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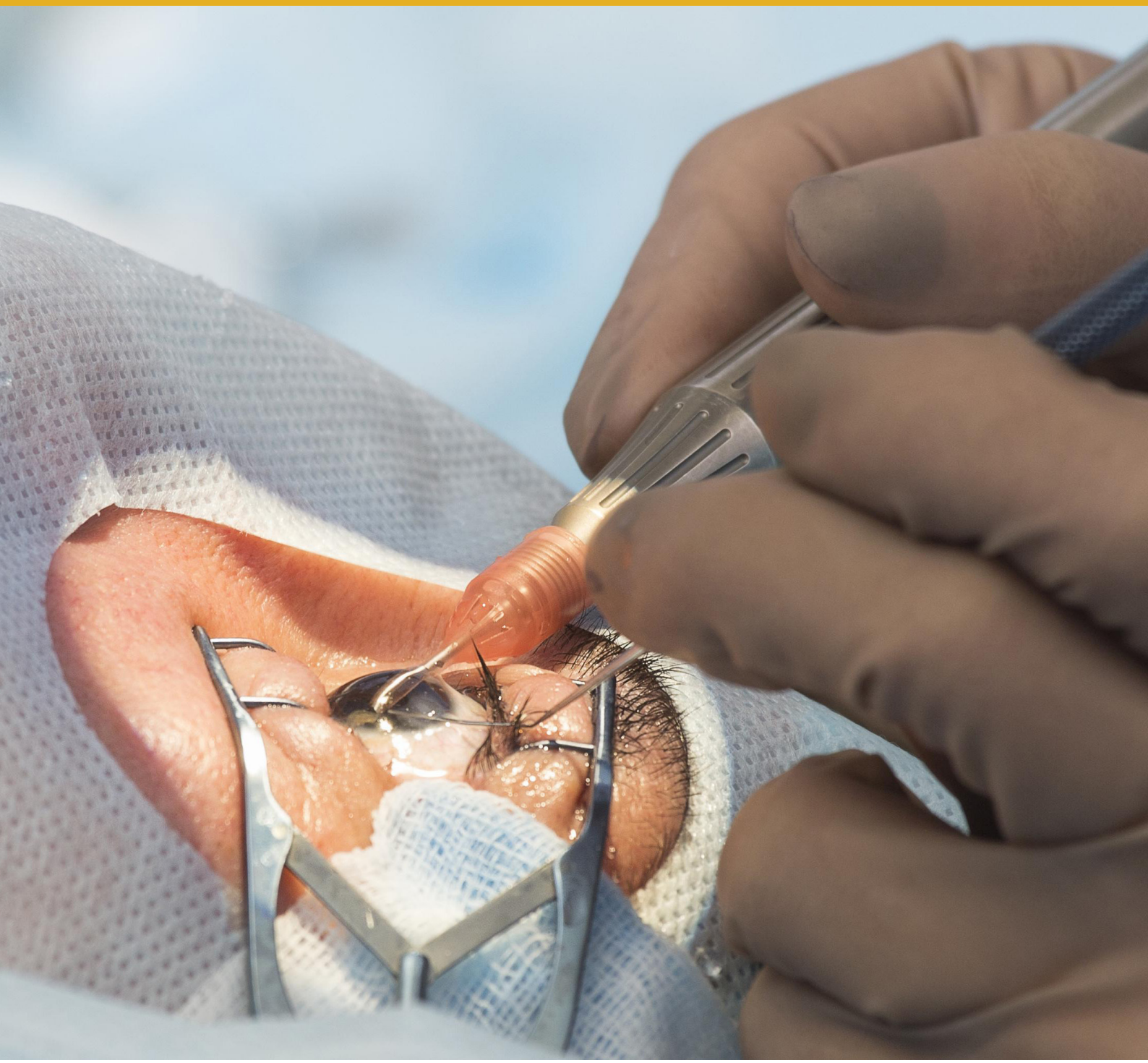

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Volume 12

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Volume 12

Foundation years journal

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AN UNUSUAL CAUSE OF UNILATERAL PAINFUL RED EYE

A Youssef, R Brennan

Abstract

A 57 year old female presented to eye casualty with sudden, severe ocular pain and marked injection of the left eye over a 5-day period, consistent with scleritis. Her symptoms were associated with nausea and photophobia. Her past medical history included seronegative arthritis, reduced appetite and 10kg weight loss over the past 6 months.

Upon examination, her vision acuity in the affected eye was reduced so she could neither see letters on Snellen chart, nor count fingers, but only perceive hand movements, 360 degrees of scleral and conjunctival injection was found, which was most significant overlying a peripheral haemorrhagic choroidal effusion seen on dilated fundoscopy. B-scan showed an inferior-nasal echogenic mass which was confirmed to be arising from the choroid on CT and MRI scans of the Orbit and associated with exudative retinal detachment.

The scleritis significantly improved with high dose oral Prednisolone. She was referred to the regional Ocular Oncology services in view of the choroidal mass. However, she re-presented 2 weeks later to Accident and Emergency with severe pain in the affected eye and a raised intraocular pressure above 90mmhg. An emergency eye enucleation was carried out and histopathology examination of the eye confirmed a malignant choroidal melanoma.

This case discusses an unusual aetiology of unilateral scleritis, and it highlights the importance of careful posterior segment examination in patients presenting with an ocular inflammation.

Case history

A 57 year old female was referred to eye casualty by her GP with 5 days history of sudden onset, severely painful red left eye. She described the pain as unilateral, periocular, stabbing in nature, associated with nausea and photophobia. Her previous medical history included seronegative arthritis which was treated with non-steroidal anti-inflammatory drugs, reduced appetite and 10kg weight loss over the past 6 months.

Vision acuity was reduced to hand movements in the left eye (6/9 on Snellen chart in the right eye). Slit-lamp examination revealed 360 degrees of scleral and conjunctival injection, which was most significant on inferior-nasal aspect overlying a peripheral haemorrhagic choroidal effusion seen on dilated fundoscopy (Figures 1 & 2). Her intraocular pressure was normal measuring 18 mmHg in both eyes on initial presentation.

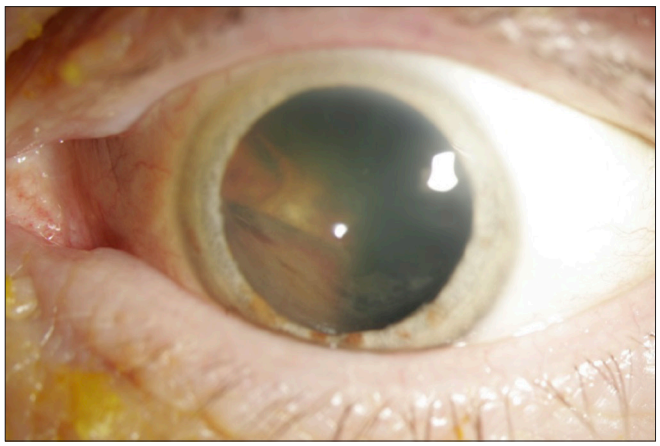

Figure 1: Mass visible in posterior segment.

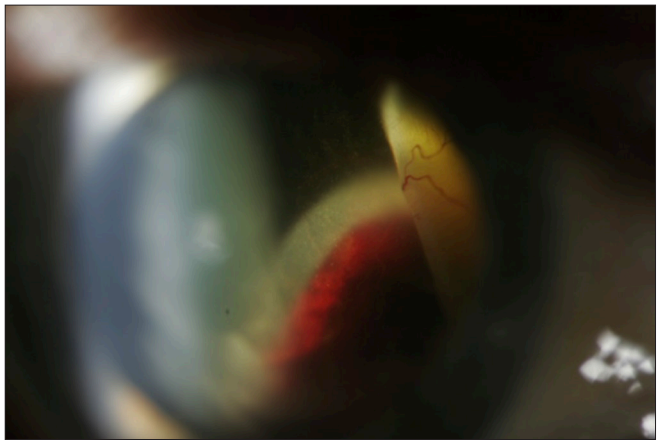

Figure 2: Peripheral haemorrhagic choroidal effusion seen on dilated fundoscopy.

B-scan ultrasonography of the globe (Figure 3) showed an inferior-nasal echogenic mass which was confirmed arising from the nasal Choroid on Computed Tomography (CT - Figure 4) and Magnetic Resonance Imaging (MRI - Figure 5) scans of the orbit and associated with exudative retinal detachment. The mass did not show any obvious extension outside the globe. There was high signal on T1 MRI, suggestive of choroidal melanoma.

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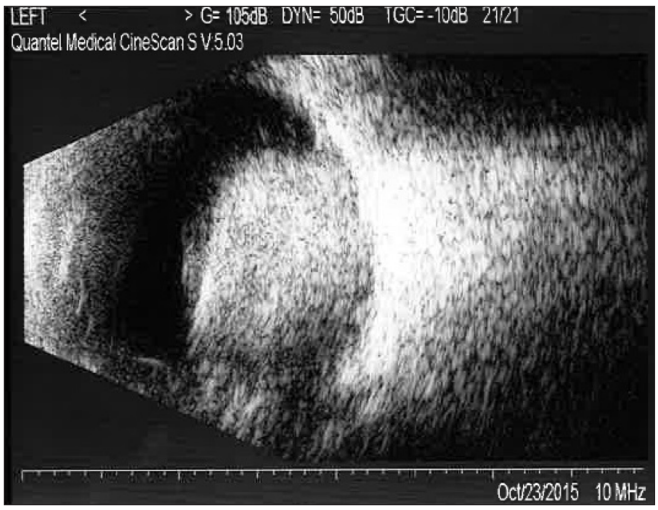

Figure 3: B-scan ultrasonography showing an echogenic mass.

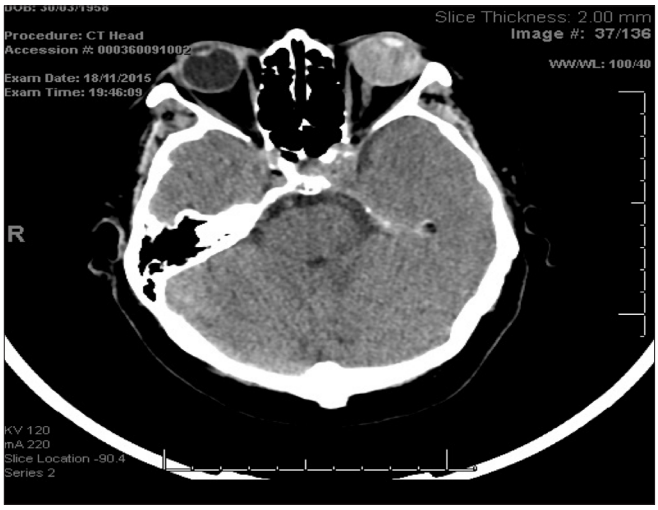

Figure 4: CT Scan showing hyperdense mass in left orbit.

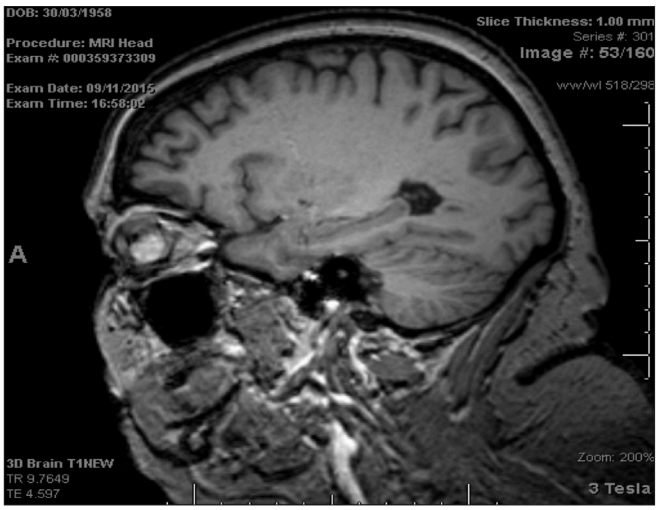

Figure 5: MRI showing intraocular mass.

Our differential diagnosis was exudative choroidal effusion secondary to scleritis or choroidal tumour, most likely melanoma associated with scleritis.

The patient was followed up closely in our clinic. An urgent regional Ocular Oncology referral was arranged. She was started on high-dose oral Prednisolone following initial presentation, which significantly relieved her pain and settled her scleritis. However, her vision did not improve, nor did the size of the choroidal mass change on follow-up.

Before being seen by the Ocular Oncology team, she re-presented 2 weeks later to Accidents and Emergency with sudden severe pain in the affected eye due to raised intraocular pressure above 90mmhg. The choroidal mass had caused an acute secondary angle closure glaucoma. This did not respond to intravenous Acetazolamide. An emergency eye enucleation was carried out and histopathology examination of the eye has confirmed a malignant choroidal melanoma.

Following enucleation, the patient had an ocular prosthesis fitted and has been monitored closely by the Oncology team. Her follow-ups included abdominal ultrasound and liver functions tests every 6 months, which have not showed any signs of metastasis.

Discussion

Scleritis usually presents with a painful red eye with or without vision loss. There are different forms of scleritis. It can be nodular or diffuse, necrotising or non-necrotising. Anterior scleritis, which is defined as scleral inflammation anterior to the extraocular recti muscle insertions is much commoner than posterior scleritis where the sclera posterior to the insertion of the rectus muscles is involved. Manifestations of posterior scleritis include serous retinal detachment, choroidal folds, or both. Symptoms of posterior scleritis include loss of vision as well as pain on eye movement (1).

Scleritis is associated with many connective tissue disorders, it is seen most commonly in women with Rheumatoid Arthritis. Other common risk factors include infections, trauma and exposure to radiation. There is no known HLA association with scleritis. Scleritis can be idiopathic (2,3).

In this case, we present an unusual cause of scleritis, a malignant melanoma. Typically, choroidal melanomas are asymptomatic when the lesion is small and sparing the macula, but they can present with decreased visual acuity, visual fields defects, flashes and floaters (4).

AN UNUSUAL CAUSE OF UNILATERAL PAINFUL RED EYE

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Slit-lamp and fundal examination

It is essential to carry out full dilated fundal examination in patients presenting with ocular inflammation, particularly when associated with reduced vision. Even though scleritis is a clinical diagnosis with signs of inflammation visible in anterior segment examination, this case has revealed a more sinister cause of ocular inflammation (5).

A diagnosis of choroidal melanoma is based on clinical features as a biopsy risks extraocular spread. The size, shape, thickness (over 2mm) and echodensitiy of the lesion are important ultrasound features. Dilated fundal examination of a suspicious lesion would show changes in colour (grey or pale/amelonotic), retinal elevation, associated subretinal fluid (exudative retinal detachment) and surface orange lipofuscin pigment (4).

Investigations

It was essential to rule out any underlying autoimmune disease, especially in this case, as the patient had previous history of seronegative arthritis. Laboratory investigations for scleritis did not show significant abnormalities in this patient These included complete blood count, erythrocyte sedimentation rate (ESR), serum autoantibody screen (including antinuclear antibodies, anti-DNA antibodies, rheumatoid factor, antineutrophil cytoplasmic antibodies), urinalysis, syphilis serology, serum uric acid and sarcoidosis screen (2).

In this case, further imaging was vital to confirm the presence of a large choroidal lesion. Scleritis can cause exudative choroidal effusion, however, detailed evaluation of the choroidal lesion by B-scan ultrasonography, CT and MRI scans of Orbits have been essential in establishing a diagnosis of choroidal melanoma which was later confirmed on histopathology analysis of the enucleated eye.

Management

The main aim of treatment of scleritis is to control the inflammation and minimise ocular tissue damage. Oral Non-Steroidal Anti-Inflammatory Drugs (NSAIDS) are usually the first line of treatment in mild to moderate scleritis. Topical steroids may reduce ocular inflammation; however, systemic steroids are particularly important in patients not responding to NSAIDS or in cases of posterior or necrotising scleritis (2).

Immunomodulatory drugs may be necessary in patients with chronic scleritis that is not adequately controlled on systemic steroids. Additionally, Rheumatology consultation and follow-up of patients on immunomodulatory drugs is recommended.

Whilst this patient presented with an unusual cause of scleritis, she responded well to systemic steroids, which reduced the ocular inflammation. On the other hand, her vision did not improve, and the size of the choroidal lesion did not change, which favoured the diagnosis of choroidal melanoma.

In this case the patient was unfortunate as she developed an acute secondary angle closure glaucoma, this is due to anterior displacement of the iris-lens diaphragm by the tumour. Therefore, she did not respond to glaucoma treatment as her aqueous outflow was totally obliterated.

Choroidal melanomas can erode through the sclera. In view of the large size of her tumour and the secondary glaucoma an enucleation was performed. Treatment options for smaller choroidal melanomas include observation, laser photocoagulation, photodynamic therapy, proton beam radiotherapy, stereotactic photon beam irradiation, plaque brachytherapy and local resection. An effective chemotherapy regimen has not yet been found. Neither enucleation nor local treatments such as radiation therapy have been shown to alter mortality rates (5,6).

Conclusion

This is an unusual case of unilateral scleritis secondary to an underlying malignant choroidal melanoma. It highlights the importance of full dilated fundal examination in patients presenting with an ocular inflammation. Multidisciplinary approach and close monitoring of patients with suspected ocular tumour is vital.

Multiple Choice Questions

1: All the following conditions could present with visual disturbance EXCEPT?

a. Scleritis

b. Episcleritis

c. Uveitis

d. Choroiditis

e. Optic Neuritis

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2: A 32 years old male presents with painful, red, photophobic left eye with mildly blurred vision for 4 days. On slit-lamp examination there are cells and flare in the anterior chamber and pupil is sluggish to react to light. What is the single most appropriate clinical diagnosis?

a. Acute angle closure glaucoma

b. Acute conjunctivitis

c. Acute dacrocystitis

d. Acute Iritis

e. Corneal foreign body

3: Which of the following is TRUE? Choroidal melanomas are usually associated with:

a. Yellow surface drusen

b. Orange lipofuscin

c. Red desaturation

d. Absent red reflex

e. Malignant melanoma of the Skin

4: Which of the following is NOT true? Scleritis can?

a. be present in a white eye

b. be painless

c. cause choroidal effusion

d. induce myopia

e. result in an ocular perforation

5: Which of the following features is likely to have higher risk of malignant transformation in a choroidal naevus?

a. Associated symptoms

b. Thickness <2mm

c. Absence of subretinal fluid

d. Presence of drusen

e. Peripheral (>3mm from Optic disc)

Answers for MCQ's

1. b - Episcelritis

Episcleritis is a benign, self-limiting condition, characterised by localised inflammation of the episcleral tissue, with or without pain. Reduced vision is not a feature of Episcleritis. Symptoms of severe pain or discharge should prompt an alternative diagnosis (7).

