Unilateral proptosis typifying the variety in orbital pathology.

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Case Presentation

A 62 year old Caucasian lady presented to the Eye Casualty Department with a two week history of a painful red right eye with increasing 'prominence' of the same.

She had a history of hypothyroidism, breast cancer treated with mastectomy and axillary clearance with subsequent further axillary surgery for recurrence, hypertension and depression which had necessitated in-patient treatment on three prior occasions.

Her right visual acuity was 6/60 improving to 6/18 with pin-hole and that from her left eye 6/6 un-aided. Her pupil reactions were normal. In particular there was no relative afferent pupillary defect. As can be seen from figure 1 here was thickening of the periorbital tissues bilaterally with the right appearing erythematous additionally. The conjunctiva on the right was injected and chemosed. Exophthalmometry measured right and left proptosis of 26 mm and 23 mm respectively. The conjunctival chemosis and proptosis prevented complete closure of this patient's right eyelids. All ocular movements on this side were limited. Her optic nerve head looked normal.

Computed tomography of brain and orbits with and without contrast was performed. Uniform enlargement of the extraocular muscles on the right which spared the muscle tendons was apparent. No abnormalities of the cavernous sinus were noted.

A diagnosis of asymmetric thyroid eye disease was made and this lady treated with 1g methylprednisolone intravenously daily for 3 days followed by oral prednisolone at a starting dose of 40mg daily and tapered slowly over 3 months. Her orbital inflammation resolved and she regained full eye movements.

Discussion

Patients with a diverse range of orbital pathologies present with a limited number of similar symptoms and signs. This can make their management interesting but challenging. Table 1 details a number of conditions, any one of which may have readily explained this lady's presentation.

As can be seen from this table imaging with computed tomography and/or magnetic resonance imaging which can be combined with angiography as necessary is essential in determination of the underlying disease process.

With such a presentation the main reasons for visual loss are given in table 2.

Examination of optic nerve function is mandatory and involves what is shown in table 3.

Similarly, an examination of lid function and in particular lid closure and an examination of the cornea is also necessary. If there is lagophthalmos and/or evidence of exposure keratopathy attempts must be made to achieve eyelid closure adequate enough to protect the cornea while the underlying pathology is addressed. Ways in which this can be achieved are given in table 4.

Orbital disease is a relatively uncommon ophthalmological presentation. Early diagnosis using clinical and radiological signs and, in some cases, histopathological findings allows for the prompt institution of appropriate management which can be sight and in such cases as those of orbital cellulitis or cavernous sinus thrombosis, even life-saving.

Figure Legend

Figure 1 Oedema and erythema of the periorbital tissues on the right is indicated by the red arrow. The conjunctiva is injected. Conjunctival chemosis which is worst inferiorly is indicated by the blue arrow.

Figure 2 Two coronal sections of computed tomography imaging of the orbits. The red arrows indicate enlargement of all four rectus muscles on the right.

