principal recommendations following their assessment (figure 1). Their recommendations give clear direction that senior doctors should be helping juniors with identifying patients who need and making resuscitation decision, especially with acute admissions.

5 NCEPOD principal recommendations

CPR consideration & consultant review

All acute admissions must have CPR status considered and recorded ideally from admittance and certainly by first consultant review. If CPR is considered inappropriate consultant involvement must occur at that time.

Care before Cardiac Arrest

Escalation to a more senior doctor where patients deteriorate following non-consultant review. If not escalated reasons for this must be documented clearly in the patient notes.

Resuscitation status

Understanding by medical staff that patients can remain for active treatment but that in the event of cardiac arrest CPR attempts may be futile. Being for active treatment is not a reason to omit consideration and documentation of decisions regarding cardiac arrest.

Resuscitation attempt

An agreed hospital plan for airway management during cardiac arrest.

Period after the cardiac arrest

All CPR attempts in hospital to be audited with assessment of what proportion of patients should have had a DNACPR decision in place prior to the arrest rather than after the first arrest.

Figure 1: 5 NCEPOD Principal Recommendations (1)

The Tracey Judgement - Discussing with patients and families

In February 2011, Janet Tracey was diagnosed with lung cancer with an estimated prognosis of 9 months. She had explicitly expressed a wish to be involved in all her management discussions. Two weeks after her diagnosis she was involved in a major road traffic accident in which she sustained a serious cervical fracture and was admitted to hospital under the Neuro-Critical Care unit. Due to chronic respiratory problems she was placed on a ventilator but did not respond to treatment for her chest infection. Following two unsuccessful attempts to wean her off the ventilator Janet Tracey's consultants were faced with the decision of how to proceed.

The decision was made to take Mrs Tracey off the ventilator. At this time, clinicians recognised the need to make a decision regarding actions in the event of cardio-respiratory arrest. Prior to being weaned off the ventilator a DNACPR notice was completed. Mrs Tracey was successfully weaned off the ventilator and her condition appeared to stabilise.

However, Mrs Tracey's daughter became aware of the DNACPR notice and given that this decision had been unknown to patient or her family she was horrified and registered her objections. This led to the subsequent cancellation of the DNACPR notice 5 days later.

A couple of days later Mrs Tracey started to deteriorate. She expressed that she did not wish to discuss resuscitation. Following medical review a second DNACPR was completed with the agreement of the family. Mrs Tracey continued to deteriorate and she died on 7th March.

The family of Mrs Tracey took the case to the High Court for judicial review. In a judgement handed down by the Court of Appeal widely covered in the national media, (2) the court ruled that Mrs Tracey's human rights were breached because there was a failure to discuss a DNACPR decision with a patient who had capacity, who had previously expressed a clear wish to be involved in discussions about her care and treatment.

The consequence of this case is that there is now a legal obligation for health care professionals to demonstrate attempts to involve patients in discussions about DNACPR. There should be a presumption in favour of patient involvement and, if the patient is not involved, there needs to be convincing reasons not to involve the patient.

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Subsequent guidance from the joint statement released by the British Medical Association (BMA), Resuscitation Council and Royal College of Nursing stated that discussions about dying and DNACPR shouldn't be avoided to attempt to spare the patient distress unless there is a good reason to believe that this would lead to psychological harm to the patient. (5)

They also advised discussion with those identified as important by the patient unless a competent patient has explicitly declined this. It should be made clear that 'their role is to help to inform the decision making process, rather than being the final decision makers' and that the decision needs to be considered from the perspective of the patient's, and not the family's, wishes. If the patient has a LPA for health and are now not competent, then legally reasonable attempts to involve the named person in discussions is required. Ultimately, DNACPR decisions are the responsibility of the medical team, but should involve discussion with patients who have capacity to do so.

The 5 Priorities

The decision not to attempt resuscitation can be a clear decision in the face of medical futility in those with irreversible conditions or clinical frailty. Nonetheless, it remains only the initial step in ensuring good quality care for those at the end of life. Again on the front pages of the national media was a controversy in palliative care, on this occasion over the Liverpool Care Pathway. In response, the Leadership Alliance for the Care of the Dying People, a coalition of 21 national organisations was established in June 2014 to develop new standards and guidance. They published their guidance in 5 priorities that clinicians should focus on during the care of the dying patient. (3)

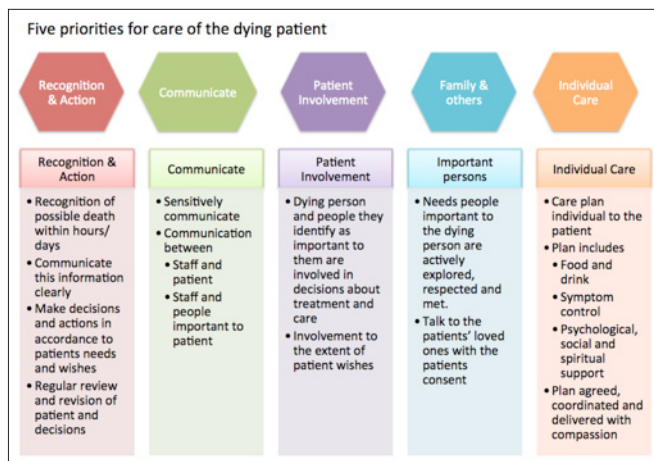


Figure 2: Five priorities for the dying patient, adapted from the Leadership Alliance for the Care of the Dying People. (3)

Case History - A real life challenge of DNACPR discussion

The situation

An 89 year old lady was presented to frail elderly multidisciplinary team (MDT) meeting. She had been diagnosed with a fungating breast cancer and was not a suitable candidate for further interventional treatment. She had further co-morbidities including frailty and dementia, although this did not yet have a formal diagnosis. She had multiple attendances to the Accident and Emergency department (A&E) over the preceding months. As she had expressed a desire not be in hospital she was presented with the aim of referring to community palliative care to explore her capacity and understanding of the situation and to make individualised future care planning.

However, before the MDT plan could be established she attended A&E again at night with dizziness and falls. After treatment in hospital she was transferred to a community hospital for rehabilitation. Two weeks later after failing to improve with rehabilitation, her family were obstructing discharge and refusing a DNACPR. Her son was adamant that she should not be DNACPR and threatened to file a complaint. He was confrontational, stating that he 'knows his rights' and had a history of making formal complaints against the health system. He demanded that the team continue her rehabilitation and that she should be 'made to do it' in order to recover.

The discussion

A family meeting was arranged with the palliative care consultant to raise concerns that she was nearing the end of her life, and to make plans given her expressed desire to not be in hospital.

During the meeting the patient's son was invited to explain his understanding of the situation. It became apparent from the conversation that he had received conflicting pieces of information from health care professionals regarding his mother's cancer prognosis. She had also never been formally diagnosed with dementia and she had been "labelled" without the patient or family being consulted about what the condition meant or entailed. To him dementia was about isolated profound memory loss and did not account for her changes in daily activity or general health.

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Each of the identified issues was acknowledged and discussed. It was explained that dementia is not just a memory illness and that it often co-existed with other and multiple comorbidities, including frailty,⁶ falls, infection risk and communication issues that affects prognosis. The natural course of her illness was also described, with an explanation of how the trajectory is of a slow global decline characterised by episodes of acute deterioration, which if she recovered from would create a new state below the pre-morbid baseline.

The Outcome

After discussing their concerns and the subsequent explanations the family agreed that they had thought she was dying but had been scared to acknowledge this. They expressed that they were relieved that someone else had said what they were thinking and felt it was no longer them 'giving up on her'. They now felt like they understood and could agree with their mother's choice to 'go home'. 'Home' was further discussed.

It was agreed that fast track to a nursing home, that could meet her medical needs and allow her to be near family, was the option in the patient's best interest given that she lacked the capacity for the decision. Ceiling of treatment and escalation decisions were set and agreed on involving oral antibiotics but no intravenous medicines, radiotherapy or CPR. The son apologised for being obstructive and was co-operative once he had understood the situational whole picture rather than just a focus on DNACPR discussions.

Approaching the discussion

In this case there were challenges presented with talking to the patient's loved ones about end of life care planning and DNACPR decisions. Polarising views existed between the clinicians and family's understanding about her current situation resulting in differing opinions on what represented her best interests.

Every good conversation starts with good listening. Until the issues and concerns raised by the family are acknowledged it is hard to enact change that will improve the disagreements arising in end of life planning. Listening to patients and their families often reveals existing conflicts in understanding, which may have led to differences in understanding the clinical situation.

There was also an underlying fear the son had of 'giving up on her' and when this was realised and addressed it helped explain some of the relatives' behaviour and difficult interaction with the doctors. Often DNACPR decisions represent the last frontier of hope for patients and their families. Therefore, it is important that DNACPR should come at the end of a wider exploration of understanding and decisions to help signal to patients and families that it is not a 'final chance to intervene' but part of an inevitable progression of disease. In this case, realising that putting the patient through CPR was likely to prolong suffering without any prognostic benefit the family were able to come to terms with the situation and participate in the discharge planning.

Ultimately, in discussions about resuscitation and end of life care communication is key. It is important to be open and sensitive. It should be clearly but sensitively sign posted that patients are actively dying without the use of innuendos that may confuse patients and families. Decisions about DNACPR and end of life are sensitive topics to all of us, so embracing the conversation early and starting at the beginning but with the end in mind will prove beneficial to clinicians, patients and their loved ones.

Our advised approach (Figure 3) incorporates elements of the discussed guidance and gives you a systematic approach to discussing with patients and their loved ones. The ABCD approach will remind you to advance plan, check baseline understanding of involved parties, to communicate sensitively and openly and finally to document the appropriate decisions.

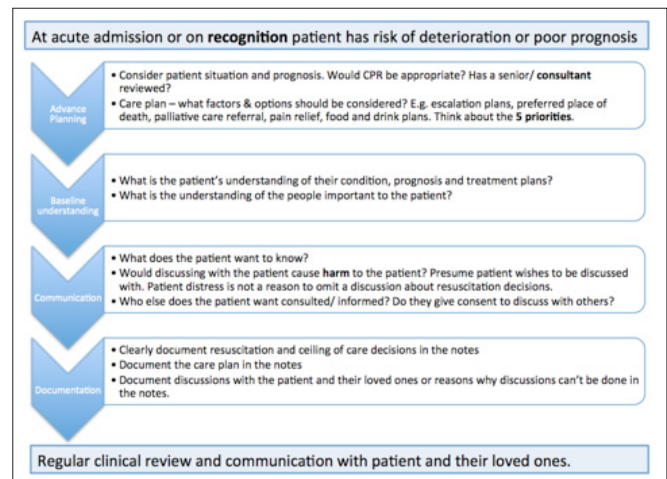


Figure 3: Our advised ABCD approach diagram for clinical approach to the dying patient by clinicians with recent guidance in mind.