2. d - Acute Iritis

The presence of white blood cells and flare in the anterior chamber is a specific sign of acute anterior uveitis (i.e. Acute Iritis). It is essential to carry out a dilated fundal examination to rule out posterior uveitis, which is managed differently (8).

3. c - Orange lipofuscin

Choroidal melanomas usually present with an elevated domed-shaped grey lesion of the choroid with ill-defined margins. A choroidal lesion is highly suspicious of melanoma if there are any of the following: colour changes (grey or yellow), thickness > 2mm, associated with subretinal fluid, an orange lipofuscin pigment, sentinel vessels and/or secondary glaucoma (4,6).

4. d - Induce myopia

Posterior scleritis may present with white eye, it does not induce myopia. A typical feature of posterior scleritis is presence of the T-sign on B-scan ultrasonography. Necrotising scleritis can be painless and result in a scleral melt (1).

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5. a - Associated symptoms

Choroidal melanomas may arise from benign choroidal naevi. If 3 or more of the following factors are present in a naevus, there is an estimated risk greater than 50% of malignant transformation (6):

- *Thickness > 2 mm.*
- *Subretinal fluid.*
- *Associated symptoms.*
- *Presence of orange pigment.*
- *Margin within 3 mm of the optic disc.*
- *Ultrasonographic hollowness.*
- *Absence of surrounding halo. Naevi usually show a surrounding clear halo consisting of atrophied retina.*
- *Absence of drusen.*

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ASSESSMENT OF ACUTE ONSET DOUBLE VISION

L Butler, M Edington, C Diaper

Abstract

Sudden onset diplopia is a relatively common presenting complaint. Given the myriad causes of double vision, it can be difficult to assess and come to a diagnosis, particularly for doctors with limited ophthalmology or neurology experience.

This article uses a validated algorithm, The Edinburgh Diplopia Algorithm, to provide a simple framework for carrying out this assessment and allow the doctor to come to the most likely diagnosis.

While any patient with true diplopia requires referral to ophthalmology for further assessment and management, it is still extremely important that the referring clinician comes to a likely diagnosis prior to referral. This not only greatly aids in the triage of these patients but also avoids unnecessary delay in the patient having appropriate investigations performed.

Introduction

Many junior doctors find it difficult to assess sudden onset diplopia and to diagnose the underlying pathology. Given that the causes of diplopia can range from simple benign conditions such as cataract to life-threatening conditions such as an aneurysm, and that most junior doctors have had less than two weeks experience in an ophthalmology rotation, (1) it is not surprising that there is some uncertainty in dealing with these patients.

This article aims to provide a simple framework for assessing patients with double vision, based on a previously published and validated algorithm: The Edinburgh Diplopia Algorithm (Figure 1) (2).

This algorithm is designed to allow the user to come to the most likely diagnosis. It is important to note that this framework needs to be used alongside a thorough clinical history and examination.

Figure 1: The Edinburgh Diagnostic Diplopia Algorithm (1).

Assessment

Initial assessment

The first step in assessing diplopia is to establish if this is true diplopia or whether the patient actually perceives blurring of their vision as "double vision".

It is important to establish that the patient is definitely seeing two separate images, rather than just shadowing around objects or blurred edges. The symptom of blurred vision will involve a different diagnostic path, which is not covered in this article.

Once true diplopia has been confirmed, the next step is to determine whether it is monocular or binocular. This can be done by asking the patient to cover each eye in turn (or doing it for them if they are unable).

If the diplopia resolves when either eye is covered but is present when both eyes are open, it is binocular diplopia. If it remains when one eye is covered then it is monocular diplopia.

Monocular diplopia

Monocular diplopia is fairly uncommon and is usually due to light diffraction, either caused by an irregularity in the cornea (such as a scar or ectasia) or lens (such as cataract) (3). If the diplopia improves or resolves when the patient looks through a pinhole, it is likely due to light diffraction, and the patient can be referred routinely to ophthalmology for further investigation.

If diplopia remains despite looking through a pinhole, cortical pathology should be considered, particularly in the presence of any neurological signs. Pinholes can be found on simple occluders (Figure 2) or can be made very easily with a plain sheet of paper and white needle or pin (Figure 3).

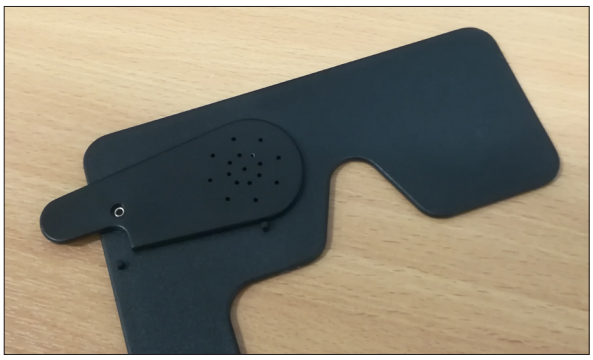

Figure 2: A pinhole occluder.

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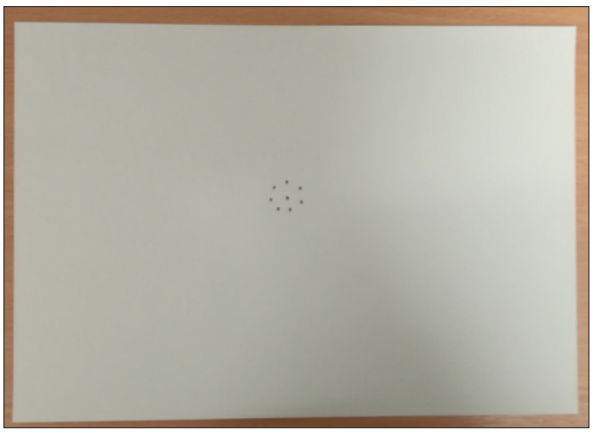

Figure 3: A makeshift pinhole occluder, created using a sheet of paper and a pin. The tip of a ballpoint pen could also be used.

Binocular diplopia

Having established that the patient has true binocular diplopia, the next question to ask is how the two images are separated: vertically or horizontally. It may be easier to ask patients if the images are "side by side" or "one above the other".

Horizontal diplopia

Sudden onset horizontal diplopia is most likely due to a cranial nerve VI (CN VI) palsy or an internuclear ophthalmoplegia (INO). (4) To separate these two diagnoses, ask the patient to look in the direction that makes the diplopia worse. Assess the position of their eyes when they do this: if they diverge then an INO is more likely; if they converge think about a CN VI palsy (Figure 4).

An abducens nerve (CN VI) palsy is the most common cranial nerve palsy due to its long path, making it vulnerable to direct and indirect lesions. The affected eye will be unable to abduct due to paralysis of the lateral rectus muscle. Visual acuity, pupil reactions, and lid position should be normal. In the absence of trauma or signs of raised intracranial pressure, the most common cause is microvascular ischemia.

If patients have a confirmed diagnosis of hypertension, diabetes, or hypercholesterolaemia, they will not necessarily require urgent imaging. Baseline blood tests (FBC, U+Es, CRP, glucose, and cholesterol) as well as blood pressure should be documented, medical treatment optimised, and patient referred for formal orthoptist assessment.

Patients with suspected microvascular CN VI palsy will likely be monitored in the first instance; if their symptoms are progressive or if there is absence of improvement after six weeks, they should have an MRI of head and orbits. (5)

INO is a disorder of conjugate lateral gaze caused by injury or dysfunction of the medial longitudinal fasciculus. The affected eye will show an impairment in abduction, while the unaffected eye will abduct with nystagmus (Figure 4).

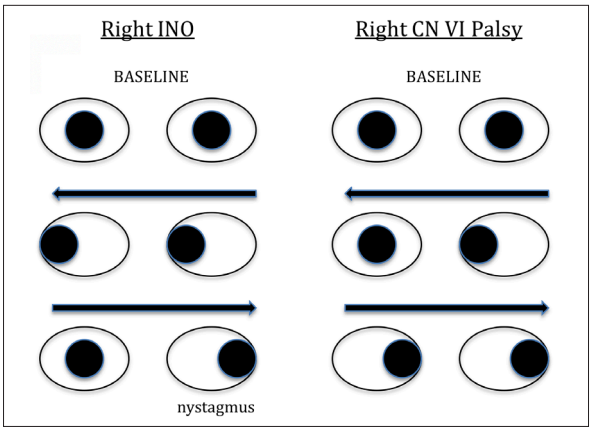

Figure 4: Schematic showing the different eye movements in INO and CN VI palsy.

The impairment may be a slowing of the movement rather than complete paralysis in abduction. (6) Depending on patient age, the most common causes of INO are multiple sclerosis or brainstem infarction (7) and these patients will need urgent brain imaging in the form of MRI.

Vertical Diplopia

If a patient describes vertical diplopia, cranial nerve III or IV palsy is most likely. In an oculomotor (CN III) palsy, the eye will be deviated down and out due to medial rectus, inferior rectus, superior rectus, and inferior oblique muscle paralysis. In a complete CN III palsy, there will also be lid ptosis and pupil mydriasis (dilation) on the affected side. The oculomotor nerve supplies the sphincter pupillae and the levator muscle.

ASSESSMENT OF ACUTE ONSET DOUBLE VISION

L Butler, M Edington, C Diaper

The pupillary fibres of the occulomotor nerve run superficially so are likely to be involved if there is a compressive lesion. Urgent imaging in the form of CT/MRI angiogram is therefore required in patients with pupillary involvement to rule out a posterior communicating artery aneurysm, particularly if there is associated headache or periocular pain.

It is possible to have a partial CN III palsy, where there is no pupil or lid involvement, or where not all the extra-ocular muscles supplied by the occulomotor nerve are affected. These cases are more likely to be microvascular in nature, are less urgent than a complete CN III palsy, and may not always require neuroimaging. (8) Always discuss such cases with an ophthalmology or neurosurgery specialist.

In a trochlear nerve (CN IV) palsy, the patient will often describe images as being 'tilted' due to paralysis of the superior oblique muscle which controls upgaze and torsion (rotation) of the eye. As the trochlear nerve is the thinnest and longest intracranial nerve, it is particularly susceptible to traumatic injury.

In the absence of head trauma, the most likely underlying cause for an acute CNIV palsy is microvascular, so a detailed history as well as baseline blood tests and blood pressure are useful. In isolation, CNIV palsy is rarely caused by aneurysms, demyelination, or tumours (9); however, MRI head should always be considered in a new onset palsy to rule this out.

It is also possible to have a sudden decompensation of a long standing congenital CN IV palsy; the patient would normally exhibit facial asymmetry and head tilt to compensate for the diplopia, so past photographs can be very useful to determine this. If facial asymmetry and head tilt are present in past photographs, no further investigation is generally required.

Another common cause of sudden onset vertical diplopia is thyroid eye disease (TED). TED causes a restrictive myopathy due to swelling and inflammation of the extraocular muscles. The most commonly involved muscle is the inferior rectus, hence the vertical diplopia. TED patients can, however, also present with horizontal diplopia if the medial rectus is affected. (10)

In this condition there are likely to be other signs of thyroid orbitopathy such as lid retraction or proptosis. There may also be a history of thyroid dysfunction. If TED is suspected it is important to carry out thyroid function tests to assess thyroid status. Always assess lid closure, visual acuity, pupil reactions, colour vision, degree of proptosis, and perform optic disc examination prior to referral as this will affect how urgently the patient will need to be seen by the ophthalmologist.

Other causes of double vision

The Edinburgh Diplopia Algorithm does not include all the causes of sudden onset double vision; only the most common ones. (3) An important cause of diplopia that is not covered and is worth mentioning is myasthenia gravis (MG). It is often referred to as "the great imitator" as it can present with a range of neurological signs that can change throughout its course. (11)

From the diplopia point of view, the vertical muscles tend to be affected the most, but the limitation is variable and does not correspond to any nerve palsy. Patients also often display unilateral or bilateral ptosis which can vary throughout the day. Fatiguability can be assessed by measuring the patient's lid position when looking straight ahead, and then asking them to look up for 90 seconds. Often one can notice an increase in ptosis towards the end of the test.

Cogan's lid twitch is another useful test. Ask the patient to rapidly refixate their eye from downgaze to the primary position (straight ahead). The sign is positive if there is overshooting of the upper lid before settling down to the ptotic position.

If there is suspicion of MG then it is useful to send a blood test for acetylcholine receptor antibodies, which will be detected in 85% of generalized MG and 60% of purely ocular MG. (12) Further discussion about diagnosis of MG is outwith the scope of this article and will most likely be carried out by an ophthalmologist in conjunction with a neurologist.

An additional diagnosis not covered by the algorithm is that of a decompensated phoria. Like MG this can present in a number of ways. Many people have a latent tendency for ocular misalignment, heterophoria, that may become apparent, or manifest, under certain conditions such as stress or fatigue. (13)

Patients with a decompensated phoria will likely describe their diplopia as being intermittent, worse in the evening, when they are tired or when they have had alcohol. Examination will reveal normal eye movements, although deviation of the eyes can be induced on cover testing. (14)

There should not be any lid ptosis. While a decompensated phoria is a relatively benign cause of diplopia, patients should still be referred to From the diplopia for an orthoptic assessment and to rule out any other, more serious conditions such as MG.

ASSESSMENT OF ACUTE ONSET DOUBLE VISION

L Butler, M Edington, C Diaper

Conclusion

The framework described above is based on a validated algorithm that aids the user in coming to a "most likely" diagnosis. It is simple and quick to use and requires no special equipment, based largely on the skill of the clinician in eliciting specific symptoms and observing simple eye movements.

Any patient with true diplopia requires referral to ophthalmology for further assessment and management. It is still extremely important, however, that the referring clinician comes to a likely diagnosis prior to referral. This not only greatly aids in the triage of these patients but also avoids unnecessary delay in the patient having appropriate investigations performed.

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ASSESSMENT OF OPTIC DISC SWELLING

M Edington, L Butler, C Diaper

Abstract

Optic disc swelling is a clinical finding that may be the result of a variety of different conditions. The aim of this article is to review these causes and consider how a differential diagnosis can be made following good history taking and examination, which subsequently will guide appropriate investigations and referrals.

Bilateral involvement (papilloedema) is most commonly due to raised intracranial pressure, but other diagnoses such as malignant hypertension, space occupying lesion, and infection or inflammation should be considered. Unilateral disc swelling is most commonly due to anterior ischaemic optic neuropathy (AION) or optic neuritis. Any patient with suspected optic nerve pathology should have a baseline assessment of their optic nerve function prior to referral to ophthalmology, so that appropriate management and follow up can be established.

Introduction

Optic disc swelling (Fig 1) is a pathological condition with a variety of causes. It is important to distinguish between bilateral and unilateral swelling in order to guide the differential diagnosis. All patients will require an in-depth history taking, as well as assessment of their optic nerve function, to determine urgency of referral to ophthalmology or other specialties and further management.

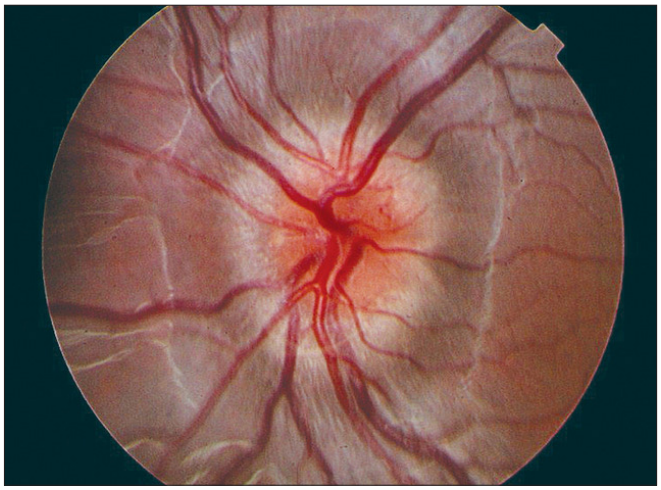

Figure 1: Classic appearance of a swollen optic nerve head, visible on ophthalmoscopy (https://creativecommons.org/licenses/by-nc/2.0/)

Optic nerve function assessment

Any patient with suspected optic nerve pathology should have a complete assessment of optic nerve function. A clinician of any specialty can carry out the majority of this assessment, and adequate baseline information upon referral to ophthalmology will be invaluable in appropriately triaging patients.

A detailed history of ophthalmic and neurological symptoms should be documented, including: characteristics, onset and duration of any headache, visual disturbance or periocular pain; past medical and ophthalmic history; and detailed drug history.

Baseline examination should include: visual acuity; colour vision (ideally using Ishihara colour plates, but if these are not available, a simple red desaturation test should be attempted); pupil reactions including checking for a rapid afferent pupillary defect (RAPD); visual fields to confrontation to map out any gross field defect or enlarged blind spot; and, finally, dilated fundus examination. (1)

This baseline assessment will usually provide enough information to establish a differential diagnosis. Patients will then be either referred to ophthalmology for further investigation with ocular ultrasound, optical coherence tomography (OCT) of the discs, and formal Goldman's visual field testing, or to the medical team for more urgent investigation with brain imaging +/- lumbar puncture.

Papilloedema

Papilloedema by definition is bilateral optic disc swelling, usually due to raised intracranial pressure. The most common cause is idiopathic intracranial hypertension (IIH). The stereotypical patient is a young overweight female (the mnemonic of 'female, fertile, fatty' is sometimes used). (2)

If the patient demographic is not typical, history should be directed towards other known causes of raised intracranial pressure, including underlying medical conditions such as migraines, hydrocephalus or previous trauma, and medications such as oral contraceptives, high dose vitamin A derivatives, and long term tetracycline antibiotics. (2)

Patients with papilloedema secondary to IIH are often asymptomatic with optic nerve swelling being an incidental finding. Others complain of pressure headaches- usually throbbing in nature, worse in the morning, and exacerbated by coughing, recumbency, or exertion. There may be associated nausea, tinnitus, and visual disturbance such as blurring or distortion, but if visual loss is present it is usually mild. (3) Patients rarely appreciate a visual field defect.