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Summary Points

- Cardio-pulmonary resuscitation was originally developed as a means to prevent primary cardiac death in patients who were otherwise healthy with little co-morbidity. Since its establishment it has formed the default conclusion in the pathway of hospital deterioration.
- For some people, progressing to CPR without intervention or acknowledgement that a patient is dying is merely prolonging the dying process.
- Following the Tracey Judgement, there is now a legal obligation on clinicians to attempt to discuss DNACPR with a presumption that the patient and their loved ones want to discuss.
- The joint statement by the BMA, Royal College of Nursing and Resuscitation Council states that end of life and DNACPR conversations should not be avoided to spare patients distress unless it leads to patient harm.
- DNACPR discussions with patients and families can be inherently challenging. Approach DNA CPR discussions openly and sensitively. Allow DNACPR discussions to come at the end of a wider exploration of understanding and decisions. In positions of differing opinions with patients and family consider where the conflict in understanding exists.

Self Test SBAs

1. Which of the following is false regarding cardio pulmonary resuscitation?

- CPR was originally developed to prevent unexpected premature deaths for patients suffering primary cardiac conditions.*
- There are poor recovery rates, with evidence demonstrating rates of less than 20%, for surviving to discharge following an inpatient cardiac arrest.*
- Patients can demand and consent to being for CPR*
- Decisions and discussions regarding CPR should be clearly documented.*
- Patients can make advanced decisions to decline CPR as treatment for cardiac arrest*

2. Which of the following is true about discussing DNACPR?

- Only consultants should discuss DNACPR with patients and families*
- If the clinician feels that the discussion of DNACPR would cause distress to the patient then it need not be discussed with the patient*
- If the family feels that the discussion of DNACPR would cause distress to the patient then it need not be discussed with the patient*
- Clinicians should discuss DNACPR with patients with a presumption that the patient wants to be involved.*
- The patient's elected next of kin should be allowed to make the decision regarding DNACPR if the patient doesn't wish to discuss*

3. Which of the following is false regarding the current legal framework associated with DNACPR?

- There is a now a legal obligation to attempt to involve patients in discussions about DNACPR decisions.*
- If the patient is not involved in discussions about DNACPR then there needs to be convincing reasons for not involving them.*
- If the patient's family disagree with the DNACPR decision, then the family's decision must be respected and DNACPR revoked immediately.*
- If the patient's family disagree with the DNACPR decision, then the clinicians' decision should stand but the family can be offered a second opinion.*
- If patient is capacitous and wishes the DNACPR decision not to be discussed with their family then it should not be discussed with the family.*

4. Which of the following is true regarding second opinions about DNACPR?

- Every patient's DNACPR decision in hospital needs a second opinion from a medical consultant.*
- If a decision about DNACPR has been made in a multi-disciplinary team meeting with more than one consultant in agreement a second opinion is not legally obligated to obtain if the patient and loved ones are not in agreement, however it remains good practice to offer.*

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c. If a patient disagrees with a clinician's decision about DNACPR there is no need for a second opinion because the patient has not given consent to be DNACPR.

d. If a second opinion is being sought for decision about DNACPR there is no need to document the first decision as it is not valid until verified by the second opinion.

e. If there is disagreement between the patient's family and clinicians about a DNACPR decision that cannot be resolved following discussion then the next step is to obtain a second opinion that must be sought from the court

Answers

1. Answer C

Like other forms of medical treatment patients with capacity are allowed to refuse medical treatment but are not in a position to request medical treatment.

2. Answer D

Following the Tracey judgement there is now a legal obligation to discuss DNACPR decisions with patients and their loved ones with a presumption of wanting to be involved in the discussion. Patients can choose not to discuss and capacitous patients can request for discussions not to be made with their family if they wish.

3. Answer C

Disagreements between patients or their loved ones and clinicians should ideally be resolved through communication. However legally clinicians make DNACPR decisions, which are medical decisions, and discussions are to involve patients and family in best interests, not to make decisions about offering medical treatment.

4. Answer B

In positions of conflict between clinicians and patients and their families regarding DNACPR a second opinion can be sought. If the decision has been made and agreed in a multidisciplinary meeting with a number of consultants present then the decision of the MDT can be considered to have included a second opinion already.

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PARAPHARYNGEAL ABSCESS

M Gilmartin, Eslam Osman



Parapharyngeal Abscess Patient Management

Initial investigations ordered included blood analysis, urine analysis, OPG and CT with contrast. Significant results included a neutrophilic leukocytosis (WBC 15.5×10^9 per L, 98% neutrophils), a random glucose of 18.8mmol/L, metabolic ketoacidosis (pH 7.296, ketonuria), hypokalaemia (3.33mmol/L) and anaemia (Hb 99g/L). Initial management included making the patient NBM, inserting a nasaogastric tube, and administering IV paracetamol, IV saline, IV flucloxacillin and IV metronidazole. The CT with contrast revealed a 2 cm abscess in the right parapharyngeal space which extended to the level of the hyoid bone (Figure 1 and Figure 2).

Abstract

We report the case of a 77 year old female who presented to our department with a sore throat, dysphagia, and right neck swelling. A contrast CT of the neck detected a 2cm abscess in the right parapharyngeal space. Due to deterioration of the signs and symptoms, we planned to drain the abscess through a transcervical approach. On direct pharyngoscopy prior to the neck incision, we found the abscess draining into the oropharynx. The tract of pus was followed into the parapharyngeal space, and the abscess was drained intra-orally. We report this case to increase the awareness of the importance of direct pharyngoscopy prior to drainage via the transcervical approach.

Case History

A 77 year old female, with a past medical history of Type 1 Diabetes Mellitus and hypertension, presented to the A&E department with a 2 week history of sore throat, dysphagia and right neck swelling. She was initially seen by the maxillofacial team for a suspected dental abscess, who then referred her on to the ENT team.

She did not appear well, with a temperature of 38.7 $\text{\textcircled{D}}$. Upon examination, she had a right tender neck swelling (Level II), trismus, restricted head movement and a hoarse voice. On flexible naso-endoscopy, she was noted to have a patent airway, normal looking tonsils, and pooling of saliva in the hypopharynx.



Figure 1: Sagittal View (parapharyngeal abscess circled).

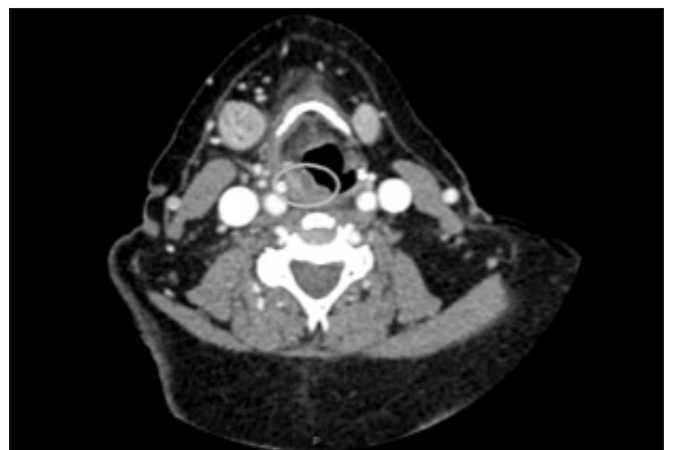


Figure 2: Axial view (parapharyngeal abscess circled).

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An emergency theatre session was booked. A sliding scale insulin protocol was started. The patient was consented for direct pharyngoscopy and an incision and drainage of the right parapharyngeal abscess. The patient was anaesthetised using awake fibre-optic nasal intubation.

Direct pharyngoscopy revealed an abnormal looking right tonsil with pus coming from the space between the tonsil and anterior pillar. The track of pus was followed laterally into the parapharyngeal space. The track was widened carefully, and the pus was drained intraorally. Pus was sent for microscopy, culture and sensitivity testing. A biopsy was additionally taken from the right tonsil.

Postoperative recovery was unremarkable. Trismus severity reduced significantly, and oral feeding was recommenced 2 days post-op. The abscess culture results came back positive for *Streptococcus Milleri* and mixed anaerobes. The patient's antibiotic prescription of IV flucloxacillin was subsequently changed to IV Penicillin G.

The histology results showed lymphoid hyperplasia and chronic inflammation of the tonsil. After the patient's glucose levels were stabilised, she was discharged from hospital, and booked into a regular outpatient clinic appointment. There have been no post-operative complications in the 3 months following surgery.

Discussion

This patient presented with symptoms typically associated with a deep neck infection and was identified promptly for swift investigation.[1] In adults the most common site for a deep neck abscess to form is the peritonsillar space, followed by the parapharyngeal space.[2]

The pre-operative management was suitably thorough, with detailed initial and on-going evaluation of the patient's airway. Previously a tracheostomy might have been considered gold-standard, due to the threatening nature of the growing swelling. However, our patient was found to have a patent airway on flexible naso-endoscopy, and a tracheostomy was not considered. CT scanning with contrast enhancement remains the best imaging modality in this situation. It can reveal the site, extension and potential source (dentition) of the abscess. Determining whether the abscess is pre or post styloid in location is an important consideration.[3]

A post styloid abscess risks invasion of the internal carotid artery, internal jugular vein, cranial nerves IX, X, XI, XII, and many other important structures. Additionally, the parapharyngeal space's relationship to the prevertebral space also enables a potential conduit for the infection to spread to the mediastinum.

These anatomical relationships demand that parapharyngeal abscesses be treated rapidly. This reduces the risk of life threatening complications (e.g.: mediastinitis, jugular vein thrombosis, Lemierre's syndrome and carotid artery haemorrhage).[4] In our case, the CT scan confirmed the diagnosis and was helpful in abscess localisation.

The use of broad-spectrum IV antibiotics for parapharyngeal abscesses is widespread. This patient initially received IV flucloxacillin and IV metronidazole. The abscess culture results came back positive for *Streptococcus Milleri* and mixed anaerobes. Her antibiotics were subsequently changed. 66.7% of isolated abscesses contain beta-lactamase producing organisms, resulting in many published protocols advocating the administration of IV co-amoxiclav.[5]

Clinicians also take into consideration the fact that many abscesses are polymicrobial, aerobic and anaerobic, necessitating appropriate antibiotic cover. Some centers have also advocated the use of IV steroids to reduce pain and other oral symptoms such as trismus; however, our patient did not receive IV steroids.

Many abscesses do not warrant surgical intervention. A recent study demonstrated that 48% (13/27) of paediatric patients with an abscess <2.5cm (CT scan) resolved with antibiotics alone.[6] Other small adult case series have demonstrated that antibiotics alone are feasible in specific situations. Anatomical location is also an important consideration: specifically whether the abscess is post or pre styloid (posterior, anterior). One adult case series showed that, of the 22 patients with a post styloid abscess location, the majority resolved with antibiotics alone.[7]

This study postulated that posterior parapharyngeal abscesses were probably the result of acute lymphadenitis and therefore bound by a capsule. Anterior styloid abscesses were considered more likely to be surgical emergencies. This is because the infection causes liquification of the pre-styloid fat. Deterioration of such a structure results in no border limiting the extension of pus into other deep neck spaces.

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Overall, the decision to operate or not warrants the consideration of many factors. In our case, additional thought was given to her immunosuppressed state (Type 1 DM and apparent ketoacidosis), and the fact that she did not immediately respond to antibiotics. In clinically stable patients, antibiotics alone are not a common initial management, followed if necessary by surgery.[8]

Traditionally such an abscess would warrant drainage through a transcervical approach. However, the cervical approach has been associated with a greater risk of complication, notably damage to the marginal mandibular nerve and pseudoaneurysm of the carotid artery[2].

Operating on an infected neck is technically difficult and associated with a prolonged postoperative recovery. We recommend that all patients listed for drainage via the transcervical approach have direct pharyngoscopy prior to the procedure. In some cases the abscess may be drained intraorally, sparing the patient from those risks associated with the transcervical approach. Today surgeons can accurately localise many abscesses radiographically and determine with good accuracy whether such an approach is feasible.

MCQs

1. Parapharyngeal abscesses can cause the following complications except:

1. Airway obstruction
2. Mediastinitis
3. Lemierre's syndrome
4. Jugular vein thrombosis
5. Peritonitis

2. Parapharyngeal abscess may be treated with:

1. intravenous antibiotics
2. transcervical drainage
3. intra-oral drainage
4. steroids
5. all of the above

3. The following investigations may be used in the management of parapharyngeal abscess except:

1. CT neck
2. US neck
3. Ig E RAST test
4. FBC
5. MRI neck

4. Acute tonsillitis if not treated, may lead to the following except:

1. Peritonsillar abscess
2. Laryngeal carcinoma
3. Parapharyngeal abscess
4. Retropharyngeal abscess
5. Airway obstruction

5. The parapharyngeal space contains the follow structure except:

1. Cranial nerve X
2. Internal carotid artery
3. Cranial nerve IX
4. Cranial nerve VIII
5. Cranial nerve XI

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Answers

1. 5 is the best option.

Peritonitis unlikely to be caused by a parapharyngeal abscess as the peritoneum is in the abdomen!

2. 5 is the best option.

All options are possible, as detailed in the discussion previously. As the complication is rare, there are few large studies comparing their relative efficacy however.

3. 3 is the correct option.

An Ig E RAST test is more important for patients being investigated for suspected allergy. A CT is first line, but you may also consider MRI and ultrasound.

4. 2 is the correct option.

Laryngeal carcinoma is not known as a complication of tonsillitis. Other common complications of tonsillitis include the options stated after the questions. Rarer complications include rheumatic fever, Sydenham's chorea, glomerulonephritis, scarlet fever and obstructive sleep apnoea.

5. 4 is the best option.

The vestibulocochlear nerve originates from the cochlea and vestibular system of the inner ear, and runs from the temporal bone in the internal acoustic meatus, to the brain stem.

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E.T. & ENT: FOREIGN BODY MANAGEMENT IN OTOLARYNGOLOGY

E Gosnell, P Murthy



E.T. & ENT: Foreign Body Management In Otolaryngology Patient Management

5 MCQs (Best of 5)

Which aural foreign body requires urgent removal due to the potential damage it can cause to structures?

- paper
- coin
- button battery
- marble
- pea

Which of the following foreign bodies can possibly be managed without input from Otolaryngology?

- coin in the oesophagus
- bead in the ear
- ingested bleach
- glass penetrating tympanic membrane
- intranasal magnets

Which of the following tools or techniques is not routinely used for removal of an intranasal foreign body?

- Parental kiss
- McGills forceps
- Tilley's forceps
- Wax hook
- Jobson-Horne probe

Which of the following are not complications associated with foreign body aspiration?

- Pneumonia
- Lung abscess
- Bronchiectasis
- Haemoptysis
- Haematemesis

Abstract

A foreign body sounds like some extra-terrestrial being but is merely an object or piece of extraneous matter that has entered the body by accident or design. Foreign bodies in the ear, nose and throat commonly present to the Emergency Department.

Most patients can be managed without specialist input but it is important to recognise the limitations within an emergency department and those patients that require urgent referral. Aspirated foreign bodies can be an airway and therefore life-threatening risk and need specialist input, as do patients who have foreign bodies with the potential to cause significant damage to normal structures such as button batteries, magnets and corrosive chemicals.

For all assessments and procedures you will need a good light, a cooperative patient as well as appropriate equipment. The first attempt will usually be the best tolerated, so if you are not confident or do not have appropriate equipment refer to an Otolaryngologist for more experienced help. Whenever a foreign body is identified, particularly in children, it is important to check other sites (i.e. both ears and both nostrils) to ensure there are no more foreign bodies hiding anywhere else!

E.T. & ENT: FOREIGN BODY MANAGEMENT IN OTOLARYNGOLOGY

E Gosnell, P Murthy

Where do ingested fish bones frequently get lodged in adult patients?

- Intranasal
- Tonsil
- Tongue
- Epiglottis
- Vocal cord

A foreign body sounds like some extra-terrestrial being but is merely extraneous matter that has entered the body by accident or design. Foreign bodies can range from harmless beads in the external auditory canal to ingested dentures and even life-threatening aspirated grapes. As an Otolaryngology trainee we are often referred patients, particularly of the paediatric age group, many of whom can be managed in the emergency department or in outpatient clinics, but recognising patients who need specialist input can be challenging. We hope to give you some tips on when to suspect foreign bodies and how to practically manage patients.

Foreign bodies in the ear

Foreign bodies of the pinna usually arise from embedded pierced earrings, which may be located in the ear lobe or the cartilaginous portions of the pinna. These patients commonly present with ear pain, swelling, redness and purulent discharge from the piercing.

The infected, embedded jewellery should be removed promptly; typically this is done under local anaesthetic but might require sedation or general anaesthesia in very young patients. Perichondritis and chondritis may complicate pinna foreign bodies, particularly when the cartilage has been pierced. *Staphylococcus aureus* and *Pseudomonas aeruginosa* are common organisms and systemic antibiotics are necessitated (refer to local antibiotics protocol).

UpToDate [1] states foreign bodies of the external ear canal are more common in children aged up to 6 years old. Common objects include beads, small toys, pebbles, tissues and paper. In adult patients it is more common to find cotton buds, insects or hearing aid components.

The type of foreign body determines the timing for removal

Button batteries, often found in toys and hearing aids, can be corrosive and require urgent removal by an Otolaryngologist. A live insect moving in a child's ear canal can cause considerable discomfort and occasionally may damage the tympanic membrane and middle ear. Insects should be killed with mineral oil, ethanol or lidocaine prior to attempted removal to prevent excess insect movement during retrieval. Foreign bodies that might have penetrated the tympanic membrane and caused damage to middle ear structures also require immediate attention whereas other canal foreign bodies can be deferred.

Often patients with ear foreign bodies are asymptomatic and can report the incident themselves or a parent may have witnessed the insertion. Some patients present with pain, deafness, bleeding or unilateral discharge. Oscopic visualisation will confirm the presence of a foreign body and allows examination of the external ear canal and tympanic membrane to ensure that there is no penetrating injury that requires specialist input. Using a headlamp will ensure sufficient light to directly visualise the object during removal.

Soft foreign bodies, such as cotton wool, may be grasped with a pair of crocodile or Tilley's forceps, whereas solid foreign bodies, such as beads, are best removed by passing a wax hook or Jobson-Horne probe beyond the object and pulling it towards you. Children need to be cooperative and it would be appropriate to refer to senior staff or consider a general anaesthetic if you have an uncooperative child, inappropriate equipment, failed attempt or suspected trauma to the eardrum.

Foreign bodies in the nose

Intranasal foreign bodies are most common in young children and often present asymptotically after witnessed insertion of inorganic objects such as beads and small toys. Porous nasal foreign bodies such as paper or foam as well as organic matter such as carrots and peas are also common but are often associated with a unilateral purulent and foul-smelling discharge. Other symptoms can include unilateral nasal obstruction, epistaxis and unilateral vestibulitis.

Removal of an intranasal foreign body is an elective procedure and the concern of migration of an inert nasal foreign body through the nasopharynx followed by aspiration into the trachea is not warranted in a normal healthy patient with intact airway reflexes. UpToDate [2] states the estimated risk for this complication is less than 6 in 10,000 cases and there are no reports of bronchial foreign bodies spontaneously arising from nasal objects in the literature. Button batteries can be associated with black purulent discharge, epistaxis, facial pain and swelling. The corrosive nature of batteries can cause significant damage to nasal structures, as can magnets that are attached across the nasal septum, and require urgent removal by an Otolaryngologist.

An auroscope can easily be used to examine a child's nose. Visualisation of foreign bodies establishes the diagnosis. Warner et al. [3] tell us they are most commonly located on the floor of the nasal passage just under the inferior turbinate or superiorly in the nasal cavity just in front of the middle turbinate.

Patients with soft or smooth nasal foreign bodies that totally occlude the anterior nasal cavity can be removed using positive pressure techniques such as asking the child to blow their nose or the 'parental kiss'. After explaining the procedure, the parent should place their mouth over the child's open mouth forming a firm seal.

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After occluding the unaffected nostril with a finger, the parent blows a sharp short puff of air into the child's mouth. There will be resistance caused by the closure of the child's glottis and the air will pass through the nasopharynx and out through the unoccluded nostril; hopefully dislodging the foreign body! Non-occlusive foreign bodies in the anterior nasal cavity can be removed with instrumentation: crocodile or Tilley's forceps for soft objects, passing a wax hook or Jobson-Horne probe beyond solid objects and gently pulling it forward.

Instrumentation of the nose may cause some trauma and brief epistaxis, which is typically controlled with direct pressure. For most patients, referral to ENT is not necessary; equipment such as a sheet for wrapping children and using topical anaesthesia or vasoconstrictor nasal drops can increase the success rate for removal. Potential indications for referral to Otolaryngology include posterior, impacted, or penetrating foreign bodies and those that cannot be removed due to poor cooperation, bleeding or inappropriate equipment.

Foreign bodies in the throat

Foreign bodies in the throat can be either aspirated or ingested and it is important to establish the former immediately due to the risk of airway compromise.

The presence of asphyxia indicates the need for immediate resuscitation and examination of the airway with input from both ENT and Anaesthetics. Basic life support manoeuvres are age dependent but include back blows, chest thrusts and abdominal thrusts. If there is no relief, direct laryngoscopy should be attempted by specialists to remove the foreign body. Persistent obstruction from a foreign body above the vocal cords may warrant intubation, accepting advancement of the foreign body into the right main bronchus for later surgical removal or needle cricothyrotomy to bypass the obstruction.

Foreign body aspiration should be suspected in children, especially those who have sudden onset of lower respiratory tract symptoms, as well as those who do not respond to standard management of other suspected aetiologies such as asthma, croup or pneumonia. A history of choking is highly suggestive but episodes can be un-witnessed or unrecalled and hence the absence of a history of choking does not rule out foreign body aspiration.

In the absence of asphyxia, radiographic studies such as chest X-ray and lateral neck soft tissue X-ray may aid in diagnosis, but normal studies do not exclude the presence. If foreign body aspiration is suspected, patients should be referred immediately to ENT for an experienced operator to perform bronchoscopy under general anaesthetic. Diagram one, taken from UpToDate [4] shows the natural course of foreign body aspiration, including the complications of acute or recurrent pneumonia, lung abscess, bronchiectasis and haemoptysis.