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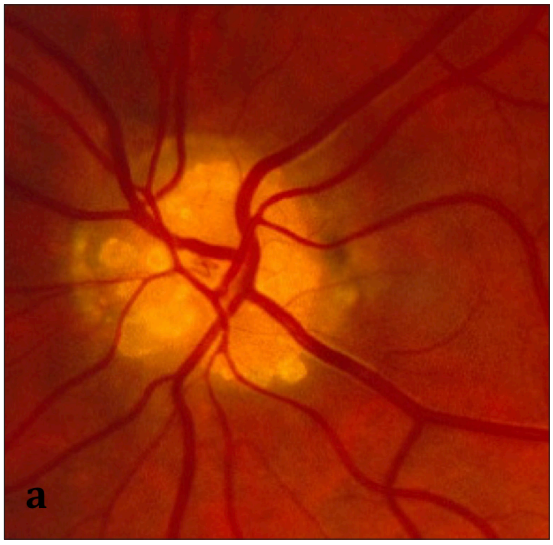
A diagnosis of IIH should not be made without ruling out other causes of bilateral optic disc swelling, including malignant hypertension, space occupying lesions, venous sinus thrombosis, and inflammatory, infiltrative, or infectious causes. If there are associated features such as headache and nausea, the patient should be assessed promptly by the medical team and investigated with MRI/MRV head and lumbar puncture (for diagnosis of IIH the CSF must be normal aside from increased opening pressure).

If the patient is completely asymptomatic, they should be referred to ophthalmology in the first instance in order to differentiate between true papilloedema and 'pseudopapilloedema', and avoid unnecessary testing. In papilloedema, the disc swelling is usually symmetrical, and nerves are often hyperaemic, with nerve fibre layer opacification. Peripapillary vessels will often be obscured, with loss of spontaneous venous pulsations, venous dilation, disc haemorrhages, and loss of distinct central cup. (4,5,6)

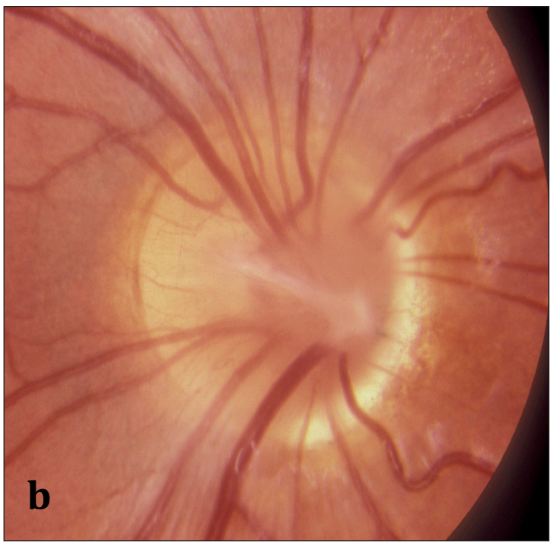
Paton's lines (circumferential retinal folds radiating from the optic nerve) may also develop. Visual acuity is usually preserved, with normal pupil reactions. Colour vision may be normal or slightly reduced, and the typical visual field defect is an enlarged blind spot, although there can be generalised constriction and even complete field loss in later stages.

Pseudopapilloedema is a benign condition, usually caused by optic disc drusen- deposits of protein, mucopolysaccharides, and calcium salts (Figure 2a). Tilted optic nerve heads (Fig 2b)7, hypoplastic discs with small cup to disc ratio, and peripapillary atrophy (Fig 2c) can also sometimes be confused for disc swelling.

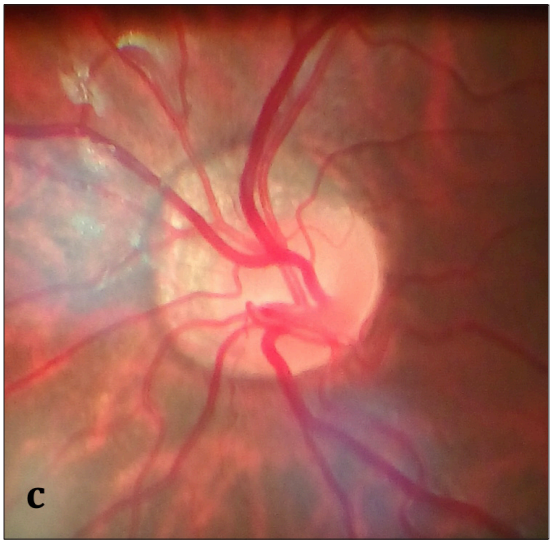
Figure 2



a - The typical, lumpy, yellowish appearance of optic nerve head drusen (http://www.mrcophth.com/opticdisccases/drusen.html)



b – The pallor of optic disc atrophy surrounding the optic disc that can be confused with swelling (Published under the creative commons liscence) (7)



c – A tilted optic disc (https://creativecommons.org/licenses/by-sa/4.0/deed.en)

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Pseudopapilloedema may be bilateral or unilateral. On examination, there should not be hyperaemia, and disc vessels should be clearly visible, although anomalous vasculature is often found (increased number of vessels at the disc, abnormal branching, and increased tortuosity). (4,5,6)

If drusen are superficial, distinct yellowish 'bumps' can sometimes be seen. Buried drusen are more difficult to assess and often additional testing with ocular ultrasound, autofluorescence, optic disc OCT, or fundus fluorescein angiography is required. (8) In instances where optic disc drusen are not clearly demonstrated, patients will still require further investigations with brain imaging +/- lumbar puncture.

Optic Neuritis (ON)

Optic neuritis is an inflammation of the optic nerve that typically affects young healthy adults, more commonly females. It can be idiopathic or a symptom of underlying demyelinating conditions such as multiple sclerosis (MS) or neuromyelitis optica (NMO). It can also be caused by certain autoimmune disorders, infections, and medications (Table 1), the clinical features of which are discussed in detail by Voss et al. (9)

Demylination	multiple sclerosis, neuromyelitis optica
Autoimmune disease	sarcoidosis, systemic lupus erythematosus, Sjögren's syndrome, Behchet's disease
Infections	herpes zoster, varicella zoster, lyme disease, syphilis, tuberculosis, cat scratch disease, β-hemolytic streptococcal infection, meningococcal infection
Drugs	quinine, tetracycline, amiodarone, ethambutol, isoniazid

Table 1: Aetiology of Optic Neuritis.

In typical ON, patients usually present with varying degree of monocular vision loss, which progresses for 7-10 days before stabilising. Most patients also report retro-orbital pain (worse on eye movement) and disturbance of colour vision. (10)

An RAPD should be present; reconsider the diagnosis if pupil reactions are normal. Up to 65% of patients will have retrobulbar neuritis and a normal optic disc appearance on presentation. (10)

If oedema is present, it may not correlate with degree of vision or field loss. Haemorrhages, cotton wool spots, and exudates are rare findings and tend to point toward a different diagnosis. Optic disc atrophy and pallor typically develops after 4-6 weeks. (10)

Patients should be referred for baseline Goldman visual fields. A central scotoma is often described as a typical finding; however, Jung et al described similar incidence of altitudinal defects, sectorial scotoma, arcuate scotoma, and enlarged blind spot in their case series. (11)

Uhthoff's phenomenon (worsening of vision with increased body temperature-for example while taking a bath) and Pulfrich phenomenon (a pendulum swinging in one plane appears to trace an ellipse due to asymmetrical optic nerve conduction velocity) have also been described but are not diagnostic of ON. (10)

Prognosis for visual recovery is generally good, and occurs over a few weeks-months. However, even if acuity improves most patients have some degree of residual abnormality in colour vision, contrast sensitivity, or visual fields. The Optic Neuritis Treatment Trial (ONTT) provides guidance on prognosis and treatment of ON. Results showed that while high dose steroids hasten visual recovery, treatment does not affect final visual outcome. (12).

Therefore, it is important to discuss options with the patient on an individual basis. As optic neuritis is the initial clinical manifestation of multiple sclerosis (MS) in up to 20% of patients, an MRI brain/orbits should be discussed. The ONTT found that the finding of white matter lesions is the strongest predictor for developing MS; presence of one or more lesions translates to an approximately 70% risk of developing MS at 15 years. (12)

Atypical ON is rare but it is important to recognize its features because prognosis and management are very different. Some of the 'red flags' include age of onset <12 or >50 years, severe visual loss (no perception of light) lasting > 2 weeks, lack of pain or pain lasting > 2 weeks, bilateral or sequential involvement, and deterioration after tapering of steroids. (9) Table 2 summarizes the features of typical versus atypical ON, which facilitate establishing a diagnosis and appropriately managing patients.

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Onset	Acute to subacute	Sudden
Age	Young adult patient (peak 15-50 years of age)	<12 or >50 years
Pain	Worse with eye movement	No pain or persistent pain > 2 weeks
Involvement	Usually unilateral	Simultaneous or sequentially bilateral
Examination	Reduced colour and contrast vision, any type of visual field defect, mild uveitis or periphlebitis possible, disc swelling with no haemorrhages or cotton wool spots	Severe vision loss (no perception of light), significant anterior and/or posterior segment inflammation, intensely swollen optic nerve head with multiple haemorrhages
Association	Multiple sclerosis	Neuromyelitis optica spectrum disorders, sarcoidosis, vasculitis and lupus; history of neoplasia
Prognosis	Generally good, VA tends to improve spontaneously over a few weeks	Usually poor; absence of recovery after > 3 weeks
Steroids	Hasten recovery but do not affect final visual outcome	No effect on recovery, relapse on tapering of steroids

Table 2: Differentiating between typical and atypical optic neuritis.

Anterior Ischaemic Optic Neuropathy (AION)

AION is a major cause of significantly impaired vision among the middle-aged and elderly population. (13) It can be subdivided into arteritic and non-arteritic causes. It is more common in Caucasians, with a slight female predominance in the arteritic group. (13) The underlying pathology is ischaemia due to interrupted blood supply from arteries supplying the optic nerve head.

Patients with arteritic AION usually complain of sudden onset vision loss, which, depending on time of presentation, can be mild to severe. There is usually marked optic disc swelling and 'chalky' pallor, with multiple flame haemorrhages and cotton wool spots. (13)

RAPD will be present, as well as a significant field defect (typically inferior altitudinal in early stages, progressing to generalised loss). The optic disc in the fellow eye should be anatomically normal. Within 6-8 weeks the optic disc develops atrophy with cupping similar to that seen in glaucoma, but with pallor of the remaining neuroretinal rim. (13) There may be other signs of ischaemia, such as central retinal artery occlusion, choroidal infarction, and diplopia secondary to extraocular muscle involvement so a full ophthalmological assessment is necessary.

Arteritic AION is most commonly caused by temporal arteritis (also known as GCA- giant cell arteritis), but it can also be caused by other vasculitides and connective tissue disorders such as polyarteritis nodosa, Wegener's granulomatosis, and systemic lupus erythematosus. (13)

One should ask specifically about history of polymyalgia rheumatica, as the two conditions often occur together. Patients with GCA typically have systemic symptoms including headache, temporal and scalp tenderness, jaw claudication, malaise, weight loss, myalgia, and fever. (13)

The temporal arteries may be promient, ropey, and tender to touch, with loss of palpable pulsation. However, all patients >50 with sudden onset unilateral vision loss and disc swelling, even without systemic symptoms, should be investigated for GCA with urgent ESR, CRP and platelet count, as delayed treatment can result in bilateral vision loss in up to 50% of cases, as well as systemic complications such as stoke or myocardial infarct.

If GCA is suspected, the patient should be started on high dose steroids immediately, without waiting for confirmation of the diagnosis with temporal artery biopsy. Additionally, it is worth remembering that while a positive temporal artery biopsy result is useful to confirm the diagnosis and justify long-term steroid treatment, a negative result does not exclude GCA. It is therefore important to rely on history taking and examination skills to guide management.

Non-arteritic AION is characterised by a variable degree of sudden painless loss of vision (usually not as severe as in arteritic AION). Visual loss is often noticed on waking, and nocturnal hypotension has been hypothesized as a possible cause. (13)

Patients typically do not have associated ocular or systemic symptoms, but should be screened for GCA with urgent blood tests. RAPD is common, but may not be present if visual loss is mild. Sectoral disc swelling, particularly of the superior disc, is common, with a corresponding altitudinal field defect (inferior or infero-nasal). (13)

However, other patterns can also occur. Disc pallor is less common than in arteritic AION; flame haemorrhages and localised arterial attenuation may be present. It is important to examine the fellow eye, as patients often have a small, crowded 'disc at risk' with anatomical predisposititon for ischaemia. (13)

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Bilateral involvement is less common than in arteritic AION (up to 20% reported in some studies), and usually occurs sequentially rather than simultanously. Atherosclerosis has been implicated as the underlying pathology, therefore management of patients' cardiovascular risk factors such as hypertension, diabetes, and hypercholesterolaemia is essential. There is no proven effective therapy, although aspirin has been suggested to reduce the risk to the fellow eye.

Other differential diagnoses

Other causes of optic nerve swelling include inflammatory (sarcoidosis), infectious (toxoplasmosis, cat-scratch disease, viral encephalitis), and compressive (thyroid eye disease, nerve sheath meningioma, glioma) optic neuropathy, as well as diabetic papillopathy.

A detailed description of these conditions is outside the scope of this article, but usually patients will have other ocular and systemic signs of a pathological process, such as fever, headache, anterior chamber or vitreous cells, retinal lesions, proptosis, or limitation of eye movements.

Conclusion

There are numerous causes for optic disc swelling. Bilateral involvement is most commonly due to raised intracranial pressure, but other diagnoses such as malignant hypertension, space occupying lesion, and infection or inflammation should be considered. Unilateral disc swelling is most commonly due to AION or optic neuritis, and patient demographics as well as a detailed history and examination will help differentiate between them. Any patient with suspected optic nerve pathology should have a baseline assessment of their optic nerve function prior to referral to ophthalmology, so that appropriate management and follow up can be established.

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ATYPICAL RETINITIS PIGMENTOSA MASQUERADING AS NORMAL TENSION GLAUCOMA

EH Shao, A Lewis, E Ansari

Abstract

A 60-year-old lady first presented to our ophthalmology department in 2003 with intermittent blurred vision. Her visual acuity was 6/12 in the right eye and 6/9 in the left eye. She had normal intra-ocular pressures and her optic nerves looked healthy.

Visual field testing showed bilateral superior arcuate defects, which were unexplained by her eye examination. An MRI head was ordered, with normal results. She was given a working diagnosis of normal tension glaucoma.

However, despite being placed on glaucoma treatment, her superior visual field defect gradually worsened over 5 years. A repeat dilated fundus examination revealed pigmented speckling of inferior retina in both eyes. She was subsequently referred for retinal electrophysiology and diagnosed with sectoral retinitis pigmentosa.

Our case highlights the importance of having a solid grasp of different patterns of visual field defects in different pathologies, and of checking that visual field defects match the clinical findings.

Background

Visual field testing is an important investigation in the evaluation of visual function of patients with many different types of pathologies, in particular those affecting the optic nerve, as well as neurological and retinal conditions. However, as the test is also highly subjective, it is essential to differentiate true, disease-related defects and abnormalities from artefact and noise.

Glaucoma affects more than 70 million people worldwide with approximately 10% being bilaterally blind [1]. As such, it is a worldwide leading cause of irreversible vision loss [2]. Because it may be asymptomatic until a relatively late stage, diagnosis is frequently delayed [2].

Glaucoma is characterised by vertical optic disc excavation or cupping secondary to ganglion nerve cell loss, characteristic visual field defects and raised intraocular pressure in most cases [2].

However, other conditions can mimic glaucoma because of the presence of optic neuropathy, or visual field defects, or both. The clinician must be vigilant in being able to differentiate true glaucoma from other conditions that can masquerade as glaucoma.

Retinitis pigmentosa (RP) is a group of conditions characterised by progressive dysfunction, cell loss, and atrophy of retinal tissue leading to sight loss and blindness. There is currently no treatment or cure for this debilitating condition.

Sectoral or atypical retinitis pigmentosa is a rare subtype, with fundoscopic findings limited to a segmental area. With its muted expression and visual field defects, it can mislead ophthalmologists in diagnosing glaucoma and neurological problems.

This case highlights the importance of careful interpretation of visual fields alongside clinical findings.

Case Presentation

A 57-year-old lady initially presented to another ophthalmology department in 2000 with bilateral fluctuating blurred vision and 'zig-zag' photopsia in the right eye. She had a background of hypertension, hypercholesterolaemia and hypothyroidism post thyroidectomy.

Her mother had glaucoma; there was no other family history of ocular conditions. She was extensively investigated and found to have bilateral early cataracts, normal optic nerves , "longstanding chorioretinal atrophic changes" in the peripheral retina of the left eye, and bilateral superior arcuate field defects. MRI head was normal and the patient was discharged from ophthalmology with reassurance.

Three years later, she presented to our ophthalmology department with ongoing visual symptoms . Visual acuities were 6/12 in the right eye and 6/9 in the left eye. She had normal intra-ocular pressures (IOP) of 18 in both eyes and her optic nerves looked healthy.

An initial dilated fundus examination revealed a posterior vitreous detachment but no other abnormality. Visual field testing showed bilateral superior arcuate defects, which were unexplained by her clinical findings (figure 1). An MRI head was repeated, with normal results. She was monitored in the general clinic, and her superior visual field defect gradually worsened over the next five years.

ATYPICAL RETINITIS PIGMENTOSA MASQUERADING AS NORMAL TENSION GLAUCOMA

EH Shao, A Lewis, E Ansari

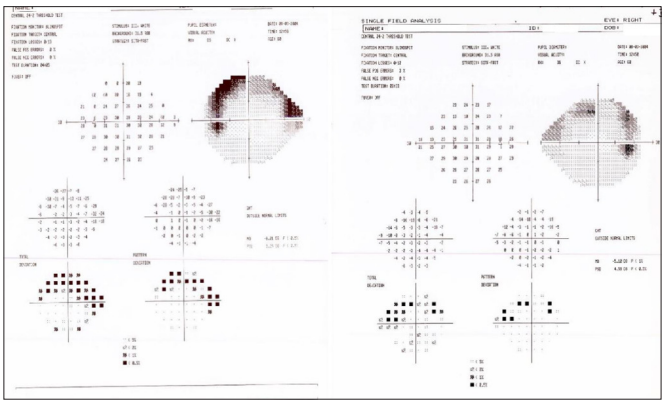

Figure 1. Presenting visual field defect.

It was finally noted by a glaucoma consultant that her optic disc appearance (cup disc ratio of 0.5 with normal neuro-retinal rims) was inconsistent with her field defects. A repeatdilated fundus examination revealed pigmented speckling of inferior retina in both eyes (figure 2).