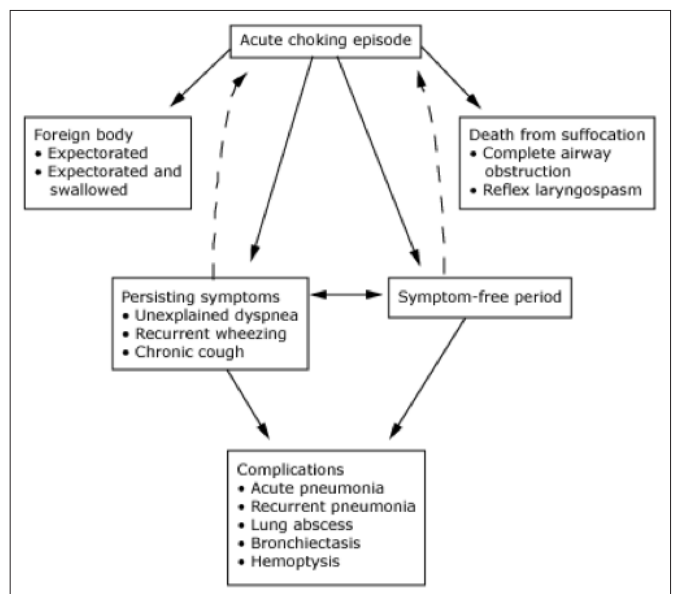


Diagram One: Natural course of foreign body aspiration.

Ingested foreign bodies commonly include fish, chicken or lamb bones in adults. If sharp objects e.g. razor blades, magnets or batteries are ingested urgent intervention is indicated. Patients present with acute onset of symptoms including constant pricking sensation on every swallow, drooling, dysphagia and localized tenderness in the neck. A potential complication of ingested foreign bodies includes oesophageal perforation which can cause mediastinitis and thoracic abscesses. Symptoms and signs include retrosternal chest pain, pyrexia, tachycardia and surgical emphysema.

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Examination with a good light as well as palpation of tonsils and tongue base (common areas for objects to lodge) can often allow identification of foreign bodies. Anaesthetizing the throat using Xylocaine spray can often assist examination as well procedures to remove visible foreign bodies. Initial investigations include an AP and lateral soft tissue X-ray of the neck (and chest if appropriate) looking for foreign bodies at common sites such as the tonsil, tongue base and posterior pharyngeal wall.

Flecks of calcification around the thyroid cartilage are common on X-ray but some foreign bodies are radio-opaque and can clearly be seen and localized using X-rays. Oesophageal perforations and surgical emphysema can also be seen on X-rays. Visible foreign bodies in the mouth, tongue base or pharynx can be removed with Tilley's forceps or McGill's forceps. Patients with airway compromise, failed removal, radiological evidence of a foreign body or a good history of a foreign body but not visible on examination should be referred to ENT for specialist input, requiring either flexible nasendoscopy or endoscopy and removal under general anaesthetic.

UpToDate [5] states ingested foreign bodies are very common in children aged 6 months to 6 years, with common objects including coins, toy parts and marbles. Fortunately most pass spontaneously and only a small proportion requires endoscopic removal. Foreign bodies such as batteries, magnets or corrosive substances for example bleach or cleaning products require urgent referral. For certain objects, X-rays and even CT scans are useful to assist locating the foreign body but urgent referral to ENT before arranging imaging is advised due to the potential damage associated.

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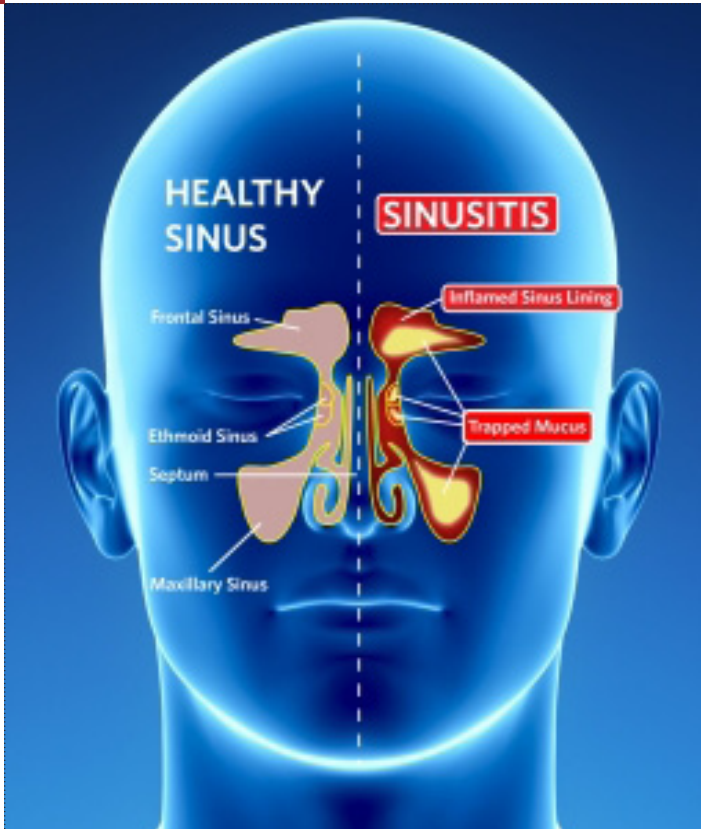
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PERI-ORBITAL CELLULITIS AS A COMPLICATION OF SINUSITIS – A CASE BASED DISCUSSION

FRM Shelton, M Pringle



Peri-Orbital Cellulitis As A Complication Of Sinusitis – A Case Based Discussion Patient Management

With a history of possible sinogenic symptoms ENT input was requested early. Examination revealed marked periorbital erythema and swelling of the right eye with proptosis, minimal eye opening, and chemosis. There was no disturbance of vision with colour vision intact, however the range of ocular movement was reduced. Flexible nasoendoscopy was attempted but not tolerated by the patient. The patient was systemically well with no evidence of central complications.

Blood investigations demonstrated an infective process with raised white cell count (WCC) 11.5 and raised CRP 45. Renal function was normal. With marked proptosis and reduced range of ocular movement there was a suspicion of a periorbital abscess and therefore urgent computer tomography (CT) of the sinuses and orbits was organised. This confirmed a sub-periosteal abscess with right unilateral sinusitis and dehiscence of the lamina papyracea. See

Abstract

Peri-orbital and orbital cellulitis is an infective process affecting the soft tissues anterior to the orbit and/or the orbital content (1). The incidence of this condition varies in literature from 0.3 -1.31 per month at specialist tertiary centres (2). Although the condition may occur in adults the incidence has been more widely seen in the paediatric population. The most common source of infection is from the paranasal sinuses, with reported rates of 60-90% of cases, but can also occur secondary to trauma, insect bites, conjunctivitis or blepharitis (2, 3).

We present a case based discussion of sub-periosteal abscess in an adult as a complication of acute sinusitis. The assessment, classification and management of this potentially devastating disease will be addressed.

Case study

A previously fit and well 70 year old female presented acutely to the ophthalmology department of a district general hospital with a 10 day history of swollen, erythematous, painful right eye. She also had symptoms of a viral upper respiratory tract infection with nasal congestion and nasal discharge predominantly worse on the ipsilateral side. There was no significant past medical history however she did describe a 30 year history of nasal congestion and facial discomfort.

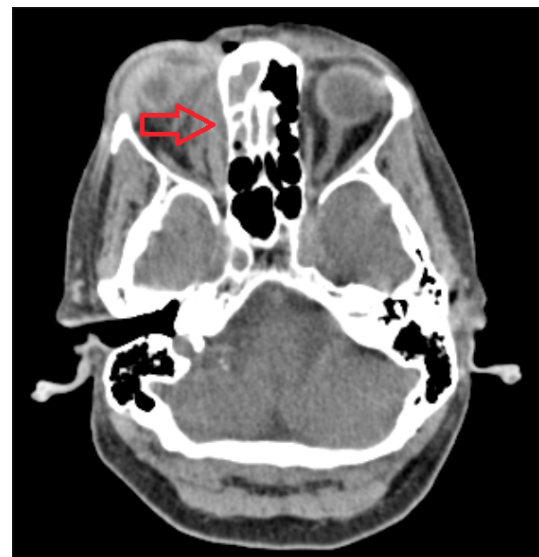


Image 1: Axial non-contrast CT of the orbits and ethmoid sinus. Proptosis and inflammation of the soft tissue in the medial wall is seen with an arrow pointing to the sub-periosteal abscess. Inflammation of the right ethmoid sinuses is also apparent.

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With evidence of an abscess, urgent external drainage of the sub-periosteal abscess and sinus washout was undertaken. The abscess was identified and drained with a corrugated drain to allow further drainage. Image 2 demonstrates post-operative clinical findings the following morning.

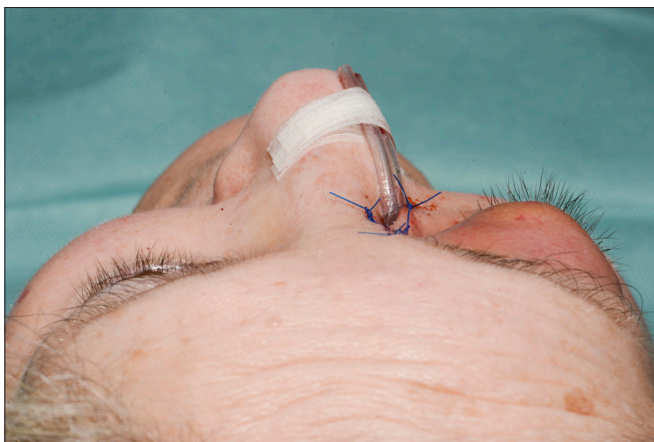


Image 2a and 2b: Clinical photographs of the patient taken the morning after emergency admission and surgery. The erythematous and swollen right eye can be seen with evidence of chemois in image 2a whilst image 2b demonstrates the proptosis.

Pus swabs from the surgery were sent for microbiology, cultures and sensitivity (MC&S) and grew a *Staphylococcus Aureus*. Ceftriaxone and metronidazole were started on admission and results of the pus MC&S confirmed these to be sensitive. Due to significant allergies to penicillin and erythromycin microbiology advice and the ceftriaxone was switched to linezolid. Nasal decongestants were also prescribed to allow further drainage.

Good outcome was obtained and the patient was subsequently discharged with follow-up by both ophthalmology and ENT.

Discussion

If not managed promptly and appropriately, periorbital infections can lead to significant morbidity and even mortality. Complications include endophthalmitis, blindness, cavernous sinus thrombosis, intracranial spread and fatality (1). With such potential devastating consequences, early recognition, prompt assessment and aggressive management of periorbital/orbital sepsis is vital.

How to assess the patient

Peri-orbital infection is a clinical emergency with potential devastating complications. In the majority of cases patients require admission for aggressive management.

Multi-Disciplinary Team (MDT) involvement

The lead specialty varies in the management of orbital sepsis however assessment should be undertaken with a MDT approach. Upile NS et al suggested that children should be admitted under the care of paediatricians with the input of ENT and ophthalmology specialist (2).

This case involved an adult and was predominantly under the care of ophthalmology however it is important to acknowledge that patients can be referred to either speciality. Literature suggests that most patients with peri-orbital cellulitis are primarily managed under the care of ENT teams (3). In this case management also required input from radiology and in particular microbiology due to her allergies.

Eye

A focused examination of the eye is required on admission. Peri-orbital infection can present with eyelid oedema, erythema, proptosis, chemois, ophthalmoplegia, decreased visual acuity and/ or ocular movement (4, 5). With ophthalmological input the eye must be assessed daily with attention to pupillary reflexes, visual acuity, colour vision and ocular movement. Particular attention should be made to red colour vision as this is the first sign of visual disturbance. The patient underwent daily ophthalmology review. Reduced ocular movement with significant proptosis and limited eye examination indicated the need for further investigations.

ENT

Flexible nasoendoscopy (FNE) of the nasal cavity should be included in the ENT examination to look for presence of nasal polyps and evidence of rhinosinusitis such as pus and mucosal inflammation. Paranasal sinus infection is the cause of 60-90% of peri-orbital infections with ethmoid sinusitis most likely to lead to abscess formation (1, 6). FNE was not tolerated in this case but the clinical history strongly indicated a sinogenic cause and was subsequently confirmed on CT imaging and at surgery.

PERI-ORBITAL CELLULITIS AS A COMPLICATION OF SINUSITIS - A CASE BASED DISCUSSION

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Neurological

With the orbit closely related to the skull base there is significant risk of intracranial complications from peri-orbital sepsis. Signs of meningitis must be excluded and a full cranial nerve examination undertaken. Evidence of any neurological deficit requires urgent CT scan with surgical drainage of any abscess.

Management

Peri-orbital infections require urgent, thorough assessment, investigation and treatment. In 1970 Chandler et al produced a classification of orbital complications secondary to sinusitis which is still readily used today (7). This describes the spectrum of presentations encountered and helps indicate the management required. Table 1 summarises this classification.

Stage	Description	Clinical description and definition	Management
I	Pre-septal cellulitis	Eyelid oedema and erythema Normal extraocular movement Normal visual acuity	IV antibiotics and daily review
II	Orbital cellulitis	Diffuse oedema of orbital contents without discrete abscess formation	
III	Subperiosteal abscess	Collection of purulent exudate beneath periosteum of lamina papyracea. Displacement of globe downward/laterally	IV antibiotics and surgical Drainage
IV	Orbital abscess	Purulent collection within orbit Proptosis Chemosis Ophthalmoplegia Decreased vision	
V	Cavernous sinus thrombosis	Bilateral eye findings Prostration Meningism	

Table 1: Chandler's classification in the sequel of periorbital cellulitis with the clinical findings and management (7, 8).

Baseline investigations

All patients should have routine baseline blood test including full blood count, urea and electrolytes and C-reactive protein (CRP). These can act as useful guide, in particular in assessing improvement however one paper reported that 50% of adult patients can have a normal white cell count (9). Therefore it is vital to not rely solely on these results when assessing the severity of the condition. This patient had an abscess at presentation but was systemically well with a minimally raised WCC and a CRP. Suspicion of an abscess was due to the clinical eye findings. In the evidence of systemic sepsis, blood culture and sensitivities should be taken preferably before, but not delaying, the administration of antibiotics (3).

Medical Management

Patients presenting with mild eyelid oedema or pre-septal cellulitis can be managed with oral antibiotics however the majority will require admission for parenteral antibiotics. Nasal decongestants should be used in the acute phase to reduce mucosal oedema and encourage natural drainage of the sinuses (3, 10). This is often commenced by ENT.

Microbiology

Management in these patients needs to be aggressive and started early. Obtaining cultures for sensitivities is valuable but should not delay administration of antibiotics. The blood cultures in this case were negative as it is in 95% of adults (3). Whilst rates of positive blood cultures are low, there is a 31-90% chance of obtaining positive bacteriology results on the surgical specimen (6). The most common pathogens seen in periorbital cellulitis are Streptococcus species, but Haemophilus influenzae, Staphylococcus Aureus and anaerobic bacteria are also frequently isolated (10, 11). Broad spectrum antibiotics such as ceftriaxone or cefotaxime should be used plus coverage for anaerobic bacteria with metronidazole or clindamycin (10). In this case ceftriaxone was used from admission and was subsequently found to be active against the Staphylococcus Aureus isolated on the surgical specimen. Metronidazole was used in addition for anaerobic cover. Close involvement with local microbiology departments is advised to narrow the antibiotic choice and avoid resistance.

Radiology

Table 1 lists the indications for urgent CT scan. CT scan was undertaken in this case due to the reduction in range of ocular movement and limited eye examination secondary to eye lid oedema restricting eyelid retraction. Radiological findings of peri-orbital cellulitis are classified according to the Chandler's Classification⁷. See table 2. A sub-periosteal abscess was demonstrated on CT imaging as seen in figure 1.

Indications for CT scan of sinuses and orbit
Clinical features of central/neurological complication
Difficulty in accurately assessing vision
Ophthalmoplegia, deteriorating visual acuity or colour vision, gross proptosis, bilateral involvement
No improvement/deterioration over 24hours
Swinging pyrexia not resolving within 36hours

Table 2: Indications for urgent CT scan based on guidelines produced by Howe L et al 2004 (3).

PERI-ORBITAL CELLULITIS AS A COMPLICATION OF SINUSITIS - A CASE BASED DISCUSSION

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Surgery

Surgical drainage is indicated when there is confirmed or clinical suspicion of an abscess. Surgery involves external or endoscopic drainage of the peri-orbital abscess with wash out of any sinus involvement. This patient had a sub-periosteal abscess (stage III Chandler's classification) which was drained surgically using an external approach with washout of the sinuses.

Conclusion

Periorbital and orbital infections should be managed from the outset with a MDT approach. Early assessment, by both ENT and Ophthalmology, and subsequent regular review are essential. A CT scan should be performed if the eye can not be examined due to excessive eyelid oedema or if there is proptosis, chemosis, pain on eye movement or reduced vision, or if there is a failure to improve after 24 hours. Aggressive treatment with intravenous antibiotics with or without surgery, will reduce the risk of the potentially serious complications which can occur in this condition.

Best of 5 MCQ's

1. Which specialty should take lead responsibility in the management of patients with peri-orbital infections:

- A. Paediatrics
- B. Ophthalmology
- C. ENT
- D. Microbiology
- E. Acute medical team

2. What is the most likely cause of periorbital cellulitis:

- A. Trauma
- B. Rhinosinusitis
- C. Blephritis
- D. Dacrocystitis
- E. Insect Bites

3. What is the most likely causative organism:

- A. *Staphylococcus Aureus*
- B. *Haemophilus Influenza*
- C. *Streptococcus species*
- D. *Fusobacterium*

4. Which of the following eye signs indicate potential abscess formation:

- A. Reduced range of ocular movement
- B. Loss of visual acuity
- C. Loss of red colour discrimination
- D. Slow/loss of pupil light reflex
- E. Proptosis

Answers

1. A/ B/ C

There is no gold standard guidelines on the specialty that should take lead responsibility. It is usual practise in paediatric patients with the paediatricians to lead the management but with early and regular ENT and ophthalmology input. In adults it is important for both ENT and ophthalmology teams to be aware of the disease and sequel as patients can be admitted under both. Microbiology advice is often sought but not as lead speciality.

2. B

In 60-90% of cases rhinosinusitis is the primary underlying cause and is more so in post septal disease (Chandler's II - V). In pre-septal cellulitis (Chandler's stage I) the other listed aetiologies are also commonly seen.

PERI-ORBITAL CELLULITIS AS A COMPLICATION OF SINUSITIS - A CASE BASED DISCUSSION

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3. C

With the introduction of the Haemophilus Influenza (HiB) vaccination there has been a reduction of cases of orbital cellulitis secondary to this pathogen. The most commonly reported causative agent are Streptococcus species however chosen antibiotics should have Staphylococcus Aureus and anaerobic cover as these are also commonly isolated bacteria.

4. A/B/C/D/E

Loss of red colour vision is an early indicator of impending visual complications and abscess and CT imaging should be arranged urgently to assess the extent of the abscess.

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CASE BASED DISCUSSION: RECURRENT EPISTAXIS AS A MANIFESTATION OF HEREDITARY HAEMORRHAGIC TELANGIECTASIA

A Sanchez, K Fenton, C McLarnon

Case Based Discussion: Recurrent Epistaxis As A Manifestation Of Hereditary Haemorrhagic Telangiectasia Patient Management

Abstract

Recurrent epistaxis is a common presentation to otolaryngology. When assessing the patient with recurrent epistaxis, less common causes should be considered, including hereditary haemorrhagic telangiectasia. This is an important diagnosis as the disease can have severe complications and a multidisciplinary approach is required. We discuss a case of a 40 year old man who presented with recurrent epistaxis since the age of 16, worsening in recent months. Due to a family history of epistaxis and cutaneous signs, hereditary haemorrhagic telangiectasia was suspected.

Case History

A 40 year male retail worker was referred to the otolaryngology clinic with recurrent bilateral anterior epistaxis. He described recurrent nose bleeds since the age of 16. In the last four months, nosebleeds were waking him from sleep several times every night. They tended to last around ten minutes before settling with conservative measures. Previously, he had had one or two nosebleeds per day, each lasting around one minute.

He had not identified any triggers for the bleeding and had undergone nasal cautery multiple times at his local hospital. Both his father and paternal grandfather suffered from recurrent epistaxis. The patient was HIV positive and had used cocaine in the past. He experienced intermittent headaches and had an iron deficiency anaemia likely secondary to recurrent epistaxis. He also reported rectal bleeding which had not been investigated at the time of presentation.



On examination, telangiectasias were visible on the patient's bottom lip, the roof of his hard palate, and throughout his nasal cavity (Figures 1, 2 and 3). No septal perforation was noted. His consultant suspected Hereditary Haemorrhagic Telangiectasia (HHT) as the cause of his recurrent epistaxis and he was subsequently listed for Potassium Titanyl Phosphate (KTP) laser cautery to the septal telangiectasias.



Figure 2: Telangiectasia on hard palate.



Figure 1: Telangiectasia on bottom lip.

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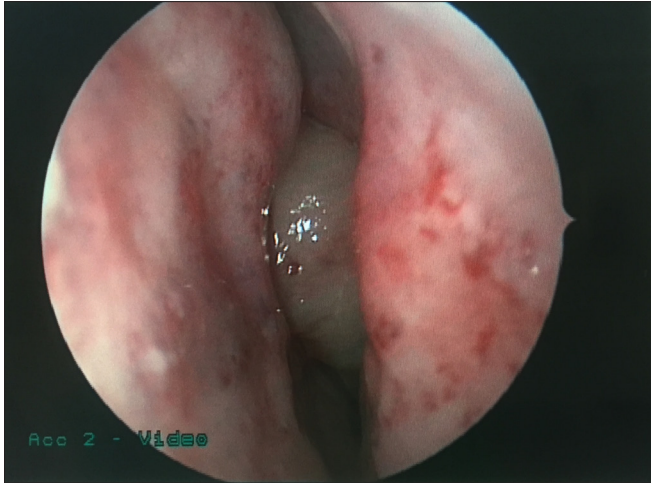


Figure 3: Telangiectasia nasal mucosa.