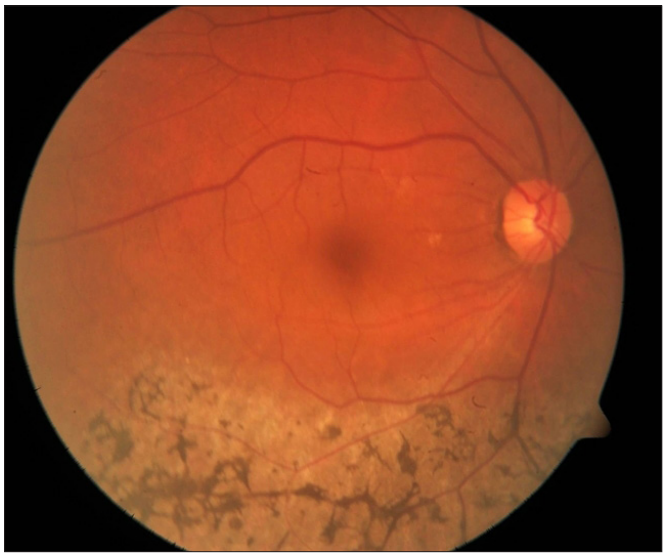

Figure 2. Right fundus photograph of sectoral retinitis pigmentosa.

The patient was referred for electrophysiology. Her ERG was abnormal with reduced b-wave amplitude [scotopic b-wave amplitude: 176.15 OD, 140.26 OS (normal 253.37 ± 57.32); scotopic b-wave amplitude: 30.73 OD, 27.64 OS (normal: 51.29 ± 10.35)] and prolonged b-wave implicit time [40.6 OD, 51.7 OS (normal: 23.32 ± 4.31)] (figure 3).

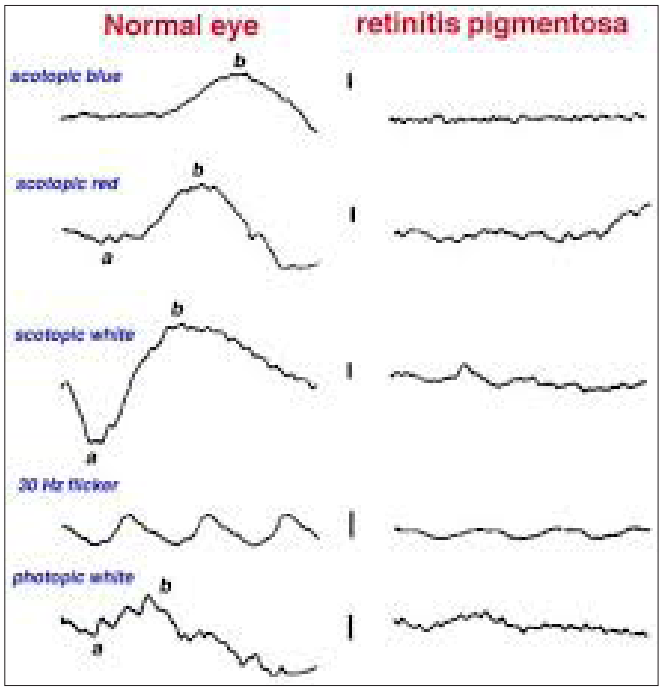

Figure 3. ERG of a normal eye and one with retinitis pigmentosa (From Clinical Electrophysiology).

She was diagnosed with sectorial retinitis pigmentosa and has since been referred for genetic counselling and online support groups. Her visual fields continue to gradually deteriorate although her central vision remains unaffected

Discussion

Retinitis pigmentosa is a blinding and debilitating disease for which there is currently no treatment. Sector or atypical retinitis pigmentosa is rare and its pathophysiology unclear. Two hypotheses have been suggested: a) anatomic or circulatory anomaly of blood vessels leading to hypo-perfusion with subsequent dysfunction and atrophy of retinal tissue, or b) localised retinal degeneration from an embryonic origin [3].

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Retinal changes are frequently muted and can be missed. As such, patients are often misdiagnosed as glaucoma due to the frequently associated peripheral field defects. In our patient, positive family history for glaucoma and no family history of retinitis pigmentosa (unusual in that the disease is autosomal dominant) also contributed to the initial misdiagnosis.

Retinochoroiditis and sectorial RP can masquerade as glaucoma because they lead to peripheral visual field defects including arcuate or wedge-like defects [5]. However, glaucoma typically presents with a characteristic vertical excavation of the optic disc and a pattern of field defects that is in keeping with areas of retinal nerve fibre loss (figure 4).

The axons maintain a retinotopic organization in the optic disc [4]. Neurones of the superior and inferior temporal retina do not mix, and respect the temporal raphe, thus glaucomatous field defects such as arcuate scotomas respect the nasal horizontal meridian [4].

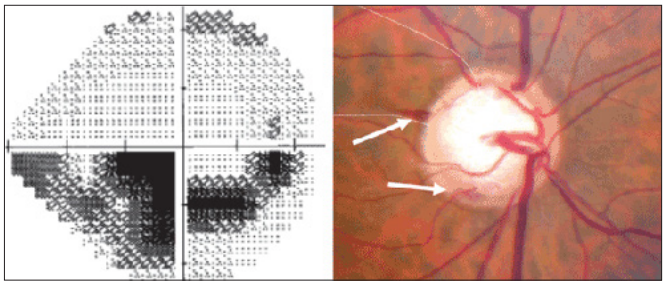

Figure 4. Glaucomatous optic disc with corresponding field defect (From Review of Ophthalmology)

Glaucomatous field defects typically "point to" the blind spot, continue in a peripheral arc (arcuate scotoma), going around the point of fixation (paracentral scotoma) before terminating abruptly at the nasal horizontal meridian (figure 5).

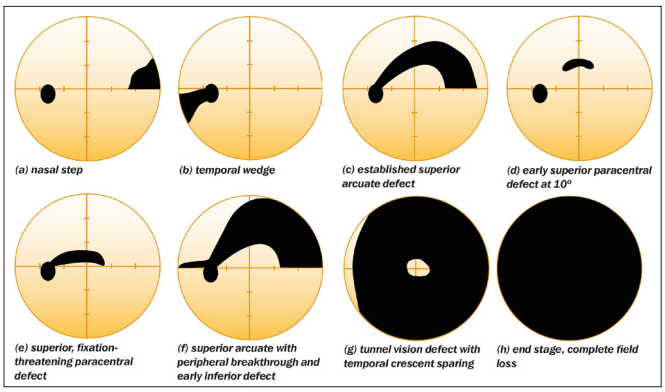

Figure 5. Glaucomatous visual field defects (From visual-field-software.com)

For several years our patient's field defect was wrongly interpreted as arcuate scotoma. By 2005, the scotoma crossed the horizontal meridian, which, in addition to normal disc appearance, should have alerted clinicians to consider an alternative pathology (figure 6).

Field defects caused by retinal lesions are frequently deep,have sharp borders, may not respect the horizontal midline, and tend to show much less variability from test to test than glaucomatous pathology [5].

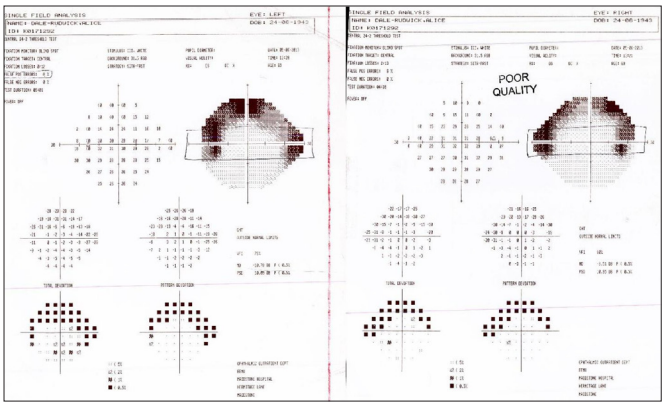

Figure 6. Progression of visual field defect

It is important to exclude neurological disease upon visual field testing, as conditions such as space occupying lesions and cerebrovascular events can result in patient mortality if left undetected without appropriate management.

Neurological conditions lead to visual field defects either side of the vertical meridian, for example homonymous superior quadrantonopias (temporal lobe lesion), inferior quadrantonopias (parietal lobe lesions), homonymous heminopias (parietal and temporal lobes, visual cortex), and bitemporal heminopias (optic chiasm) (figure 7) [6].

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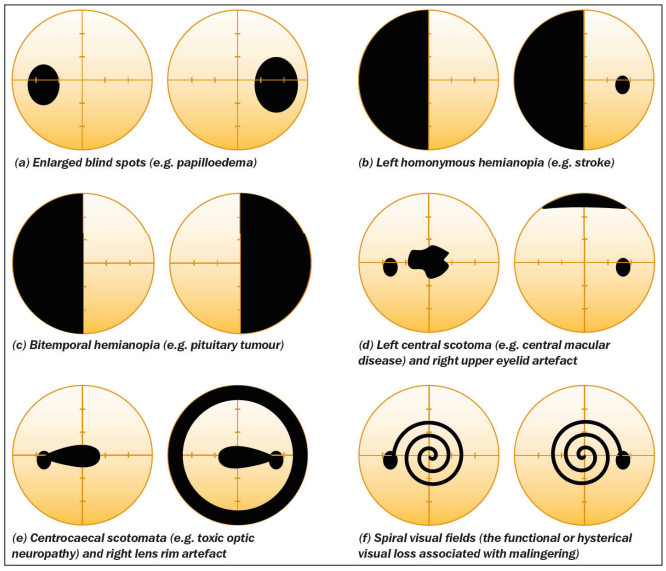

Figure 7. Non-glaucomatous visual field defects (From visual-field-software.com)

Ischaemic optic neuropathy results in altitudinal visual field loss, which can be misdiagnosed as glaucomatous field defect especially if the patient has co-existing glaucoma. It is important to note that large, sudden visual field changes are not typical in glaucoma.

Such changes are usually secondary to other pathology: neurological disease eg. optic neuritis, or vascular occlusions in the retina. If a large change is seen and part of the field loss seems hemianopic or occurs in the other eye as well, neurological causes need to be ruled out [4].

Often, several diseases coexist in the same eye, e.g. glaucoma and retinal disease, and it becomes important to determine whether encountered field loss is caused by one disease or the other as this will affect patient management. Dilated fundus examination is not routinely done in glaucoma clinics after the initial assessment; however, if optic disc appearance does not correspond to visual field defects, a dilated fundoscopy is mandatory to rule out other pathology.

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HOW GOOD A CATARACT SURGEON ARE YOU? THE IMPORTANCE OF RISK ASSESSMENT IN CATARACT SURGERY

M Hussain, S Patra

Abstract

The National Ophthalmology Database (NOD) was established by the Royal College of Ophthalmologists (RCO) in 2010 to collect data from electronic medical record (EMR) systems and perform the National audit for all cataract surgery undertaken in the National Health Service in England and Wales (1).

For the purpose of the National audit, two primary indicators of surgical quality were benchmarked; the rate of rupture of posterior lens capsule (PCR) during surgery and the rate of visual acuity (VA) loss attributable to the surgery (2,3).

This paper discusses how to perform a risk assessment of a patient undergoing cataract surgery in the context of PCR and VA loss. It also explains the meaning and application of risk stratification and risk adjustment in clinical practice.

Introduction

Cataract surgery involves replacing the cloudy natural crystalline lens of the eye with a clear artificial lens implant. The most common technique, known as phacoemulsification, involves removal of the cataract through a small wound (1.8 to 2.4 mm wide), using ultrasound energy.

The lens capsule, an epithelial membrane which encloses the lens, is opened anteriorly via a step known as capsulorhexis, while the posterior capsule of the cataract is left intact to provide the main support for the artificial lens implant.

The commonest indication for cataract surgery is reduced vision leading to difficulty with performing daily tasks. The main aim of cataract surgery in majority of cases is to improve visual function and according to the 2017 NOD audit report 94% of eyes with no ocular co-morbidities achieved a vision of 6/12 or better after cataract surgery (1).

Risk Assessment

Risk assessment of patients undergoing cataract surgery is a valuable clinical skill. It can reduce surgical complications and improve surgical outcomes. It aids the selection of appropriate cases for trainees based on their level of experience. It is used to risk-adjust surgical outcomes to reflect the case-mix complexity of individual surgeons. It will enable the surgeon to discuss the patient specific risks of a proposed procedure during informed consent (1,4,5).

The most common complication that can occur during cataract surgery is posterior capsular rupture (PCR), with or without vitreous loss (2). It is usually associated with increased morbidity such as the need for additional surgical interventions, a greater number of follow-ups, and a greater risk of post-operative complications.

The National Cataract Surgery audit of 55,567 cataract surgeries performed in the NHS between November 2001 to June 2006, identified several risk factors associated with PCR (2) (see Table 1).

In the audit the overall rate of PCR was 1.92%, but the risk was significantly higher if the cataract surgery was performed by a junior trainee (5.1%) rather than a more experienced surgeon (2).

The loss of VA following cataract surgery, is defined as loss of 3 logMAR lines VA or doubling of the visual angle (3). There are several risk factors associated with loss of VA (Table 2). The risk factor associated with the greatest loss of VA following cataract surgery is the occurrence of PCR during surgery (2). Although VA loss occurred in only 1.24% of patients undergoing cataract surgery, eyes with PCR during cataract surgery had a 5.74 times increased likelihood of VA loss (3).

The National Cataract Surgery audit also demonstrated that there was a cumulative increased risk of PCR and VA loss for patients when multiple risk factors were present (2,3). The predicted probability of PCR occurring in a complex patient with multiple risk factors increased from 0.76% to 50% or more when compared to a patient with a baseline risk profile (ie: no significant risk factors) (2).

It is therefore important that a thorough pre-operative risk assessment is carried out in all patients undergoing cataract surgery.

Age	Increasing age
Gender	Male
Diabetic status	Diabetic retinopathy
Pupil size	Small pupil (One that fails to dilate with pre-operative mydriatic eyedrops to at least 6 mm)
Axial length of the eye	Greater than 26 mm (long or myopic eye)
Grade of cataract	Brunescent/white cataract (advanced end-stage cataracts)
View of the fundus	No fundal view/ vitreous opacities (due to advanced cataracts, vitreous haemorrhage, small pupils etc)
Integrity of zonules	Pseudo-exfoliation (PXF)/ phacodonesis/ trauma (conditions associated with weak or compromised zonular support)
Integrity of the posterior capsule (PC)	Trauma, posterior polar cataracts, intravitreal injections (conditions with pre-existing damage to the PC)
Drugs	Patient taking Doxazosin
Ability to co-operate and lie still	Learning difficulties, dementia, claustrophobia, anxiety, Parkinson's, head tremor, language difficulties
Ability to lie flat	Any condition which makes it difficult for the patient to lie flat for at least 20 minutes
Surgeon grade	Trainee

Table 1: Risk factors associated with posterior capsule rupture during cataract surgery.

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Age	Greater than 70 years
Diabetic status	Diabetic retinopathy
Pupil size	Small pupil (One that fails to dilate with pre-operative mydriatic eyedrops to at least 6 mm)
Axial length of the eye	Equal or less than 23 mm (Shorter or hypermetropic eye)
Surgical complications	Posterior capsule rupture
Ocular co-morbidities	Age related macular degeneration (AMD) corneal pathology amblyopia previous vitrectomy
Surgeon grade	Trainee

Table 2: Risk factors associated with visual acuity (VA) loss attributable to cataract surgery.

Risk Stratification & Risk Management

Risk stratification is a process which allows patients to be categorised into high, moderate and low surgical risk tiers. It is based on accurate recording of all relevant risk factors and usually involves sophisticated algorithms although clinical judgment can play an important role.

According to the National Institute for Health and Care Excellence (NICE) guideline NG77 all cataract surgeons should consider using a validated risk stratification algorithm to identify people at increased risk of complications during and after surgery (4). The results of the risk stratification should be explained to the patient at the time of informed consent and used to help the patient make decisions about their care (4).

Several algorithms have been developed to stratify risk in patients undergoing cataract surgery (6-9). The National Ophthalmology Database (NOD) risk stratification model, is based on a large retrospective multivariate regression analysis derived from previous National Cataract Audit data and is incorporated into the commonly used electronic medical record (EMR) programmes (MediSIGHT, OpenEyesTM). If the pre-operative risk data is recorded in the EMR, the patient will be risk stratified automatically and the information is used to plan the surgery (Scenario 1 & 2).

The majority of risk factors associated with both PCR and VA loss are non-modifiable such as the patient's age, axial length, and the presence of ocular co-morbidity (2,3). However, prior knowledge of risk factors helps to anticipate intraoperative technical challenges, and take appropriate precautions such as modifying surgical or anaesthetic techniques, using specialised equipment or timely onward referral to the relevant sub-specialty teams (2,10).

The key modifiable risk factor for PCR or VA loss is the level of surgeon experience. Risk stratification of patients helps to allocate cases based on surgeon experience (2, 12). The selection of appropriate cases for trainee surgeons combined with close supervision and the use of work-based assessments such as Objective Assessment of Surgical and Technical Skills (OSATS) promotes a safe step-wise system of surgical training whereby case complexity is increased as the trainee gains in surgical experience (12).

Patient A is 90 years old and presents with a white cataract, a small pupil and pre-existing macular degeneration.

During informed consent the higher risk of PCR and VA loss should be discussed, as well as the guarded visual prognosis due to the macular degeneration. The surgery should be done by a senior surgeon.

Surgical techniques to reduce the risk of complications such as Trypan Blue dye to stain the lens capsule and mechanical pupil stretching devices (Malyugin ring or iris hooks) to enlarge the pupil should be used. Close monitoring of the macular degeneration is warranted.

Scenario 1: Managing a patient with HIGH surgical risk.

Patient B is 68 years old and presents with difficulty driving caused by a moderate degree of cataract. Patient B has mild diabetic retinopathy and no other surgical risk factors.

During informed consent the higher risk of PCR and VA loss should be discussed in addition to the risk of progression of the diabetic retinopathy as a result of surgery. The surgery can be allocated to a trainee with the appropriate level of experience but must be performed under close supervision.

The diabetic retinopathy should be stabilised prior to surgery and monitored closely post-operatively along with the use of topical non-steroidal anti-inflammatory agents.

Scenario 2: Managing a patient with MODERATE surgical risk.