He was referred to the pulmonary vascular clinic where telangiectasias were also noted on the patient's fingertips (Figure 4). No pulmonary arteriovenous malformations (AVMs) were detected on Chest X-Ray and oxygen saturations were 98%, both lying and standing.



Figure 4: Telangiectasia on finger.

Given the strong clinical suspicion of HHT, he has been offered genetic testing for definitive diagnosis, which is still awaited at the time of writing. Should he experience ongoing headaches, he may require imaging to exclude cerebral AVMs. He will be followed up in ENT outpatient clinic six monthly.

Discussion

Background

HHT, also known as Osler-Rendu-Weber disease, is a multi-system vascular dysplasia. It is caused predominantly by mutations in HHT1 (ENG) or HHT2 (ACVRL1) genes and is typically autosomal dominantly inherited but may also arise as a point mutation [1]. Other genes such as SMAD4 have also been implicated [2].

Its prevalence is approximately 1 in 5,000 but may be underdiagnosed [2]. The hallmark of the disease is mucocutaneous telangiectasias and AVMs in the gastrointestinal tract, liver, lungs and central nervous system. These AVMs can give rise to intracerebral haemorrhage and other potentially fatal bleeds. Telangiectasias are capillary dilatations near the surface of the skin or mucous membranes which appear as small red dots or spider-like red lines.

Recurrent epistaxis is the most common presentation of HHT and often first presents in early adolescence [2]. The epistaxis can range from mild to severe and often worsens with age. The nosebleeds result from rupture of the fragile telangiectasias in the nasal mucosa. Cutaneous telangiectasias typically occur several years after the onset of recurrent epistaxis.

Epistaxis - Initial Assessment

All patients should be assessed using an ABC approach [3]. A full set of observations should be undertaken to establish if the patient is haemodynamically compromised. Significant epistaxis requires wide bore intravenous access and fluid resuscitation should be administered if there is evidence of shock. Full blood count and coagulation screen should be sent to identify anaemia and coagulopathy respectively. The patient should be group and saved as they may require transfusion.

Conservative management of epistaxis comprises leaning the patient forward, pinching the soft part of the nose and applying ice to the bridge of the nose and nape of the neck to induce vasoconstriction. Mild epistaxis may be successfully treated with silver nitrate cautery. If the bleeding persists despite conservative measures, proceed to nasal packing.

Once the patient is stabilised, a history should be taken. This should ascertain the onset, duration, side and severity of the bleed. Establish if it is anterior or posterior and identify potential precipitants including, but not limited to trauma, hypertension and anticoagulation.

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Differential

Recurrent epistaxis can also present in coagulation disorders, inflammatory disease and nasal neoplasm [3]. Patients with telangiectasia should be questioned for other features of CREST (Calcinosis, Raynaud's, Esophageal dysmotility, Sclerodactyly and Telangiectasia) syndrome as well as HHT.

Diagnosis and Investigation

The Curaçao Criteria can be used to aid diagnosis of HHT [4]. The four part criteria consist of 1) recurrent spontaneous epistaxis, 2) mucocutaneous telangiectasias, 3) internal arteriovenous malformations and 4) having a first degree relative with HHT. A definite diagnosis is given if three or more criteria are present; the presence of two criteria suggests a 'possible or suspected diagnosis' of HHT; and HHT is unlikely if there are less than two present, although this should not exclude a diagnosis.

This patient demonstrated two of the above criteria and a family history strongly suggestive of HHT, if not a definitive diagnosis. It is suspected that the Curaçao Criteria may lead to underdiagnosis of HHT in younger children who are yet to present with epistaxis or mucocutaneous telangiectasia and those patients who have developed HHT via a point mutation rather than inherited it [2]. As such, genetic testing should be offered to these individuals. If genetic testing cannot be offered, these individuals should be investigated for presence of AVMs [2]. It is recommended genetic testing be offered to family members of patients with confirmed HHT, even if asymptomatic [2].

HHT Management

The management of HHT includes symptomatic treatment, identification of AVMs and mitigating the complications of AVMs [1].

Given the multisystem nature of HHT, a multidisciplinary team approach is indicated. ENT input is required for recurrent epistaxis whilst Respiratory, Hepatology, GI and Neurology services may be needed to screen for AVMs. Should an AVM be identified, it must be regularly monitored as they are prone to growth. Haematology input may be required for the treatment of associated anaemias.

Over three quarters of all HHT patients will eventually develop recurrent epistaxis [2]. Humidification and nasal lubricants may slightly reduce their frequency [2]. If nasal packing is indicated in HHT epistaxis, it should be lubricated to minimize damage to nasal telangiectasias [2]. Those with severe symptoms should be referred to an ENT surgeon with clinical experience of HHT. Treatment options include cautery, dermoplasty and closure of the nasal cavity (Young's procedure) [2,5].

KTP laser cautery combined with submucosal injection of the monoclonal antibody Bevacizumab showed a statistically significant decrease in epistaxis frequency and number of blood transfusions required compared to KTP laser cautery alone [6]. Tranexamic acid and estrogens have improved patients' perceptions of their symptoms, if not clinical outcomes such as haemoglobin levels [2,7,8].

Those cases of HHT caused by mutations in the SMAD4 gene require rigorous gastrointestinal screening as this variant of HHT includes features of juvenile polyposis [9]. This involves colonoscopy starting in middle teenage years, to be repeated every one to two years [2].

Multiple Choice Questions

1. How is HHT diagnosed?

- a) Light's Criteria
- b) Curaçao Criteria
- c) Well's Criteria
- d) Ranson's Criteria
- e) Hereditary Haemorrhagic Telangiectasia Criteria

2. How is HHT inherited?

- a) Autosomal recessively
- b) Autosomal dominantly
- c) Point mutation
- d) A and C
- e) B and C

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3. Which of these is the most common symptom of HHT?

- a) Recurrent epistaxis
- b) Haemoptysis
- c) Haematemesis
- d) Haematochezia
- e) Intracerebral haemorrhage

4. A patient with a known diagnosis of HHT presents with an anterior nosebleed that has not settled with conservative measures. What is the most appropriate next step?

- a) Foley's catheter and BIPP
- b) Nasal cotton wool
- c) Lubricated nasal pack
- d) Silver nitrate cautery
- e) Young's Procedure

5. A mutation in which gene is associated with a syndrome encompassing HHT and juvenile polyposis?

- a) ENG
- b) ACVRL1
- c) SMAD4
- d) BMPRII
- e) None of the above

Multiple Choice Answers & Teaching Notes

1 - b

2 - e

3 - a

4 - c

5 - c

Answer explanations

1. The Curacao Criteria consists of criteria that can be used to aid in a diagnosis of HHT. They are recurrent spontaneous epistaxis, mucocutaneous telangiectasias, internal AVMs and a first degree relative with the disease. Three or four criteria give a definitive diagnosis; HHT is unlikely if less than two are met.

2. HHT is inherited autosomal dominantly or via point mutations, most commonly via mutations in HHT1 or HHT2 genes.

3. HHT can cause all of the symptoms mentioned, but most commonly presents with recurrent epistaxis. Patients with suspected HHT presenting with epistaxis should be questioned about bleeding from other sources, and should be screened for AVMs should they have any suggestive symptoms, for example headaches or gastrointestinal bleeding.

4. Patients with HHT presenting with epistaxis should be managed acutely as per any epistaxis patient. This should include an ABC assessment to ensure haemodynamic stability. If conservative measures fail, proceed to nasal packing, ensuring that in HHT the packs are lubricated to prevent damage to the fragile telangiectasias in the nose.

5. Mutations in the SMAD4 gene can lead to a version of HHT which includes juvenile polyposis. It is important that these patients are identified in order to conduct gastrointestinal screening from the teenage years to assess for polyps, as these can be a precursor to gastrointestinal cancers.

CASE BASED DISCUSSION: RECURRENT EPISTAXIS AS A MANIFESTATION OF HEREDITARY HAEMORRHAGIC TELANGIECTASIA

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CASE BASED DISCUSSION; SIMPLE SORE THROAT? THE IMPORTANCE OF THOROUGH CLINICAL ASSESSMENT IN ACUTE TONSILLITIS

H Saeed, G Shalom, T Ellis



Case Based Discussion; Simple Sore Throat? The Importance Of Thorough Clinical Assessment In Acute Tonsillitis Patient Management

He had no previous episodes of tonsillitis or peritonsillar abscess. Examination findings were bilateral painful level two lymph node swellings, mild trismus and bilateral exudative tonsils. The Uvula was central and there was a patch of presumed oral candidiasis on the hard palate. There was no dysphonia and neck movements were normal. Salient blood tests results are highlighted in table 1.

The suspected diagnosis of acute tonsillitis with oral candidiasis was made and the patient was admitted for intravenous (IV) fluid therapy, antibiotics (benzylpenicillin), oral fluconazole and analgesia. The patient was given 24 hours of IV antibiotics. His oral intake and pain improved allowing discharge with oral antibiotics (penicillin V), antifungal (nystatin) and analgesia.

Unfortunately, the patient re-presented 14 days later. He was once again assessed by the on-call ENT doctor. On this occasion GM was showing signs of a systemic inflammatory response; temperature of 38.6, pulse of 130 beats per minute, blood pressure of 107/56, respiratory rate of 17 and sats 100 percent on room air. His symptoms of sore throat had worsened after completing his course of oral antibiotics. GM complained of odynophagia, fevers, drenching night sweats and decreased oral intake. Once again bilateral exudative tonsillitis, oral candidiasis and cervical lymphadenopathy were found on examination.

Initial ECG revealed a sinus tachycardia and a venous blood gas was grossly normal. The working diagnosis was recurrent bacterial tonsillitis with secondary sepsis. A "sepsis six" management plan was instigated by both the A+E and ENT team. The patient was transferred to the ENT ward for further treatment and investigations.

Abstract

We describe a case of a 22-year-old male presenting to the ear, nose and throat (ENT) team with symptoms and signs of tonsillitis with sepsis. Routine management is discussed followed by further investigations to diagnose a serious underlying condition. Good aspects of clinical care by junior members of the ENT team are highlighted as well as areas where patient care could have been improved. Important questions and answers regarding the management of tonsil pathology are given to act as a learning tool for foundation year doctors.

Case History

A 22-year-old male (GM) of Polish descent presented to our accident and emergency with symptoms and signs of sore throat. He was promptly referred to the ENT on call for further assessment for presumed acute tonsillitis. GM had a past medical history epilepsy which was controlled by bi-daily levetiracetam. He had no other known medical history nor allergies to medication.