Diabetic retinopathy	Management of diabetic retinopathy Used of peri-operative non-steroidal anti-inflammatory eye drops
Small Pupil size	Intracameral phenylephrine Pupil stretch or sphincterotomy Viscoelastics such as Healon 5 Iris hooks Malyugin ring
White cataract	Pre-operative retinal B-scan ultrasound Tryphan blue dye
Inability to co-operate	Consider sedation or general anaesthetic
Weak zonules or damaged PC	Consider early referral to vitreoretinal team Capsular tension rings Alternative lens implants
Ocular co-morbidities	Cataract surgery performed by a team specialised in managing the specific co-morbidity Careful monitoring of co-morbidity
Trainee Surgeon	Appropriate case selection Close supervision Sufficient surgical time

Table 3: Minimising surgical risks

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Risk Adjustment

The case-mix data from the National Cataract Surgery audit showed that PCR rates ranged between 2.62% for those surgeons performing the most complex case-mix and 1.17% for those performing the least complex cases (13). This is particularly relevant when complication rates are audited against national benchmark figures and between individual surgeons (1).

The question is, who is the competent surgeon, Surgeon A, who reports higher rates of PCR or VA loss, but operates on a more complex case-mix than his/her peers or Surgeon B who has a less complex case mix and reports lower rates of PCR or VA loss? To help answer this question the NOD cataract audit applies a risk adjustment formula to produce risk-adjusted rates of PCR and VA loss for individual surgeons (1,14).

If Surgeon A's case-mix is of higher complexity then after adjusting for risk the PCR and VA loss rates will be reduced. If, however, Surgeon A's rates of PCR or VA loss remain above the benchmark rates despite risk-adjustment, it would signify that Surgeon A has a 'worse than expected' surgical outcome. On the other hand, surgeon B may also have a 'worse than expected' surgical outcome if his case-mix was of very low complexity and after adjusting for the lower risk the rates were higher than the benchmark rates.

The current benchmark rates for Consultant surgeons are 1.1% for PCR and 0.9% for VA loss (1) The accuracy of the risk-adjusted rates of PCR and VA loss is dependent on surgeons accurately recording the relevant risk factors for all their surgical cases.

Conclusion

The two benchmarked indicators of surgical quality, PCR and VA loss, are associated with known risk factors and early recognition will significantly decrease the chance of PCR or VA loss occurring, separately or concurrently.

Risk assessment and stratification aids the surgeon to take preventative measures to minimise complications. It allows appropriate allocation of cataract cases according surgeon experience and promotes a safer step-wise approach to surgical training. It also has the benefit of helping patients to make informed choices about their surgery. Risk adjustment allows individual surgical outcomes to be audited and compared to the National benchmarked rates as well as between peers. It is evident that accurate and meticulous pre-operative risk assessment will determine how good a cataract surgeon you are and can be.

MCQ

1. It is important to carry out a detailed assessment of pre-operative risks in patients undergoing cataract surgery because

A. It can improve surgical outcomes

B. It helps plan the surgical procedure and mitigate risks

C. It helps select appropriate cases for trainee surgeons

D. All of the above

E. All except C

2. The most important modifiable risk factor for posterior capsule rupture (PCR) and visual acuity (VA) loss is

A. Small pupil size

B. Patient unable to lie still

C. Inexperienced surgeon

D. Diabetic retinopathy

E. None of the above

3. The National Ophthalmology Database (NOD)

A. was established by the Royal College of Ophthalmologists

B. are responsible for collecting and analysing data for the National cataract audit

C. benchmark the indicators of surgical quality for cataract operations in England and Wales

D. report risk-adjusted rates for individual surgeons

E. All of the above

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4. The current benchmark rate for posterior capsule rupture following cataract surgery for a Consultant Eye Surgeon is

A. 5.1%

B. 2%

C. 1.1%

D. 0.9%

E. None of the above

5. Risk adjusting the rates of PCR and VA loss associated with cataract surgery is important because it

A. helps to recognise differences in case-mix complexity and identify acceptable variations in surgical outcomes

B. lowers rates of PCR and VA loss and improves surgical outcomes

C. increases rates of PCR and VA loss in complex cases

D. None of the above

E. All of the above

Answers

1. Answer: D

Pre-operative risk assessment and stratification will improve surgical outcomes by reducing the risk of posterior capsule rupture and visual loss.

The surgeon can plan the procedure according to the risks and this may include modifying a surgical technique or using specialised equipment. Less complex cases are usually reserved for the junior trainee surgeons (see Table 3 and Scenarios 1,2).

2. Answer: C

The National Cataract Audit showed that the risk of both PCR and VA loss was much higher with a junior trainee surgeon with less experience. It was recommended that risk stratification of cataract patients be used to select cases appropriate for trainees to minimise the risk of complications and help in providing a safer step-wise approach to surgical training.

3. Answer: E

Cataract surgery is the most frequently undertaken NHS surgical procedure with approximately 390,000 cataract operations undertaken in England and 16,000 in Wales during 2015-2016.

The Health Quality Improvement Partnership (HQIP) commissioned the National Ophthalmology Database (NOD) Cataract Audit to report on all NHS funded cataract surgery in England and Wales (1).

4. Answer: C

The NOD audit for 2016-17 has re-set the benchmark rate for PCR and VA loss after cataract surgery performed by a Consultant Eye Surgeon to 1.1% and 0.9% respectively. The previous benchmark rates were 2% and 1.5% respectively and had been derived from pooled data for surgeons of all grades.

5. Answer: A

The risk adjustment formula used by NOD is shown below. It helps recognise differences in case-mix complexity for individual surgeons.

Risk-adjusted rate = Benchmark rate x (Observed rate/ Expected rate)

Ideally, the ratio of the observed to the expected rate should be the equal to 1, the observed rate being the reported figure for a individual surgeon and the expected rate the figure that would be expected in a population with the same level of complexity as the surgeon.

If the observed rate is higher than the expected rate (ratio >1) then the risk-adjusted rate would be higher than the benchmark while if the observed rate is lower than the expected rate (ratio <1) then the risk-adjusted rate would be lower than the benchmark.

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ETHICAL CONSIDERATIONS WHILE LISTING FOR CATARACT SURGERY

Y Levene, Z Cheng, J Shankar

Abstract

This case-based discussion of an elderly lady with cataract and Fuchs' endothelial dystrophy describes the four pillars of medical ethics which allow us to provide 'Good Medical Practice' as highlighted by the GMC. It discusses how these are pertinent to our case and day to day in the ophthalmology department.

Case history

An 81-year-old lady was referred to our ophthalmology outpatient department routinely by her optician for consideration of cataract surgery.

She currently lives alone, independently and manages fairly well. However, she has noticed she is struggling to read and is concerned about things deteriorating and losing her independence. She has had no previous ocular history though says she remembers it being mentioned at the optician she has 'pearly metal' colour to her eyes.

Past medical history includes diet-controlled type 2 diabetes and osteoarthritis. On examination her left eye was an amblyopic eye with a visual acuity of 6/36, her right eye visual acuity of 6/18. She had bilateral Fuchs' endothelial dystrophy and both eyes had posterior subcapsular cataract. Intraocular pressures were 14mmHg bilaterally and fundal examination bilaterally was normal.

There was extensive discussion surrounding listing her for cataract surgery given her other eye being amblyopic, then the added ocular pathology of Fuchs' endothelial dystrophy adding risk to the surgery and that currently she feels herself she is managing well. Her daughter attended with her and it was decided they would go home with some further information to think about things. She has a follow up appointment in 3 months' time.

This case highlights some of the ethical considerations for ocular surgery which will be further discussed.

Discussion

The four pillars of medical ethics are of daily importance in the ophthalmology department. They are autonomy, beneficence, non-maleficence and justice (1).

Pillar of medical ethics	Definitions as per Medical Protection Society (1)
Autonomy	Respect for the patient's right to self determination
Beneficence	Aiming to help patients
Non-maleficence	To do no harm
Justice	Treating patients equally and equitably

Table 1: Definitions of four pillars of medical ethics, from Medical Protection Society (1)

Each principle is important and should be considered when making decisions about patients care in order to comply with Good Medical Practice (2).

Autonomy

In terms of our patient's autonomy in this case, her main aim was to continue independent living. She was unduly concerned about the risks given her other eye being amblyopic and the diagnosis of Fuchs' endothelial dystrophy and wished to have longer to make her decision, as is her right.

She was provided with information about her condition and the risks posed with surgery. Firstly, as Fuchs is characterised by loss of endothelial cells there are greater intra-operative risks due to the poor view (3). Secondly there is a post-operative risk of worsening Fuchs owing to further endothelial loss during surgery (3). This could negate some of the vision that would be gained by removing the cataract.

In this case there are clear arguments for both having and not having the surgery and as always, the decision of whether to go ahead with the surgery if offered, is the patients.

What is important to note is that to enable patients to make autonomous decisions we must provide appropriate information and allows them to make an informed decision, taking into account their life values. This may lead to a patient making what a clinician would see as an 'unwise' or illogical decision but this is their right (4).

There have been patients who cannot bear the idea of having their eyes operated on and so despite having significant cataracts that would likely gain a real improvement in their vision from surgical intervention, have decided against having surgery.

Many patients with cataracts also have other ocular pathology such as age-related macular degeneration (AMD)). In this case the information patients are provided is with a guarded prognosis, that the vision may improve but other pathology means it will not be fully regained or cure the other pathology (5). In this case some patients are happy to go ahead as they feel that any increase in vision is very important to them while others may feel they do not wish to take risks or undergo surgery if it will not fully resolve all of their visual problems.

Beneficence and non-maleficence

Just like the blades Ophthalmologists hold, the intervention we provide is a "double- edged" sword. We as doctors have a moral obligation to help our patients, however the treatment we offer will inevitably risk harming them (6). We must consider the two principles jointly to bring the most benefit (beneficence) without causing undue harm (non-maleficence) to the patient.

In order to achieve this, we must have sufficient knowledge and experience of the disease to profess (hence professional) the benefit and risk of harm we can bring. As foundation doctors, it is more important to recognise the limits of our expertise in the field and involve a more experienced team member early on in the case.

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One of the ways we can practice beneficence is by acting as an advocate for your patient, as highlighted by a GMC report (2). This involves promoting the interests of the patient, ensuring the patient access the right expertise and representing the view of the patient in discussions with other team members.

Another way we practice beneficence is by ensuring effective communication with both our patients and our colleagues. It is essential for the right information to be delivered in an appropriate manner while listening to and addressing any patient concerns appropriately (7). By ensuring the patient has understood the issues around the decision they have made, we have also enabled them to make an autonomous decision.

One of the roles of a foundation doctor is to protect patients, by being the "guardian" (6). This involves ensuring that safety guidance is adhered to and practicing within your level of competence of knowledge and skills thereby incorporating the principle of non-maleficence by trying to avoid harm to the patient.

More generally in ophthalmology this is achieved by identifying potential safety risks or difficulties that could happen during the individual's surgery and try to take steps to alleviate this. It is also encompassed in the 'Duty of Candour' in 'Good Medical Practice', being open and honest with patients about mistakes made (2).

In our case, our patient was seen by a Foundation Year 2 doctor initially. By acting as an advocate, the patient was discussed with, and reviewed by the Specialist Trainee and subsequently the attending Consultant.

The Consultant had sufficient knowledge and experience to explain to the patient the options of treatment and overall benefit and risks of surgery. The Consultant and Specialist Trainee acted as communicators to ensure the patient received relevant information and enabled the patient to make an autonomous, informed decision.

This can also be seen in patients with AMD that are offered and undergo cataract surgery. By informing them of the latest literature and offering cataract surgery, we maintain the principle of beneficence. Patients with AMD and cataracts report improved visual acuity and contrast sensitivity after cataract surgery (5).

Therefore, in order to be maintaining these ethical principles, people with existing ocular pathology should still be considered for cataract surgery if there is opportunity to improve their vision in some way.

In summary, we can ensure we practice beneficence and non-maleficence by:

- *Acting as advocates*
- *Acting as guardians*
- *Acting as communicators*

Justice

Looking at distributive justice in healthcare is about both "equity and equality" (Medical Protection Society) (1). Equality tries to ensure that everyone is given the same things to enjoy a healthy life, but some people will inherently need more resources than others- this is equity, giving individuals what they need to maintain a healthy life (6). The NHS does not have unlimited resources and therefore in trying to deliver true equity we may face challenges (4).

The waiting list time for cataract surgery varies from hospital to hospital. There are often phone calls from patients and letters from opticians to expedite surgery. It is therefore important when listing patients to evaluate the extent to which vision is compromised and having an effect on daily activities, work or ability to maintain an independent life and list urgency as appropriate.

In our case our patient was amblyopic in one eye which would generally mean an increased urgency if the patient had only one good eye, however our patient felt she was coping well so this would not necessitate expediting surgery.

Distributive justice can also be seen when looking at clinician time, in general it is agreed that the doctor should spend sufficient time with patients that is needed for them to take a history, examine and make a management plan that is communicated to the patient (1). Some patients will take up more clinician time due to difficult clinical scenarios that require more discussion such as with our patient and this can generally be ethically justified (4).

However, there are instances where patients who have complained previously receive more attention and time than other patients, not based on clinical need but on trying to avoid another complaint. While this may be understandable it may be considered to go against the principle of justice.

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Balancing the Ethical Pillars

Each of the ethical pillars are well defined and each can be applied to clinical practice individually. The real challenge is in scenarios where these principles conflict. It is argued that these principles are "prima facie", meaning they are all binding unless they conflict with each other, in which case we must choose one over another. We as doctors need to balance these principles in practice. For example:

1. A patient exercises his/her autonomy to refuse a treatment that you know will bring an overall benefit(beneficence).

2. You wish to advocate for your patient (beneficence) to have an expedited cataract surgery, but this may delay other patients who have been on the waiting list (justice).

3. A patient failed to attend eye clinic (autonomy) on multiple occasions, wasting valuable consultation sessions for other patients (justice), but not offering new appointments may cause harm (non-maleficence).

These are just some day to day examples where the ethical principles play conflicting roles in decisions doctors make. In such situations it is often helpful to discuss with colleagues and come to an appropriate well-reasoned decision.

It is commonly accepted, which also applied in our case, that patient autonomy prevails above beneficence (3). Despite the fact our patient was functionally affected by her cataract, she felt she would have greater benefit from monitoring her current cataract than risk worsening eyesight, and the team respected her view.

Conclusions

• Ophthalmologists are bound by the four pillars of medical ethics and these should be considered when seeing patients in the eye department and considering listing for surgery.

• Providing appropriate information for patients allows them to make autonomous decision.

• There may be times when ethical principles conflict, discussion with colleagues is often helpful.

Test yourself

1) Autonomy can best be described as

a) Acting in order to cause the least harm to your patient

b) Acting to try and benefit the patient

c) Allowing your patient to make decision's based on their life values

d) Not allowing your patient to make unwise decisions

e) Trying to use resources fairly

2) With regards to pre-operative cataract considerations

a) AMD should be considered an absolute contraindication to cataract surgery

b) Amblyopic patients should not be listed for surgery

c) People who ask to for surgery to be expeditated should always be listed as urgent

d) Anyone with other ocular pathology should be fully discussed and where appropriate listed with a guarded prognosis

e) Patient's opinion on how their cataract is affecting their lives is secondary to their visual acuity

Answers

1 - C, this is a definition of autonomy.

This question is concerned with the four pillars of medical ethics. A is describing non-maleficence, B is describing beneficence. D is not true; an autonomous decision does not have be a wise decision as long as the decision fits with the patient's life values. E is describing justice

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2 -D

This is good medical practice and is needed for appropriate informed consent for the surgery. It should be explained how the other ocular pathology can influence both surgery and outcomes.

A, this is not the case, as discussed AMD patients have been shown to derive benefit from cataract surgery but it must be made clear that surgery will not fix their distortion or central visual loss associated with AMD.

B, amblyopic patients arguably should be considered for surgery if they have a potentially reversible condition affecting their good eye.

C, listing urgency should be based on clinical need not at patient request, principle of justice.

D, visual acuity is always secondary to the effect of vision on patient's daily activities though they will often go hand in hand.

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PRE-SEPTAL & ORBITAL CELLULITIS

Y Levene, M Abdimalik, J Shankar

Abstract

This case-based discussion of an immunocompromised, 57-year-old male presenting with septic shock secondary to pre-septal cellulitis outlines the presentation, diagnosis and management of such patients. There are contrasting points in presentation between pre-septal and orbital cellulitis.

Examination of the former reveals normal vison and pupils accompanied with unrestricted eye movements due to the infection not anatomically breeching beyond the tarsal plate. Other differences in presentation and management are reviewed.

Specific learning points for foundation doctors are made throughout, identifying key referral points and a multitude of specialties to involve in patient care – Ophthalmology, ENT, Maxillofacial and ITU.

Case History

A 57-year-old male presented to A and E with unilateral, extensive soft tissue oedema and erythema surrounding his left eye.

History of presenting complaint revealed that the patient had noticed some minor swelling the previous evening, which upon waking had worsened to an extent that he was unable to see out of that eye due to the restrictive effects of the oedema. This was accompanied by intense pain.

Upon reflection of the past days events he recalled sustaining a scratch above his left eye by a tree branch whilst gardening. He continued to worsen clinically becoming pyrexic. He denied upper respiratory tract symptoms.

Past medical history: *Chronic renal failure with failing renal transplant.*

Medications: *Azathioprine, Tacrolimus. No known drug allergies identified.*

Examination: *Unable to part the lids to open the eye or even assess for consideration of performing a lateral canthotomy by A and E. Ophthalmology assessment below.*

Vital signs were:

- *Temp 38.2° C.*
- *Heart rate 124 beats/minute.*
- *Blood Pressure 90/68 mmHg.*
- *Respiratory Rate 20 breaths/minute*

A diagnosis of sepsis from pre-septal cellulitis was made and the appropriate protocol initiated. This involved completing 'The sepsis six' - blood cultures and ABG with lactate taken, urine output monitoring, IV antibiotics within the hour (cefuroxime and metronidazole commenced), IV fluids and oxygen administered. Ophthalmology were contacted for referral immediately and subsequently ITU and ENT.

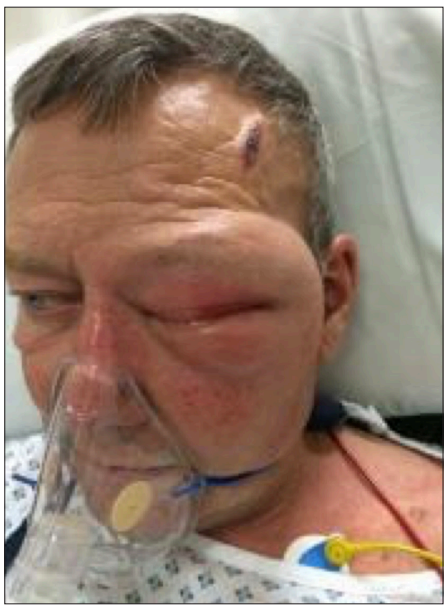
Ophthalmology examination at initial presentation showed a bilateral visual acuity of 6/9 (no change with pinhole), with normal, painless eye movements and pupil reactions. There was no relative afferent pupillary defect present. Patient had full colour vision with Ishihara plates (14/14).

Gross swelling of the upper and lower eyelids and soft tissue surrounding the left eye were noted with pre-auricular lymphadenopathy. All assessments discussed were facilitated by a nurse parting the eyelids due to the surrounding swelling. Furthermore, due to the severe sepsis, slit lamp examination was not feasible or appropriate. Fundoscopy at the bedside revealed a brief glimpse of a normal optic disc.

Blood tests: *He was admitted to ITU with septic shock requiring level 3 care in the form of intubation and ionotropic support to maintain blood pressure.*

Blood test	Result	Normal reference range
Na	136	(135-145) mmol/L
K	4.8	(3.5-5.0) mmol/L
Hb	75	(130-170) g/dL
Platelets	49	(150-400) x 10^9/L
WBC	13.7	(4.0-10.0) x 10^9/L
Neutrophils	11.1	(2.0-8.0) x 10^9/L
CRP	270	(<5) mg/L
Urea	13.1	(1.2-3.0) mmol/L
Creatinine	233	(80-110) mg/dL
Lactate	1.9	(<2) mmol/L
Blood culture	No Growth	

Urgent CT orbit showed small peripherally enhancing collections (post contrast) seen in the anteroinferior aspect of the left orbit, with small extension towards the insertion of left lateral rectus. Marked soft tissue swelling overlying the left orbit, maxillary and temporal bones was apparent however the orbital septa remained unbreached.



Picture 1: Initial presentation when seen by ophthalmology ST1 doctor (with patient consent to have photograph taken and published)

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Outcome

He remained in ITU for 10 days with daily ophthalmology review to check the swelling, eye movements, pupil reaction and cornea. From an ophthalmological point of view, he responded well to treatment with reduced swelling by 24 hours of IV antibiotics. Unfortunately, his kidney function continued to decline, and he has been on dialysis since and is awaiting a further renal transplant.

This was partly attributed to the contrast needed for the CT scans but it was decided and discussed with all the teams and the patient that the CT scans were vital investigations and his renal team felt he would be needing dialysis soon anyway.

Why is this important to me as a foundation doctor?

A recent study showed 72% of doctors contacted initially for patients with pre-septal cellulitis were foundation doctors or core trainees. Moreover, not all hospitals have comprehensive ophthalmological guidelines available for junior doctors to refer to. Therefore, having some prior knowledge of the presentation and initial management is useful. Early referral for specialist advice to ophthalmology is essential (1).

When referring to ophthalmology use a systematic approach as you would with any referral. SBAR (situation, background, assessment and your recommendation) is a useful tool to use to structure your conversation to ensure both the clarity of information you are giving as well as what you are asking for in return. For example, your recommendation could be that you require telephone advice, management advice or a specialist review.

Ophthalmology will want to know, based on your history and examination, whether you think it is likely to be pre-septal or orbital cellulitis, in particular how far the swelling and erythema extend, if there are reduced eye movements (ophthalmoplegia) or obvious abnormal protrusion of the eye (proptosis) and if the patient is likely to require admission or is septic.

It is important to document visual acuity and pupil reactions (including any relative afferent pupillary defect (RAPD)) at presentation in order to monitor response to treatment. If you feel unable to assess RAPD ask a senior or ophthalmology doctor to do so.

Recording visual acuity and pupil reactions is also good practice for legal reasons; if invasive ophthalmological procedures are to be performed (such as lateral canthotomy to reduce orbital pressure) it is prudent to know the ophthalmological state of the eye pre and post procedure.

Anatomy: *The orbital septum is a membranous sheet that originates from orbital bone margins and fuses centrally with the tarsal plates. The sheet acts as a barrier that separates eyelid fat from orbital fat and the orbicularis oculi muscle preventing the spread of pathogens (2).*

Pre-Septal cellulitis (Peri-orbital cellulitis)

This is an infection that is anterior to orbital septum with symptoms generally limited to swelling, warmth and tenderness of eyelids. Patients often present with fever, malaise and irritability particularly in children.

Ptosis can also occur. These symptoms alone are suggestive of local infection that have not passed beyond the orbital septum (3). Therefore, patients with pre-septal cellulitis have normal vision and pupils with unrestricted ocular movements.

This primarily affects young patients with signs limited anteriorly in the eyelid and surrounding soft tissue (4). The orbital septum is not fully developed in young children. This increases risk of progression of infection significantly (5). Additionally, infection can progress to cavernous sinus leading to thrombosis and intracranial infection (6).

As with all infections and the case above, be aware of atypical and serious presentations in immune-compromised patients.

Causes: *Infection with staphylococcus and streptococcal species is the most common but infection with Haemophilus influenza can still occur. Beta-hemolytic streptococcus is particularly associated with facial trauma, oral and sinus surgery (7).*

Pre-Septal cellulitis occurs following spread from local infection as follow:

- *Sinusitis*
- *Upper respiratory tract infections*
- *Trauma around the eye (laceration)*
- *Dental abscess*
- *Local impetigo*
- *Insect bite*
- *Recent surgery around the eye*
- *Endogenous spread in immunocompromised patients*

Symptoms:

- *Fever and malaise.*
- *Normal white sclera.*
- *Erythema around the eye.*
- *Tenderness and oedema of the eyelid and*
- *Swelling and induration (hardening of soft tissue).*
- *Inflamed lids but no proptosis.*

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Investigations
If uncomplicated further investigation may not be required but ENT opinion should always be sought if spread from sinuses is suspected.

Immune compromised patients require sepsis screening. Additionally, there should be a low threshold for CT orbit to check for orbital cellulitis. This can detect spread beyond the septum early on and avoid any serious complication.

Management
Uncomplicated pre-septal can be treated with oral flucloxacillin with regular check-ups by ophthalmology to check for improvement and monitor visual acuity.

If complicated (immune compromised, septic or severe) admission and IV antibiotics will be required, as in the case above.

Orbital Cellulitis

Posterior spread can lead to orbital cellulitis. This is a serious infection that can result in loss of sight with significant life-threatening complications. Visual symptoms are the most frequent complaint and as such ophthalmology review should be sought.

Such symptoms are indicative of orbital cellulitis (infection in the eye) with offending pathogen spreading beyond the orbital septum (8). It presents as follows:

Signs and symptoms:

- *Proptosis (eyes bulging anteriorly).*
- *Painful or limited ocular movements (ophthalmoplegia).*
- *Reduced visual acuity.*

Investigations:

- *Sepsis screen bloods: FBC, U&E, CRP, Lactate and ABG.*
- *Blood culture.*
- *Percutaneous aspirates.*
- *CT orbit if posterior spread suspected.*
- *Lumbar puncture if meningitis suspected.*

Patients can present septic with orbital cellulitis and SEPSIS SIX should be followed within an hour of arriving at hospital:

Take three: *Blood culture, Urine output, Lactate*

Give three: *IV Antibiotics, IV fluids and Oxygen*

ENT opinion needs be sought urgently and consider involving maxillofacial and ITU early in the case

Management
Intravenous antibiotics are effective in only 62% of cases. The rest may require paranasal sinus or orbital surgery. Antibiotic therapy should allow cover for both streptococcal and h. influenza as per local protocol (9). If in doubt always discuss with microbiology who will advise on the most appropriate antibiotics.

Further Complications (10)

Meningitis.

Encephalitis and cerebral abscess.

Sepsis.

Key messages

- *Sepsis is possible from pre-orbital and orbital cellulitis*
- *Pre-orbital cellulitis does not present*

with visual problems but orbital cellulitis does

- *Involve ophthalmology early on in the case*
- *Always document visual acuity at initial presentation*

and before any procedures such as lateral canthotomy

Questions

1) What is the gold standard imaging for orbital cellulitis?

A) X-ray facial bones
B) B-scan ultrasound
C) CT orbit
D) OCT macula
E) Fundus photography

2) Which set of signs/symptoms would be most indicative for orbital cellulitis?

A) Proptosis, ophthalmoplegia, eyelid swelling
B) Erythema, eyelid swelling
C) Eyelid oedema, conjunctival congestion and injection
D) Eyelid inflammation, discrete nodule, insidious onset
E) Photophobia, reduced visual acuity, red eye

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Answers

1) C, this is the gold standard imaging to evaluate and diagnose orbital cellulitis and can demonstrate oedema, thickened sclera, proptosis and sinusitis (11).

A is useful initially when consider fractures including inferior wall 'blow out' fractures, though CT orbit is still gold standard for this.

B is readily available in ophthalmology departments and remains a useful tool particularly for evaluating retinal and choroidal abnormalities such as retinal detachments or choroidal nevus.

D is used for assessing conditions such as age related macular degeneration and diabetic maculopathy. E is used to take fundus photographs where a fundal abnormality is detected and used to monitor progression of such abnormalities.

2) A, these signs are highly indicative of orbital cellulitis, along with changes to pupil reaction including RAPD, reduced visual acuity, other signs of sepsis including fever and tachycardia.

B and C are more indicative of pre-septal cellulitis or conjunctivitis.

D is more likely to be a chalazion or cyst anE is most likely to be anterior uveitis.

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SMALL EYE BIG PROBLEM

LW Khoo, S Srinivasan

Abstract

This case report is on a 65 year-old hypermetropic gentleman who presented with acute angle closure glaucoma. In this report we discuss the clinical presentation, diagnosis, management of this patient and discuss the etiology, risk factors and management of acute angle closure glaucoma.

Case History

A 65-year-old man was referred to the eye casualty with a history of sudden right eye pain and blurry vision along with right sided headache. Six hours prior to the eye casualty visit, he attended his optician for a routine eye exam during which the optician installed mydriatic eye drops (Phenylepherine 2.5 % and Tropicamide 1 %) to dilate the pupils for a posterior segment examination.

Past ocular history was significant for hypermetropia and his spectacle prescription was +7.50/+0.50 x 175 in the right eye (RE) and +7.00/+0.50 x 25 in the left eye. Visual acuities were 6/12 with pinhole for the RE and 6/4.8 for the left eye (LE).

On slit lamp examination, the RE showed corneal oedema, shallow anterior chamber and a fixed semi- dilated pupil. The intraocular pressure (IOP) measurements were 77 mm Hg in the RE and 16 mm Hg in the LE (normal range 10 - 21 mmHg). Gonioscopy of the RE was difficult due to the corneal edema. Gonioscopy of the LE showed grade one narrow angles 360 degrees.

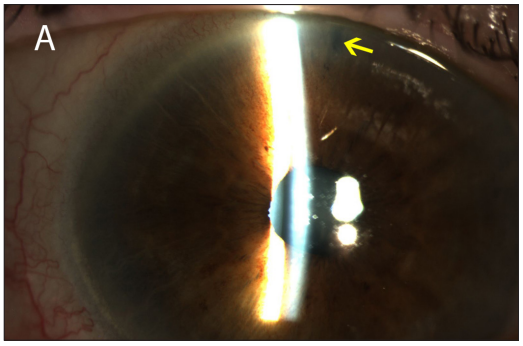
The patient was diagnosed with acute angle closure glaucoma in the RE (secondary to the installation of mydriatic agents). He was admitted and treatment was immediately commenced with intravenous acetazolamide 500 mg along with topical IOP reducing agents (Dorzolamide/Timolol, Pilocarpine, Bimatoprost and Apraclonidine)

However, as the IOP remained elevated at 60mmHg two hours after the above treatment, 250mg of intravenous Mannitol and oral acetazolamide 250 mg tid were added to the treatment regimen. Pilocarpine eye drops 2 % tid was also prescribed to the LE to constrict the pupil.

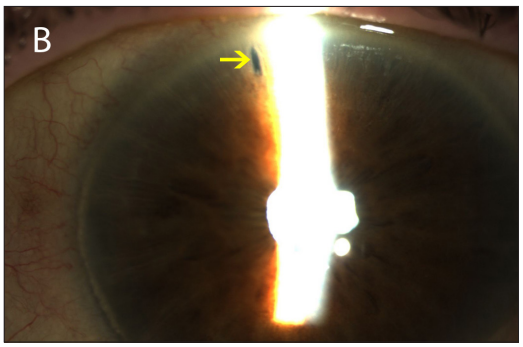
Several hours after commencement of IV Mannitol, the patient developed severe back pain. Upon further questioning, the patient revealed a past medical history of benign prostate hypertrophy and a diagnosis of urinary retention was made. An urgent ultrasound scan of the abdomen revealed an enlarged bladder.

Urinary catheter was inserted by the on call urologist, which resulted in immediate relief of his urinary symptoms. Following intravenous mannitol, the IOP in the RE reduced to 24 mm Hg. The following day, the patient underwent YAG laser peripheral iridotomies.

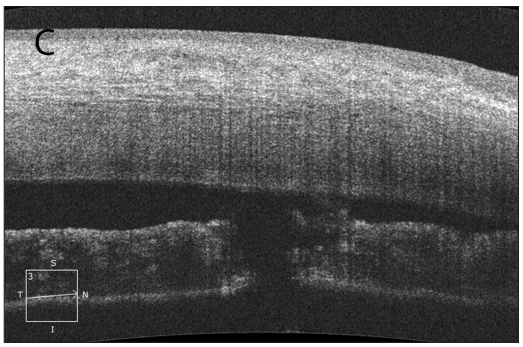
Figure 1



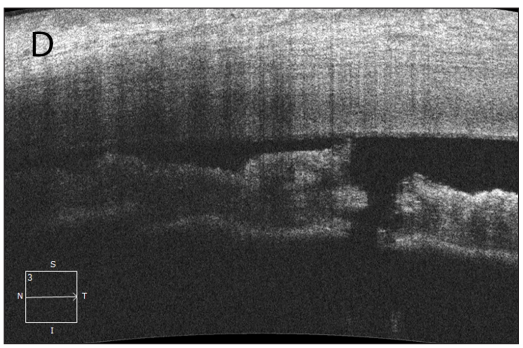
A: Peripheral iridotomy (arrow) in RE



B: Peripheral iridotomy (arrow) in LE



C: Anterior segment optical coherence tomography demonstrating patent peripheral iridotomy in RE



D: Anterior segment optical coherence tomography demonstrating patent peripheral iridotomy in LE

SMALL EYE BIG PROBLEM

LW Khoo, S Srinivasan

Discussion

Acute angle closure glaucoma (AACG) is an ophthalmic emergency that could lead to blindness if left untreated. In AACG, there is acute elevation of IOP due to the occlusion of the trabecular meshwork by the peripheral iris (1).

Common symptoms of AACG include blurry vision accompanied by haloes, periocular pain and headache. In addition to IOP of >30mmHg, slit lamp examination would also reveal signs such as conjunctival redness, fixed mid-dilated pupil and corneal oedema (1).

Several risk factors have been identified for development of AACG. Older age is a known risk factor with the average age of patients developing AACG being 60 years and over. Females and individuals of the Far eastern and Indian Asians ethnicity are also more prone to this condition.

A family history of AACG further predisposes one to AACG. Hypermetropic patients are also more likely to develop AACG (1). Other precipitating factors such as watching television in a dim room, iatrogenically induced mydriasis, adoption of semiprone position and acute emotional stress have also been identified.

In our patient's case, being >60 years old and hypermetropic had already placed him at high risk of developing AACG. The dilated fundoscopy examination with his optician tipped the scales and led to this AACG attack.

As high IOP could lead to irreversible damage to the optic nerve, the main aim of management of IOP is to reduce IOP with medical treatments such as systemic or topical carbonic anhydrase inhibitor, topical beta-blockers, topical prostaglandin derivatives (Bimatoprost), topical alpha-2 agonists (Apraclonidine), miotics (Pilocarpine) and osmotic agents (Mannitol).

Osmotic agents such as Mannitol are employed in resistant AACG not responding to other topical treatments. It achieves short term reduction in IOP by creating an osmotic gradient that draws water out from the vitreous into the blood.

However, it could lead to side effects such as cardiovascular overload and urinary retention. Hence, it needs to be used in caution in patients with cardiac, renal or prostate disease (1). In our patient, although Mannitol had successfully reduced the IOP, he developed urinary retention secondary to it.

Once the acute attack of AACG is under control, peripheral iridotomy is usually performed to allow an alternative route for the flow of aqueous humour, allowing normalization of the pressure gradient between the anterior and posterior chamber.

In the emergency department without equipment or skills for IOP measurement, diagnosis of AACG can sometimes be delayed and misdiagnosed as migraine. Migraine can present with similar symptoms to AACG including blurry vision, conjunctival redness, periocular pain and headache.

The key to differentiating AACG from migraine in such a situation is in the examination of pupils. In AACG, the pupil is fixed and mid-dilated while in migraine, the pupils are reactive to light. It is crucial to diagnosis and manage AACG in a timely manner as delayed diagnosis could lead to irreversible optic nerve damage and blindness.

MCQs

1. The following are risk factors for development of AACG except:

A. Myopia

B. Family history of AACG

C. Old age

D. Female

E. Asian ethnicity

SMALL EYE BIG PROBLEM

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2. Mannitol should be used with caution in patients with the following medical condition:

A. Hypertension

B. Heart failure

C. Diabetes

D. Rheumatoid Arthritis

E. Eczema

3. The following medications could be used to manage AACG except:

A. Beta-blockers

B. Prostaglandin analogues

C. Carbonic anhydrase inhibitors

D. Alpha-blockers

E. Miotics

Answers

1. A. Myopia

Hypermetropia is a risk factor for development of AACG. Individuals with hypermetropia have smaller eyes that are shorter in length. The intraocular structures are spaced closer together, predisposing the individual to shallow anterior chambers and narrow occludable angles.

2. B. Heart failure

Mannitol is an osmotic diuretic. It should be used with caution in individuals with heart failure due to the increased risk of pulmonary oedema.

3. D. Alpha-blockers

Alpha agonists instead of alpha-blockers are used to manage AACG. Alpha agonist decreases aqueous humour production and reduces resistance to aqueous outflow.