At initial assessment GM showed signs of systemic upset; a tachycardia of 113 BPM, a temperature of 36.6, a blood pressure of 108/59, a respiratory rate of 16 and oxygen saturations of 99% on room air. He gave a four-week history of progressive sore throat associated with intermittent night sweats. His GP had prescribed oral antibiotics (Penicillin V) which had not curtailed the symptoms. The pain had led to a minimal food and fluid intake.

CASE BASED DISCUSSION; SIMPLE SORE THROAT? THE IMPORTANCE OF THOROUGH CLINICAL ASSESSMENT IN ACUTE TONSILLITIS

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Haematological and biochemical markers had worsened since GM's initial presentation (table 1). Once again infectious mononucleosis screen was negative. During both presentations a neutropenia with lymphocytosis was present, and atypical mononuclear cells with reactive lymphocytes were seen on the blood film. Additionally, in between presentations the following results became available; total serum IgG level was elevated at 25.94 g/L (6.00-16.00), and a paraprotein blood level revealed a polyclonal increase in gamma globulins.

Investigation	Units & Range	1 st presentation	2 nd Presentation
WCC	10 ⁹ /L (4.0-11.0)	7.2	10.2
RBC	10 ¹² /L (4.6-6.0)	3.5	2.10
Hb	g/L (130-180)	120	74
Neutrophils	10 ⁹ /L (1.8-7.5)	<1	<1
Lymphocytes	10 ⁹ /L (1.5-4.0)	6.9	9.8
Haematocrit	(0.4-0.54)	0.34	0.204
MCV	fL (80-96)	97.2	96.9
MCH	Pg (27-32)	34.2	35
Platelets	10 ⁹ /L (130-400)	74	43
Infectious Mononucleosis screen (IM)		Negative	Negative
CRP	Mg/L (0-8)	77	187

Table 1

Blood tests results at initial and second presentation. Note the decline in haemoglobin and platelets with persisting lymphocytosis with neutropenia.

The abnormal blood findings prompted the ENT team to gain an urgent haematological review. A repeat blood film showed abnormal lymphoid cells and further investigations were instigated to exclude an acute leukaemia. The patient was transfused packed red cells, isolated in a side room with barrier nursing and switched to IV Tazocin and gentamicin therapy to treat suspected neutropenic sepsis. Urgent transfer of GM was arranged to the tertiary haematological unit for further management.

Provisional cell markers (figure 1 & 2) were suggestive of acute leukaemia with double lineage (both lymphoblastic and myeloblastic). This diagnosis was supported by bone marrow aspiration. The diagnosis was sensitively discussed with the patient by members of both the ENT and haematology team. Further management at the haematology unit involved continued barrier nursing, on going antimicrobial cover and the instigation of chemotherapy. Cytogenetics and trephine biopsy are awaited to confirm exact cell lineage.

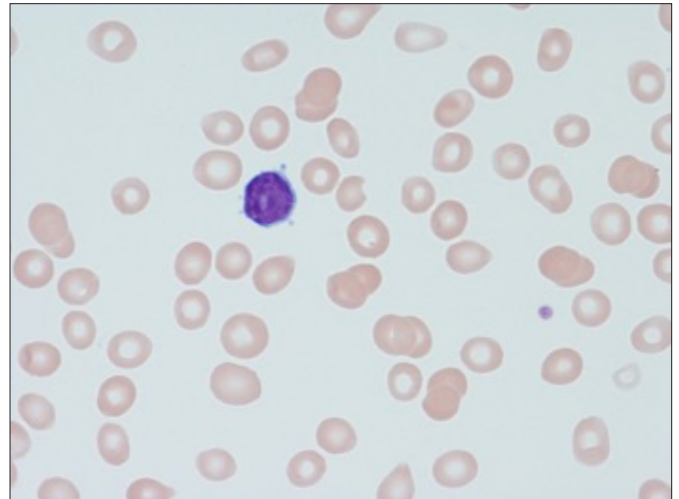


Figure 1: GM's blood film showing an abnormal lymphoblast (arrowed) which could indicate an acute lymphocytic anaemia.



Figure 2: GM's blood film showing an immature myeloblast (arrowed). This suggests a potential for a mixed acute lymphoblastic and myeloblastic leukaemia.

CASE BASED DISCUSSION; SIMPLE SORE THROAT? THE IMPORTANCE OF THOROUGH CLINICAL ASSESSMENT IN ACUTE TONSILLITIS

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Discussion

The above case highlights several examples of good clinical practice and reminds foundation years doctors of the potential pitfalls when presented with “common” pathologies. At 1st presentation, clinical assessment was thorough and accurate for several reasons; current clinical guidelines regarding when patients should be admitted with tonsillitis were adhered to, the potential diagnosis of peritonsillar abscess was excluded early on by detailed examination and important investigations such as an infectious mononucleosis (IM) screen were obtained early.

The recent SIGN guidelines state that patients with progressive difficulty with swallowing, increasing pain, or severe systemic symptoms should be admitted. If a Peritonsillar abscess was missed, subsequent treatment would be ineffective until the abscess was recognised and drained. Knowing IM is absent early on allows for the option to give amoxicillin containing antibiotics safely if necessary. Additionally, if IM had been missed there may have been implications involving patient safety; the viral illness typically takes 6-8 weeks to resolve and may require monitoring of LFT's in the community as hepato-splenomegaly is a recognised complication of glandular fever.

At second presentation, good clinical care was apparent in that sepsis was recognised early and the subsequent evidence based “sepsis six” care bundle was instigated without delay. Furthermore, severely deranged haematological investigations were repeated, scrutinised further and compared with previous. Junior members of the team promptly discussed their concerns regarding the blood dyscrasia and recurrent symptoms with ENT and Haematology consultant colleagues.

As a result, the patient received appropriate senior multi-disciplinary management and the juniors were able to learn through effective discussion and guidance from the Consultants. Additional examples of efficient patient management included liaising effectively with microbiologists and the bed managers to ensure a side-room was available to treat neutropenic sepsis, effectively communicating with the tertiary haematology unit and working with the haematology nurse to schedule a meeting with the patient so that bad news could be broken in an appropriate manner. Overall therefore, this prioritisation and attention to detail by the ENT team members provided an efficient flow to patient care.

On reflection, there are areas where initial assessment could have been improved. It is difficult to conclude that the differential of acute leukaemia should have been explored further at initial presentation; the patient's age, initial findings of a lymphocytosis, abnormal lymphoid cells and thrombocytopenia on the blood film could have all pointed to a diagnosis of glandular fever (the result of the IM screen would not have been known at this stage).

Additionally, the mild anaemia on first presentation could have been longstanding or associated with a systemic inflammatory response. However, neutropenia coupled with oral candidiasis in a usually fit young man and a history specific of night sweats should have prompted the ENT team to recognise a potentially immunocompromised individual and therefore arrange pertinent investigations. At this stage perhaps the unusual clinical presentation should have been discussed sooner with ENT seniors. Further emphasis could have been put on chasing the outstanding serum IgG and paraprotein levels. This may have allowed for early liaison with the GP and further testing to exclude an acute leukaemia prior to second presentation.

Test Yourself;

Question 1

An 18 year old student presents to the A&E department with a 1 week history of sore throat and temperature. She is no longer eating or drinking. On examination she has bilaterally enlarged tonsils with whitish exudate. There is also marked cervical lymphadenopathy. She also complains of lethargy, malaise and pain in the right side of her abdomen. She has been to her GP last week who prescribed her with a 10 days course of antibiotics.

What is the most likely explanation of her symptoms?

- A. Bacterial tonsillitis
- B. Epstein Barr
- C. General viral upper respiratory tract infection
- D. None of the above

CASE BASED DISCUSSION; SIMPLE SORE THROAT? THE IMPORTANCE OF THOROUGH CLINICAL ASSESSMENT IN ACUTE TONSILLITIS

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Question 2

From the following list select one or more of the organisms responsible for acute tonsillitis?

- A. Group B streptococcus
- B. Haemophilus influenza
- C. Pneumococci
- D. Viral infection
- E. All of the above

Question 3

A 27 year old teacher is referred by his GP for a sore throat that has not resolved after 7 day course of amoxicillin. On examination his tonsils are bilaterally inflamed with some signs of exudate. He has significantly raised cervical lymph nodes, and a widespread itchy maculopapular rash. The patient is febrile and has not been eating or drinking.

What is the most likely diagnosis?

- A. Infectious mononucleosis
- B. Peritonsillar abscess
- C. Bacterial tonsillitis
- D. Parapharyngeal abscess
- E. Viral upper respiratory tract infection

Question 4

A 16 year old girl is referred to A&E after complaining of coughing up small amounts of blood. She is 7 days post adenotonsillectomy. There is whitish slough on the back of her throat but no active bleeding. The patient is afebrile, has a blood pressure of 125/82 with a heart rate of 78. She does not feel unwell and is only complaining of mild post-operative pain at the back of her throat.

What should be the management plan?

- A. Place patient on emergency theatre list to diathermy the area to stop further bleeding.
- B. Admit the patient to ambulatory care and monitor for a few hours before sending home.
- C. Reassure patient and send her home with safeguarding advice.
- D. Admit the patient, get intravenous access and start IV antibiotics and IV fluids.

Question 5

A mother attends your clinic with her 5 year old daughter who has been complaining of a sore throat. You diagnose bacterial tonsillitis. Mum reports that in the last year her daughter has had four bouts of tonsillitis and asks if her daughter can undergo a tonsillectomy to cure the problem once and for all.

What is the best management plan?

- A. Place patient on emergency tonsillectomy list.
- B. Treat with antibiotics, reassure and send patient home.
- C. Place patient on routine tonsillectomy list.
- D. Explain to the mother that daughter is too young for a tonsillectomy and you can review her again once she reaches six years of age.

Answers

Question 1

This patient most likely has a diagnosis of glandular fever caused by the Epstein Barr virus. Glandular fever can present with enlarged tonsils with exudate, fever and feeling generally unwell. The deciding factor in this case is the patient has been prescribed antibiotics a week ago with no clinical improvement. This is because glandular fever is a viral infection, and will not respond to antibiotics. Additionally, the patient is complaining of right upper quadrant pain which is likely to liver capsular stretch, as glandular fever is associated with hepatosplenomegaly.

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Question 2

The answer to this one is 'E'. All of the above can cause tonsillitis. In bacterial tonsillitis the most common organism is group B strep, hence why a penicillin based antibiotic is usually prescribed. This demonstrates the point that health professionals should not automatically prescribe antibiotics for sore inflamed throats and irritated tonsils. The whole clinical picture should be looked at to decide if it's a bacterial or viral in origin. The CENTOR criteria can help doctors differentiate between bacterial and viral tonsillitis.

Question 3

This patient has infection mononucleosis also known as glandular fever. The maculopapular rash is a common reaction to amoxicillin when given in glandular fever. This does not mean the patient is allergic to penicillins, it's an interaction in the body between the virus and the antibiotics. It is for this reason all suspected tonsillitis patients are started on phenoxymethylpenicillin, to prevent a reaction in the event they are affected by glandular fever.