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THE RED EYE & SYSTEMIC INFECTION

AJ Simpson, A Krishnakumar, HDJ Hogg, MP Clarke

Abstract

Endophthalmitis is intraocular infection and inflammation resulting in devastating visual consequences. It is most commonly exogenous where infection is introduced following intraocular procedures or trauma.

However a small proportion of cases are endogenous, where infection is spread from other areas of the body and this carries a high mortality rate. Early recognition of endophthalmitis and urgent referral to ophthalmology can help save sight and lives.

Case History

A 70 year old lady was referred by her GP to the eye emergency department with a left sided red eye and monocular vision loss. She complained of having had flashing lights and a strange colour in her vision the previous day as well as pain and photophobia.

The morning of presentation, she woke up being unable to see out of the left eye. The visual acuity in the right eye was 6/9 and in the affected left eye she was only able to count fingers. On slit lamp examination, there was left eye conjunctival chemosis, corneal oedema and a 2mm hypopyon with anterior chamber fibrin. Due to the pronounced ocular inflammation it was not possible to gain a fundal view in the left eye.

To visualise the posterior segment of the globe, an ultrasound B scan of the eye was carried out which revealed debris floating in the vitreous. Our patient had no significant past ocular history and no previous ocular surgery. Her past medical history included recurrent biliary tree infections and back pain that had progressed over recent weeks to reduce her mobilisation.

As acute ophthalmic management, a vitreous biopsy was taken and intravitreal ceftazidine and vancomycin were given along with empirical systemic ciprofloxacin for a diagnosis of endogeneous endophthalmitis. Frequent steroid and mydriatic eye drops were commenced to reduce the intraocular inflammation and to reduce the patient's photophobia.

The vitreous biopsy grew streptococcus pneumoniae and under the guidance of the microbiology team, the patient's antibiotics were changed to intravenous meropenem and linezolid. A referral was made to the infectious diseases team to further investigate for an underlying systemic infection.

Magnetic resonance imaging of the spine was carried out to investigate our patient's back pain and she was found to have a large psoas abscess which had spread from infection of the obstructed biliary tree.

The psoas abscess was drained under radiological guidance and her back pain improved as did her ability to mobilise. The biliary tree was drained percutaneously and the patient is currently receiving long-term antibiotics under the supervision of the infectious diseases team. Most recently the patient's visual acuity had deteriorated to light perception only in the left eye and vitrectomy is under consideration.

Exogenous endophthalmitis

Endophthalmitis is infection and inflammation affecting the inner layers of the eye. Endogenous endophthalmitis as described in the case above, where infection in the eye arises from elsewhere in the body, accounts for around 5% of endophthalmitis. (1)

Exogenous endophthalmitis, where infection is introduced into the eye either through intraocular surgery or trauma, makes up the remainder and so it is important for primary care practitioners to recognise it in order to make emergency ophthalmic referral and minimise the impact to vision.

Exogenous endophthalmitis is likely to present with a unilateral red and painful eye with reduced vision. The key element in the history is the possibility of penetrating eye trauma or recent intraocular surgery or injection.

Although endophthalmitis rates are less than 1 in 1000 for both cataract surgery and intravitreal anti-vascular endothelial growth factor injection these are the commonest causes of endophthalmitis as hundreds of thousands of these procedures are performed each year in the UK. (2,3)

Without experience in slit-lamp biomicroscopy identifying the signs of endophthalmitis may be challenging. However, on close unaided examination conjunctival injection may be noticed along with hypopyon, recognised as a flat fluid level of inflammatory cells overlying the lower margin of the iris.

This hypopyon can progress rapidly in exogenous endophthalmitis, with the level rising notably over the course of several hours.

THE RED EYE & SYSTEMIC INFECTION

AJ Simpson, A Krishnakumar, HDJ Hogg, MP Clarke

The commonest pathogens causing exogenous endophthalmitis are coagulase-negative Staphylococci such as Staphylococcus epidermidis and Staphylococcus saprophyticus. (1) Endophthalmitis following intravitreal injection is 3 times more likely to be streptococcal which is thought to contribute to lower average visual outcome observed in post intravitreal injection endophthalmitis. (1)

Initial management varies between units but begins with an emergency vitreous tap, to establish the infecting organism and antibiotic sensitivities, followed by intravitreal antibiotic injection. (4) Pars plana vitrectomy is usually performed to reduce the bacterial load and clear the visual axis.

Endogenous endophthalmitis

In endogenous endophthalmitis, infection enters the eye following haematogenous spread of extraocular infection. Endogenous endophthalmitis is a sight-threatening ophthalmic emergency and has a high mortality rate related to disseminated infection if not identified and treated appropriately. (5)

The threat to sight and life emphasises the importance of awareness of this condition to clinicians. Patient groups who are at risk include those who are diabetic, immunocompromised, intravenous drug users and those with indwelling catheters or urinary tract infections. (6)

In the Western world, bacterial endogenous endophthalmitis is most commonly due to gram-positive bacteria such as Staphylococci. (7) This is in contrast to Asian countries where gram-negative species are more common. (8) Endogenous endophthalmitis can also be caused by fungal infections, such as Candida and Aspergillus species. (9)

Endogenous endophthalmitis is not linked to ocular surgery or trauma and will present with a red eye and painful visual loss. There will likely be photophobia and floaters. Note that these patients may be systemically unwell with features of sepsis and due to the systemic nature of the condition the endophthalmitis may sometimes be bilateral.

On examination, there may be a reduced red reflex, conjunctival injection, corneal haziness and hypopyon. It can be difficult to diagnose endogenous endophthalmitis due to the variability of the way in which it presents.

Investigation of this condition should include blood cultures and culture of vitreous samples to help identify and target the causative organism. The infectious diseases team should be involved to investigate and treat the patient for their underlying systemic infection.

At the time of vitreous biopsy, intravitreal antibiotics are also given and systemic antimicrobials should be targeted to the suspected organism. Pars plana vitrectomy, a surgical procedure to remove the vitreous gel, can be used for treating endogenous endophthalmitis and has proved to have a better outcome in terms of visual acuity and rate of enucleation. The ophthalmic complications of endophthalmitis include sight loss and in the worst cases there may be need for enucleation. (10, 11)

Summary

Endophthalmitis can result from infection introduced mechanically into the eye (exogenous) or through haematogenous spread from elsewhere (endogenous). Whilst the presentation may be similar, exogenous cases will have a history of eye surgery or trauma and endogenous cases will be more systemically unwell. This is an ophthalmic emergency and should be referred immediately for diagnosis and treatment.

Questions

Question 1: A 72 year old gentleman, with no prior ophthalmic history besides hypermetropia, presents with a 1 day history of a left red and painful eye with blurred vision and a pupil that is mid dilated and unresponsive to light. What is the most likely diagnosis?

a) Acute anterior uveitis

b) Angle closure glaucoma

c) Bacterial keratitis

d) Endogenous endophthalmitis

e) Exogenous endophthalmitis

THE RED EYE & SYSTEMIC INFECTION

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Question 2: A 65 year old lady presents to accident and emergency with a 1 day history of a left red and painful eye with blurred left eye vision only.

You learn she has a history of wet macular degeneration for which she received her 15th intravitreal injection to her left eye 1 week ago. Using a Snellen chart you find her right eye vision is 6/9 and left eye 6/60. What should the immediate next step of management be?

a) CT chest, abdomen and pelvis

b) Full blood count with CRP

c) Orbital X-ray

d) Peripheral blood cultures

e) Referral to on call ophthalmology service

Question 3: Which of the following patients is most likely to have endogenous endophthalmitis if they develop a red painful eye with visual loss and hypopyon?

a) A patient post cataract surgery

b) A patient who has recently had an intravitreal injection

c) A patient post pars plana vitrectomy

d) A septic patient on ITU

e) A patient following trabeculectomy

Question 4: Which is the most likely initial treatment for a patient presenting with endophthalmitis?

a) CT scan

b) Enucleation

c) Pars plana vitrectomy

d) Ultrasound B scan

e) Vitreous biopsy and intravitreal antibiotics

Question 5: Which condition is associated with a high mortality rate?

a) Acute anterior uveitis

b) Bacterial keratitis

c) Endogenous endophthalmitis

d) Glaucoma

e) Subconjunctival haemorrhage

Answers

1. Answer 1 – B

With no history of prior trauma or ocular surgery endophthalmitis is extremely unlikely. Hypermetropia and cataract are both significant risk factors for angle closure glaucoma and patients usually present with a fixed mid dilated pupil.

2. Answer 2 – E

Having recently had an intravitreal injection this patient is at risk of exogenous endophthalmitis and the features of red eye and painful vision loss is in keeping with this. Urgent referral to ophthalmology for diagnosis and management is indicated here.

3. Answer 3 – D

Endogenous endophthalmitis arises from extraocular infection such as sepsis. The other options are intraocular procedures and may cause exogenous endophthalmitis.

4. Answer 4 – E

Vitreous biopsy and intravitreal antibiotics is the best answer. Ultrasound B scan and CT scan are not treatments but the ultrasound scan can help visualise the posterior segment of the eye and CT scan may reveal underlying infection. Pars plana vitrectomy may be suitable for some patients but not all. Enucleation is usually a last resort.

5. Answer 5 – C

This is due to systemic infection.

THE RED EYE & SYSTEMIC INFECTION

AJ Simpson, A Krishnakumar, HDJ Hogg, MP Clarke

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AN APPROACH TO LACRIMAL GLAND MASSES

E Yang, V Lee

Abstract

The lacrimal gland is located anteriorly in the supero-lateral aspect of the extraconal space of the orbit, within the lacrimal fossa. It has two parts, a larger orbital lobe that extends deep into the orbital septum and a smaller palpebral lobe. Lacrimal gland diseases more frequently affect the orbital lobe, causing supero-lateral eyelid swelling.

A systematic approach to diagnosis and management of lacrimal gland masses is crucial. The pathology of these masses can be completely benign but can also be one of the first presenting features of a systemic and potentially life threatening disease that would need multidisciplinary input.

In this article, we discuss some common causes of lacrimal gland masses, how to differentiate them, the different approaches to obtaining a biopsy, and how to recognise any systemic associations and red flags.

Case History 1

A 59-year-old lady presented to the eye clinic with recurrent episodes of pain and swelling in her left upper eyelid orbital area. This had been present for the last 4 years, and the symptoms of pain were intermittent initially, but when she presented to our clinic she reported having daily attacks of pain and swelling in that area, which was not relieved despite maximal doses of oral non-steroidal anti-inflammatory medication.

She had no other systemic illness, autoimmune disease, nor any significant family history. On examination, visual acuity and colour vision were full, with no relative afferent pupillary defect (RAPD), and full range of movement of the extraocular muscles, although with pain on eye movements.

MRI head and orbits showed "a well circumscribed, 11mm, avidly, slightly heterogeneous enhancing mass within the lacrimal gland appearing to involve both the orbital and lacrimal lobes. There is no inflammation of the adjacent extra coronal fat. The lesion itself is marginally more T1 hyperintense than the adjacent muscle. There is no local bony scalloping or destruction. The right sided lacrimal gland appears normal... the left lacrimal gland is highly suspicious of a tumour".

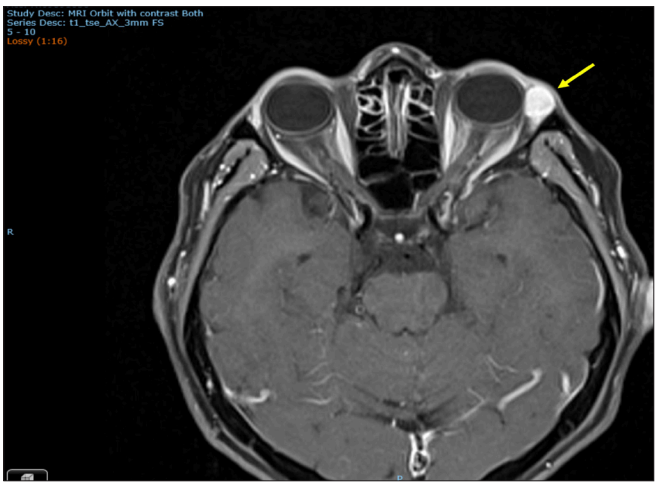

Figure 1: T1-weighted MRI brain and orbits showing a left sided well-circumscribed mass (yellow arrow) adjacent to the left globe.

In this case, the patient's history of intermittent pain and inflammation is more typical of dacryoadenitis (1), which is a common inflammatory and self-limiting lesion of the lacrimal gland. If this was the presumed case then a biopsy alone without complete removal would be adequate.

However, given the imaging diagnosis of a probable lacrimal gland tumour, which could be benign or malignant, complete removal of the mass would be required for a diagnostic sample, to prevent seeding of tumour cells into the orbit(2).

This can be done most optimally through a lateral orbitotomy approach, involving partial removal of the lateral orbital wall to fully access both the orbital and lacrimal lobes of the lacrimal gland (Figures 2 and 3).

AN APPROACH TO LACRIMAL GLAND MASSES

E Yang, V Lee

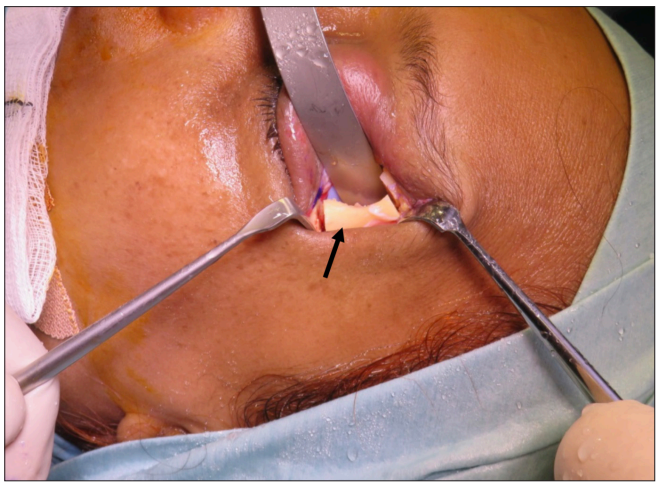

Figure 2: Retraction of the upper eyelid skin incision and deep tissue dissection to reveal lateral orbital wall (black arrow). The bony rim has been sawed down in preparation temporary removal. A blunt retractor protects the globe.

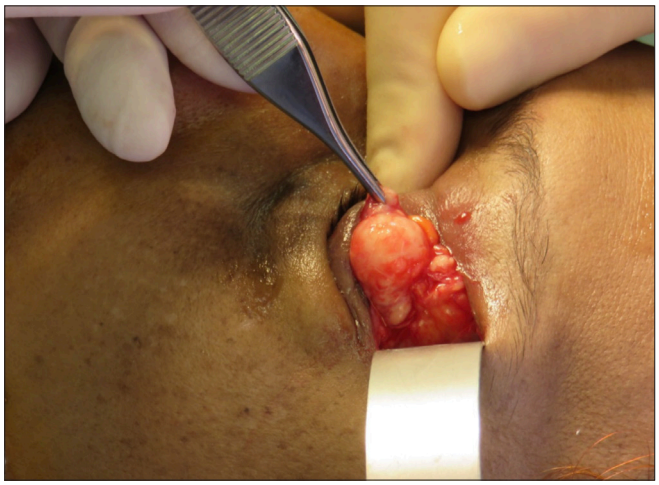

Figure 3: The above section of lateral orbital wall has been removed. Gentle tissue dissection reveals a large (1.2cm) well circumscribed, rounded mass, which was completely removed and sent for histology.

The lateral orbital wall section is replaced with small plates and screws after tumour removal.

Histology confirmed a benign mixed tumour (pleomorphic adenoma) with no atypia nor evidence of malignancy. The patient has now recovered from surgery, and is happy to be finally asymptomatic and rid of her persistent upper eyelid swelling. She has however developed significant dry eye and needed to have regular tear supplementation.

Discussion 1

A pleomorphic adenoma is the commonest lacrimal neoplasm, and accounts for nearly 20% of all lacrimal fossa lesions (3). They originate from epithelial and mesenchymal tissue, and can affect either orbital or lacrimal lobes, but most commonly the orbital lobe (4).

It is usually painless, therefore this patient's history was concerning for a more malignant and invasive lesion. There is small potential for malignant transformation (5). Because of the risk of tumour seeding, surgical removal of the whole tumour with an intact pseudocapsule, as in this case, is the recommended treatment (2).

With lacrimal gland removal, patients usually have resultant dry eye syndrome, which can be easily treated with regular artificial tear drops. Prognosis is excellent with complete surgical excision.

This case highlights the appropriate lateral orbitotomy approach to ensure entire tumour excision, and the need to exclude more malignant and potentially fatal causes such as an adenoid cystic carcinoma, mucoepidermoid carcinoma, or pleomorphic adenocarcinoma. These malignant tumours have a very poor prognosis with high mortality (3).

Case History 2

A 44-year-old lady presented to the ophthalmology clinic following a referral from her GP for a "right upper eyelid fullness" which had been present for at least one year. The swelling was painless, very slow growing, she had no double vision or other visual disturbance, no other medical history, specifically no known autoimmune disease nor any significant family history.

On examination, there was no proptosis as measured using an exophthalmometer, full range of eye movements, with a right upper-lateral eyelid swelling. On lid eversion the lacrimal lobe of the lacrimal gland was seen, with some conjunctival infiltration.

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She had an MRI head and orbits, which showed "A right preseptal soft tissue mass, adjacent but of slightly different signal characteristics to the right lacrimal gland is confirmed. It shows heterogeneous signal return with patchy contrast enhancement. There are no specific diagnostic features." (Figure 4).

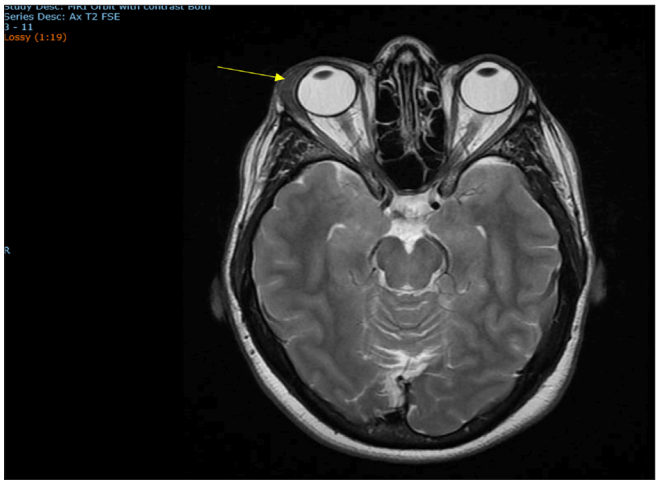

Figure 4: T2 weighted axial scan showing right eye superior-lateral soft tissue mass (yellow arrow) external to the globe.

Given the chronicity and slow enlargement of the mass, a biopsy for a diagnostic sample was necessary. Due to the MRI findings showing that the involvement was predominantly in the lacrimal lobe, a trans-conjunctival approach (Figure 5) for a biopsy to access the lacrimal lobe of the lacrimal gland was performed.

This is generally not a preferred approach, as there is a higher risk of damage to the lacrimal ductules. Also, lacrimal disease in general tends to affect the lacrimal lobe less frequently than the orbital lobe.

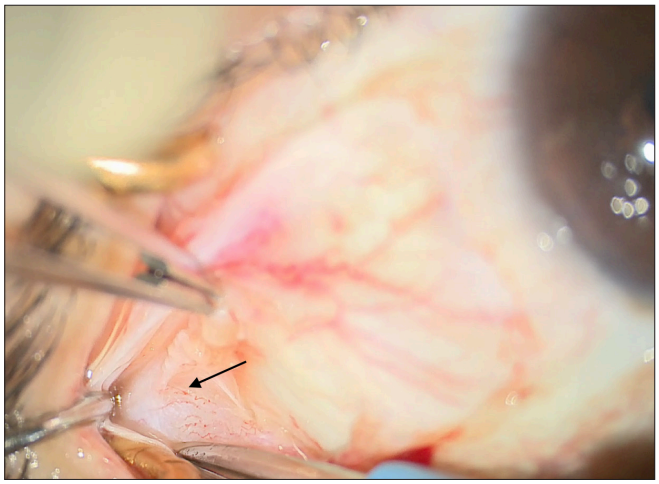

Figure 5: View under the operating microscope (Zeiss) on the superior-lateral aspect of the right eye. Conjunctiva and the underlying Sub-Tenon's layer has been dissected to reveal a white, minimally vascularized fatty tissue likely associated with lacrimal gland tissue, shown with black arrow.

The biopsy sample showed only fibrovascular tissue with no signs of malignancy. This patient was then started on oral non-steroidal anti-inflammatory medication, which helped to relieve some of her symptoms of eyelid heaviness and fullness.

Discussion 2

This patient's case highlights the unfortunately common event of diagnostic uncertainty. Although there were no features of malignancy, the option of a re-biopsy should be discussed with the patient.

A painful lacrimal mass can often be a sign of perineural invasion, and signs of involvement of other structures in the orbit such as the optic nerve (optic nerve involvement often manifests with positive RAPD, reduced visual acuity, colour vision and visual fields) and the extraocular muscles (double vision, restriction of eye movements) can be a sign of orbital malignancy.

Our patient had completely normal visual function and no other medical history. She remained well and clinically unchanged and the MRI scan showed little change at six months although the mass persisted.

A repeat biopsy was advised through an external palpebral approach (incision through upper lateral lid crease), to yield more diagnostic lacrimal tissue. However, the patient declined this and remains under observation.

Case History 3

A 28-year-old Ecuadorian lady presented to eye casualty with a painless right upper temporal eyelid swelling (Figure 6) which was intermittent over the last six months. She had previously been fully treated for TB and was on no medications and had no other systemic illness or family history.

Visual acuity, colour vision and eye movements were all normal at presentation. On examination, there was a small multi-lobulated swelling of the right lacrimal gland on eyelid eversion (Figure 7), which was non-tender and mobile, with healthy normal conjunctiva and overlying vessels.

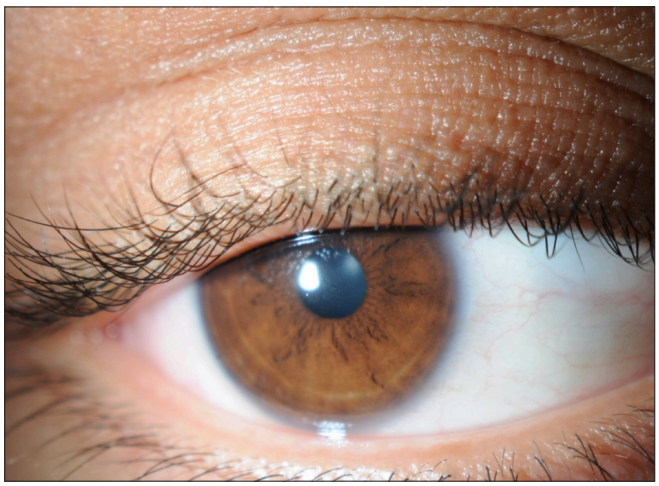

Figure 6: Very mild right upper lateral eyelid swelling on presentation.

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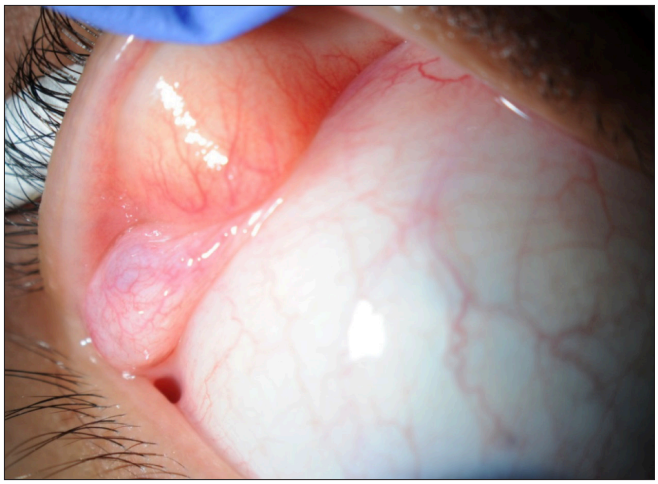

Figure 7: On upper lid eversion a small lobulated mass is seen, with no conjunctival infiltration.

She then developed diplopia with an increased right upper lid swelling, and was sent for orthoptic assessment a few weeks later, which showed right eye proptosis and restricted elevation of the right eye, suggesting a mass that was restricting globe movement.

An urgent CT orbits was requested (Figure 8), which showed "there is enlargement of the right lacrimal gland associated with proptosis as well as mild enlargement of the right superior rectus and possibly of the right lateral rectus muscles. The underlying bony changes suggest a subacute process. The differential diagnosis includes lymphoma, but further characterization is required".

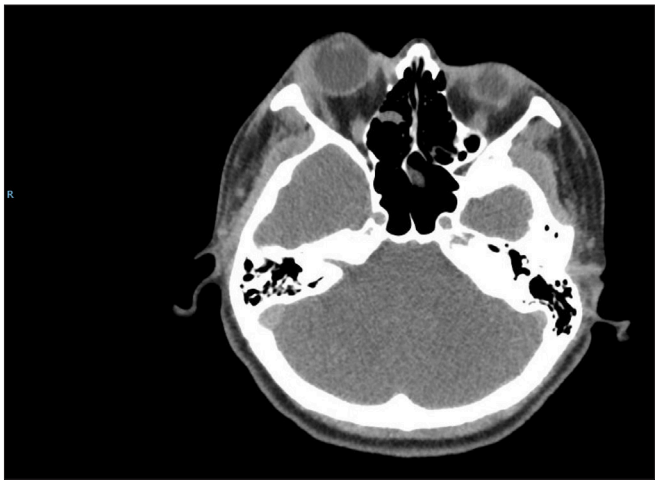

Figure 8: Right sided enlargement of lacrimal gland and recti muscles, shown on CT orbits with contrast. A differential of lymphoma is suggested.

A biopsy of the lesion was obtained through an external skin crease approach, which showed lymphocytic and plasma cell infiltrate, with no definite evidence of neoplasm nor lymphoma, with scanty IgG4+ plasma cells.

She was started on a reducing dose of oral prednisolone with a presumed diagnosis of orbital pseudotumour. Given her previous history of TB, she was referred to the haematology team for a systemic work up.

She was then found to have deranged renal function, and was eventually diagnosed by the medical team with ANCA positive cresenteric necrotising glomerulonephritis with acute renal failure.

She was admitted as an inpatient on the renal ward for plasmapheresis with cyclophosphamide infusions. At present, she is well on treatment, but is on renal dialysis and awaiting a renal transplant.

Discussion

This case highlights a mild lacrimal swelling as the first sign of a severe systemic disease. In this situation, a good history, follow-up plan, baseline investigations and prompt referral to appropriate multi-disciplinary teams was important in diagnosis and management.

Red flags in this case were the fact that the disease was evolving, with the development of double vision and proptosis, which suggests increasing orbital involvement and growth of the lesion. This again raises the importance of a thorough orbital exam at every follow up to assess important structures in the orbit, as summarised in Table 1:

Main structures within the orbit	Symptoms of dysfunction	How to assess function
Optic nerve	1. Reduced visual acuity 2. Reduced colour vision 3. Visual field defects – often subtle and not symptom 4. RAPD 5. Optic nerve changes eg. Pale disc, atrophy, swelling	1. Use distance visual acuity chart for both eyes eg. Snellen 2. Use Ishihara test plates and compare both eyes 3. Confrontational visual field testing, or formal Humphrey visual field testing for documentation 4. Swinging light test – the pupil that dilates more is the eye that has optic nerve compromise 5. Dilated ophthalmoscopy
Extraocular muscles	1. Binocular diplopia 2. Painful eye movements	1. Check that double vision resolves with one eye closed (binocular). Is it horizontal/vertical/oblique double images? Check for range of eye movements, document in which direction of gaze makes diplopia worse 2. Extraocular muscle restriction is more likely with pain on eye movements
The globe	1. Proptosis	1. Ask if patient has noticed change in facial appearance Use of exophthalmometer to document
Lacrimal gland	1. Painful/painless swelling of upper outer eyelid	1. Ask for any change in size of the lump, any change in tear production and dry eyes.

Table 1: Table describing a systematic approach to assess important structures within the orbit.

Case Based Discussion

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Clinically, it is important to be aware of systemic associations of lacrimal gland disease, so that useful baseline tests and imaging can be carried out, to direct referral to different specialties.

Table 2 describes a non-exhaustive differential of important systemic diseases that can manifest in the lacrimal gland with or without orbital signs and symptoms, and how to approach and refer if there are any concerns.

Differentials of lacrimal gland involvement as part of a systemic disease	**Features**	**History and exam**	**What to do:** **Always:** **1. Photos, CT/MRI** **2. Refer to Ophthalmology, consider biopsy**
Orbital inflammatory pseudotumour - Lacrimal gland inflammation - Can also involve extraocular muscles	Depends on structures involved – usually limited to orbit - Usally unilateral	Full history and Orbit exam (Table 1)	Refer to rheumatology: if severe will need immunosuppressants Blood tests: full rheumatology/autoimmune screen: - FBC, U&Es - ESR, CRP - TFTs - Rheumatoid factor - ANCA - ANA - Serum ACE - Imaging Joint care with ophthalmology
Sarcoid	Enlarged lacrimal gland, orbital inflammation (Diagnosis of exclusion)	Systemic history - Weight loss, fatigue - Cough, chest pain, SOB - Skin lesions - Joint pains - Genitourinary symptoms - Any family history Full eye and orbit exam	Rheumatology/autoimmune screen (as above) Often will need referral to chest physicians and joint care with rheumatology with ophthalmology input, will need to consider other ocular manifestations of sarcoid (uveitis, scleritis).
Inflammation eg. Wegener's granulomatosis /vasculitis, Ig4 disease	Usually painless Enlarged lacrimal gland, orbital inflammation (diagnosis of exclusion)	Systemic history	Rheumatology/autoimmune screen (as above) Will require serum Ig4 levels Referral to rheumatology/haematology, joint care with ophthalmology
Sjogren's syndrome	Very dry eyes Very dry mouth Usually bilateral lacrimal glands enlarged	Joint pains Enlarged parotids Lethargy	Rheumatology/autoimmune screen (as above) Referral to rheumatology, joint care with ophthalmology.

Table 2: Table summarizing some important associations of lacrimal gland involvement as part of a systemic disease.

It describes important features in the history and examination, and a basic summary of baseline investigations and further management.

Test Yourself

1. Of the options below, which is the most unlikely to be a symptom of invasive orbital disease?

a) Decreased vision.

b) Painful eye movements.

c) Seeing flashing lights and floaters.

d) Proptosis.

e) Double vision.

2. You are a Foundation Year 2 doctor in A&E. You are seeing a 60-year-old patient complaining of painful left sided eyelid swelling with double vision.

On further questioning and thorough systemic examination, you determine that the vision in her left eye has gradually worsened, her left eye appears more prominent than her right, and she reports unintentional weight loss over the last 2 months and generalised weakness. Out of the options below, which is your first initial course of action?

a) Suggest to the patient that they will need a biopsy sample taken of her eyelid today for diagnosis.

b) Call your Registrar and discuss your findings. Request a full set of bloods and an autoimmune screen, arrange for urgent CT orbit imaging.

c) Discharge the patient to her GP as she is systemically well.

d) Call the radiologist and request an immediate CT chest/abdo/pelvis to investigate weight loss.

e) Refer the patient to the next outpatient ophthalmology clinic.

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Answers

1. Answer: Option c)

Flashes and floaters in a patient's vision are more likely a symptom of a retinal tear or detachment, which does not usually point towards dysfunction of the structures within the orbit.

The other options could all suggest orbital pathology. Optic nerve dysfunction can cause decreased vision. extraocular muscle invasion/restriction can cause proptosis and double vision, and a mass large enough to push the globe forward can cause proprosis. Please refer to Table 1.

2. Answer: Option b)

This is a situation that any foundation doctor could encounter. This patient likely has developed an orbital lesion that is threatening optic nerve function, given her proptosis, double vision and decreasing vision.

This requires urgent orbital imaging for diagnosis and treatment as optic neuropathy can often be irreversible.

There is also suggestion here of systemic involvement given her weakness and weight loss, which could represent an autoimmune disorder, or more seriously, a sign of metastatic disease to the orbit.

This would require a full set of baseline blood tests including inflammatory markers and an autoimmune screen. This patient is also potentially unstable and complex, and your registrar should be your first port of call.

2. Incorrect answers:

a) Biopsy samples are never taken without appropriate imaging.

C) This patient requires urgent imaging and should not be discharged.

D) CT chest/abdo/pelvis is not warranted at this point as there is complete uncertainty about the diagnosis, without first orbital imaging and other blood tests for other organ dysfunction.

E) Again, the patient ideally should have adequate orbital imaging first, and might need a full systemic work up before discharging to an outpatient clinic.

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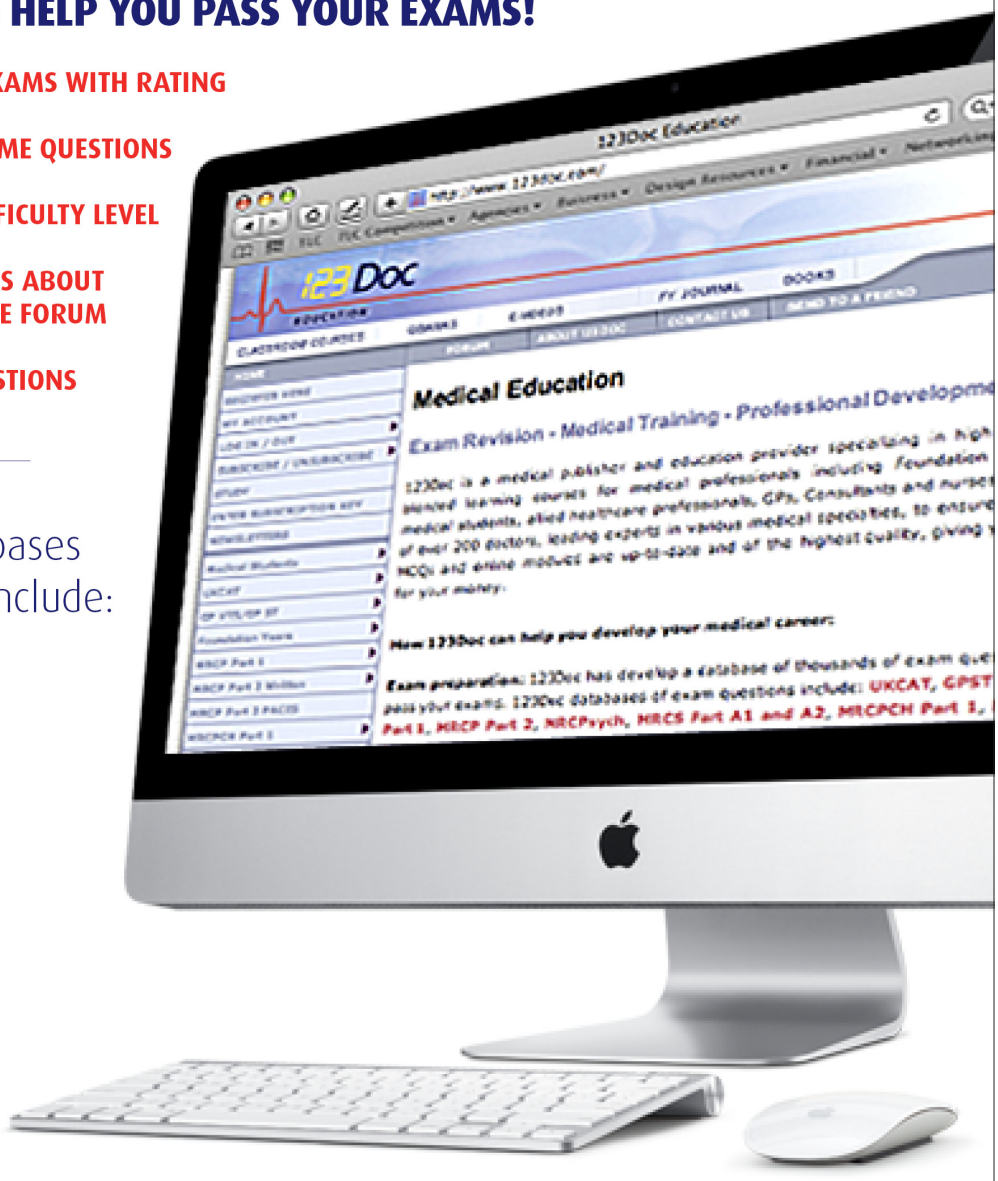

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