Question 4

The answer to this scenario is 'D'. The patient is likely suffering a secondary haemorrhage complication due to the recent tonsillectomy. There are two types of post-tonsillectomy haemorrhages. A reactive (primary) haemorrhage occurs in the first few hours following the procedure and may require return to theatre for arrest of post-tonsillectomy haemorrhage. A secondary haemorrhage occurs within 14 days after the procedure and is usually due to infection. The protocol in this sort of presentation is to admit and give antibiotics. Even if a small amount of blood is seen post tonsillectomy, patients should be admitted for observation for approximately 24 hours; a small bleed may be a herald bleed. Brisk haemorrhage may shortly follow!

Question 5

The second option 'B' is the correct choice. According to NICE the indications are 1. Seven episodes of bacterial tonsillitis in the past year. 2. Five episodes of bacterial tonsillitis in last two years or 3. Three episodes of bacterial tonsillitis per year for last three years. Additionally, patients who have more than 1 episode of peritonsillar abscess should have tonsillectomy. Other absolute indications are children with confirmed obstructive sleep apnoea and suspected malignancy. There are other rarer indications for tonsillectomy not discussed here.

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CASE BASED DISCUSSION: SUDDEN SENSORINEURAL HEARING LOSS

H Zhang, A Bhattacharyya

Case based discussion: Sudden Sensorineural Hearing Loss Patient Management

Abstract

This case based discussion centres around a 61-year-old gentleman with sudden unilateral deafness. Here, we outline the clinical assessment of hearing loss including history and examination, a differential diagnosis, the appropriate investigations and treatment of this condition.

Case history

Mr. HB, a 61-year-old chef presents to the urgent care centre with a 48 hour history of rapidly progressing, left side hearing loss.

What are the key points in your history?

What would you like to do next?

1. History of presenting complaint

- When did the symptoms start?
- Any related otological symptoms?
- Otagia
- Otorrhoea
- Tinnitus
- Vertigo
- Facial weakness
- Is this the first episode?
- Any preceding head trauma?
- Any associated nose/throat symptoms?
- Any preceding coryzal symptoms?
- Any chance of a foreign body (e.g. hearing aid battery)
- Any systemic symptoms such as weight loss/loss of appetite?



2. Past medical history

- Co-morbidities, specifically
 - Diabetes
 - Hypothyroidism
 - Clotting disorders
 - Arteriosclerosis
 - Neurological disorders, e.g. Multiple sclerosis
- Previous traumatic head injury
- Previous ear surgery

3. Drug history

Pay special attention to any ototoxic medications. The main ones include:

- Aminoglycosides (Gentamicin)
- Macrolides (Erythromycin)
- Loop diuretics
- Salicylates
- Quinine
- Antineoplastics (cisplatin)

4. Social history

Occupation and noise exposure is important. A comprehensive sexual history should also be taken to determine risk factor to sexually transmitted infections.

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5. Family history

- Anyone in the family with autoimmune disorders?
- Neurofibromatosis/ vestibular schwannomas?
- Coagulation disorders?
- Otosclerosis?

Mr HB denies related otological symptoms. He mentions that a week previously he had coryzal symptoms, which he put down to looking after young grandchildren. He denies any preceding head trauma. He is otherwise well, takes amlodipine 5mg od for hypertension and no other medication. He does not have any family history of illnesses. He has been married for 30 years and denies sexual promiscuity.

How would you proceed with the consultation?

A full ENT examination, paying close attention to the ears:

1. Start with the better ear. This is so you have a comparison for what is considered normal for the patient. Always ask if the patient has pain before proceeding. Begin with the outer ear, examining the pinna, post-auricular region, external auditory meatus. Check for tragal tenderness.
2. Examination of the external auditory canal (EAC) and tympanic membrane (TM) is achieved with an otoscope. Hold this like a pencil and examine each side with your ipsilateral hand (ie. Left ear should be examined with the otoscope in the left hand, and vice versa). Pulling the pinna superiorly and posteriorly will straighten the EAC and aid in visualisation of the TM.
3. Repeat this on the contralateral ear.
4. Assess for facial weakness.
5. The rest of the ENT examination in primary care includes examining the oropharynx and anterior rhinoscopy.

You examine Mr HB's ears and the external ear, EAC, and TM are normal on both sides. The rest of the examination is also unremarkable. What is your next step?

Tuning fork tests

1. Rinne's test.

2. Weber's test.

An understanding of tuning fork tests and their interpretation is vital to an ENT examination. Table 1 summarises the results of Rinne's and Weber's tests.

	Weber's	Rinne's Left	Rinne's Right
Normal	Central	AC > BC	AC > BC
Left CHL	Left	BC > AC	AC > BC
Right CHL	Right	AC > BC	BC > AC
Left SNHL	Right	AC > BC	AC > BC
Right SNHL	Left	AC > BC	AC > BC

Table 1: Tuning fork test interpretation.

Please note the false negative Rinne's test. This occurs in profound SNHL, and the patient would report BC > AC in the affected ear. This is because of the transmission of sounds to the contralateral cochlear through the skull base.

Performing tuning fork tests on Mr HB, you find that Weber's test lateralises to the right. Rinne's test is positive bilaterally. What does this mean?

This shows that Mr HB has sensorineural hearing loss on the left.

Which investigations would you consider referring Mr HB for?

1. Audiological testing
2. Blood tests
3. Imaging

How would you proceed with initial management?

In the acute setting, a patient diagnosed with sudden sensorineural hearing loss is treated with corticosteroids. This needs to be commenced with caution, as its benefits need to be balanced carefully with its risks on the patient's comorbidities (e.g. diabetes). Prompt discussion with ENT specialists and a plan for follow up in the ENT clinic is essential.

Discussion

Sudden sensorineural hearing loss (SSNHL) is commonly defined as a sensorineural hearing loss of greater than 30dB over 3 contiguous pure tone frequencies, occurring within a 3-day period (1). Fortunately, it usually presents unilaterally and has a good prognosis in terms of hearing recovery.

The theory behind its pathophysiology has been considered to be from one or more of labyrinthine viral infection, vascular compromise, immune mediated inner ear disease, and intracochlear membrane ruptures. The condition has an equal sex distribution, with an incidence of 5-20 cases per 100,000 and a peak incidence in the sixth decade of life (5).

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Infection	Inflammation	Tumour	Vascular	Endocrine	Toxins
Bacterial – Meningitis, syphilis	Sarcoidosis	Vestibular Schwannoma	Emboli	Hypothyroidism	Antibiotics (aminoglycosides)
Viral – Mumps, CMV, VZV	Granulomatous polyangiitis	Temporal bone metastases	Hypercoagulable states	Diabetes mellitus	Cisplatin
	Cogan syndrome	Carcinomatous meningitis	Post-radiation therapy		Loop diuretics

Table 2: Aetiological Factors in Sudden Sensory Neural Hearing Loss.

SSNHL is an otological emergency. It is important in the acute setting to distinguish between sensorineural and conductive hearing loss, thereby prompting appropriate treatment. The history is vital, to determine the course of symptoms, risk factors for hearing loss, or a definable underlying disease. The term sudden hearing loss encompasses SSNHL.

Although the majority of cases are deemed idiopathic, there are identified underlying aetiologies (Table 2) (6). Clinical examination of the external auditory canal and the tympanic membrane may be normal, which makes the diagnosis more likely. Any reversible causes should be treated for, and it is important to examine the patient in general to determine any differential diagnoses. Pure tone audiometry is mandatory in all patients, and this requires referral to audiology or ENT centres.

Investigations include blood tests; full blood count to look for signs of infection; autoimmune screen including ANA, ESR, pANCA/cANCA, ACE; INR and APTT; thyroid function tests; fasting glucose; screening for syphilis.

Magnetic resonance imaging (MRI) of the internal auditory meati (IAMs) with gadolinium enhancement is the imaging modality of choice to look for cerebellopontine angle masses, most commonly vestibular schwannoma.

There is no preferred treatment for SSNHL. It is important to counsel the patient on potential permanent hearing loss. Adjunctive management using hearing aids may be needed, and this topic must be approached sensitively.

Treatment should be tailored to the findings on history, examination and investigations. Any reversible causes, such as ototoxic medication, infection, poorly controlled diabetes should be promptly managed. If no definable aetiology is found, the treatment should be balanced with the patient's comorbidities in terms of their risks.

The most common medical therapy is corticosteroids. The recommended regimen is prednisolone 1mg/kg per day, or 60mg maximum in adults, for 10 days followed by gradual tapering of dose (2, 7, 10). There is evidence to suggest that intratympanic injection of corticosteroids, in combination with oral therapy, can be more effective (3, 7). Either way, careful discussion with the patient and urgent referral to an ENT department is necessary before consideration of this.

Other treatments that have shown to be effective include vasodilators such as Carbogen (5% carbon dioxide) inhalation (4), and hyperbaric oxygen therapy (8). Both of these, in theory, increase blood flow to the cochlea. Evidence is also given in randomised controlled trials to suggest that antiviral therapy, e.g. acyclovir and valacyclovir, are effective in conjunction with steroids (9).

MCQ (Single Best Answers)

1. What is a commonly used definition of sudden sensorineural deafness?

- > 30dB hearing loss across 2 frequencies occurring within 24 hours
- At least 35dB hearing loss across 3 frequencies occurring within 48 hours
- > 30dB hearing loss across 3 frequencies occurring within 48 hours
- >30dB hearing loss across 3 frequencies occurring within 72 hours
- Any hearing loss occurring within 72 hours

2. SSNHL usually presents in:

- Children
- Young adults
- Middle aged
- The sixth decade of life
- There is no distribution in age

3. Tuning fork tests for a patient with right sided SSNHL would show:

- Weber's lateralising to the right, Rinne's positive bilaterally
- Weber's central, Rinne's negative on the right, positive on the left
- Weber's central, Rinne's positive on the right, negative on the left
- Weber's lateralising to the left, Rinne's positive bilaterally
- Weber's lateralising to the left, Rinne's negative bilaterally

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4. Initial treatment for SSNHL should be:

- Vasodilators
- Corticosteroids, paying careful attention to comorbidities
- Hyperbaric oxygen therapy
- Intratympanic injections of corticosteroids
- Surgery

5. Appropriate referral to radiology for imaging can be considered in cases of SSNHL to look for vestibular schwannoma. The most appropriate imaging modality is:

- X ray temporal bone
- CT Head
- MRI of internal auditory meati with gadolinium enhancement
- Non-contrast MRI head
- Angiography

Answers

1. Answer D

This is a commonly used definition, although no strict criteria exists.

2. Answer D

Most commonly, SSNHL presents in the sixth decade of life. There is an equal distribution between young adults and the elderly.

3. Answer D

A right-sided SSNHL would mean that Weber's lateralises the contralateral side, the left. Rinne's would show that AC>BC, even in SSNHL, and therefore be POSITIVE bilaterally.

4. Answer B

Evidence suggests that prompt treatment with high dose corticosteroids is the preferred method.

5. Answer C

MRI IAMS with gadolinium enhancement is the first line imaging modality for cerebellopontine angle lesions.

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