Figure 1

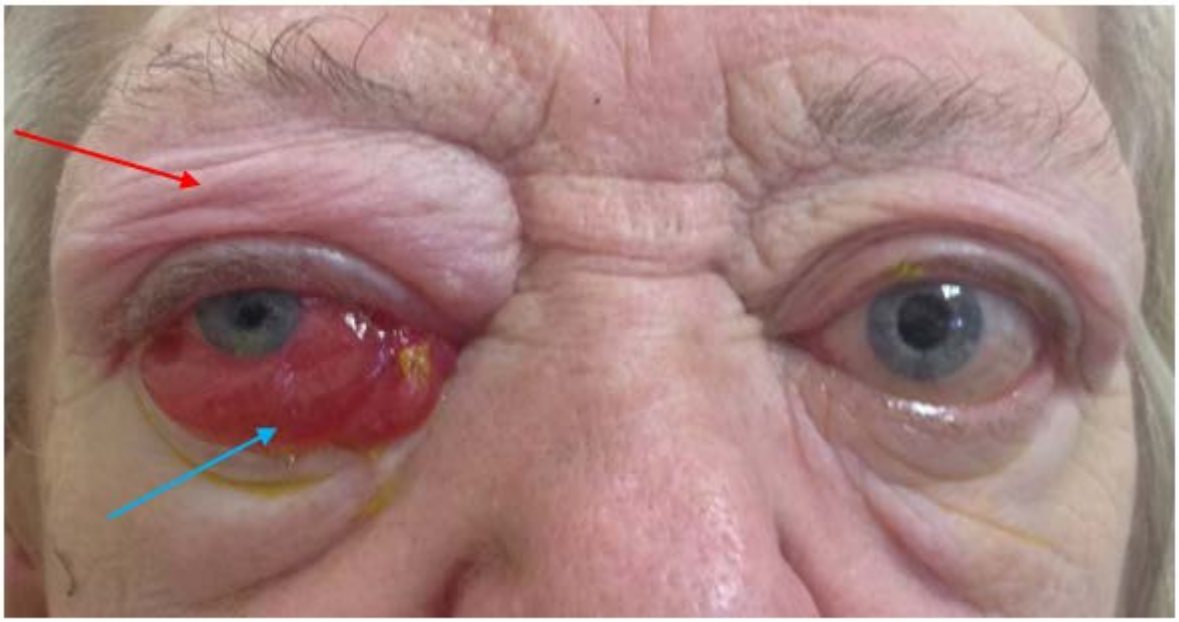

Figure 2

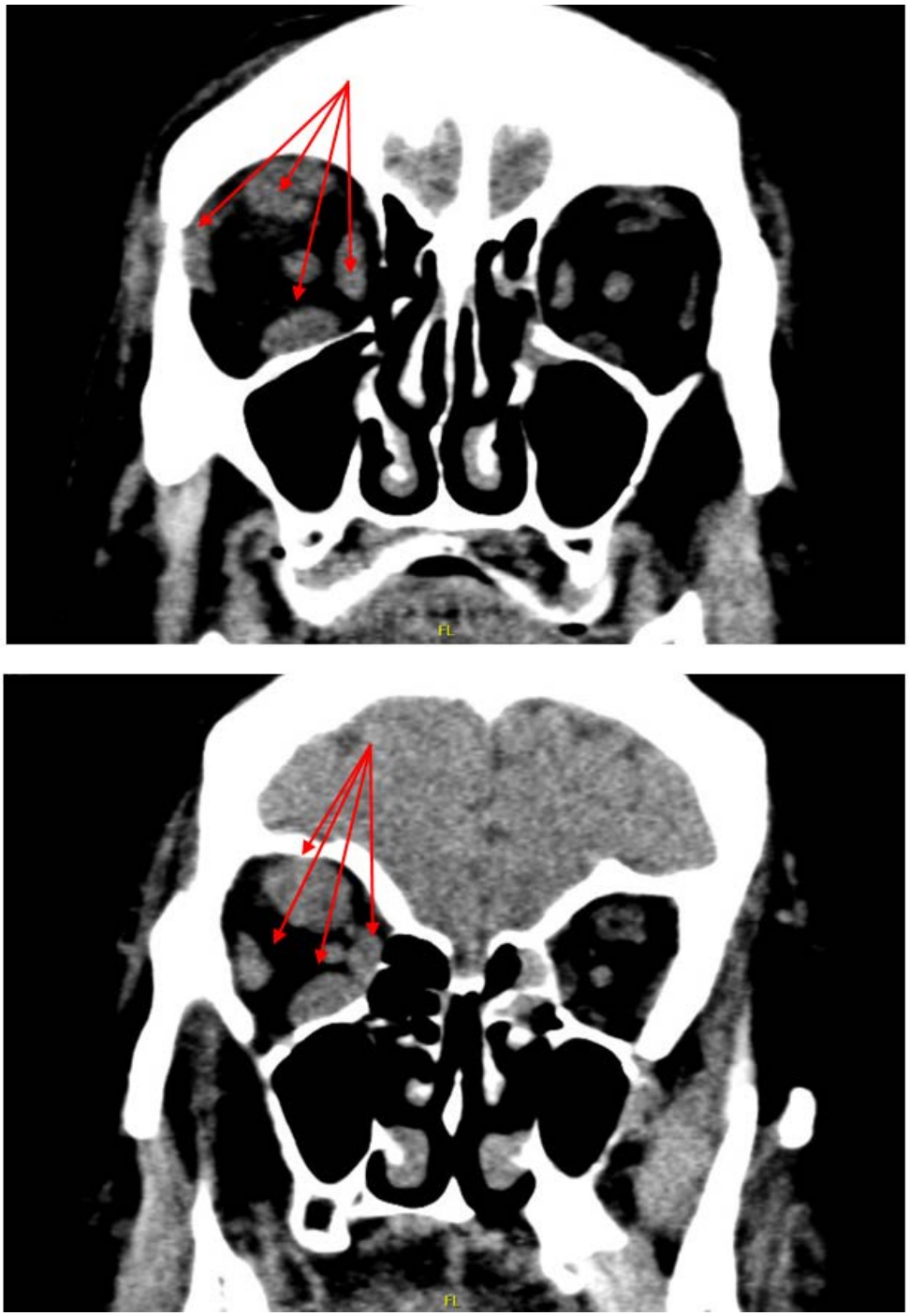

Table 1 The acutely inflamed orbit

Diagnosis	Pathophysiology	Imaging	Management
Thyroid eye disease	There is inflammation of the extraocular muscles, interstitial tissues and orbital fat and a consequent osmotic imbibition of water with an increase in the volume of the orbital contents as T-cells directed cells of the thyroid gland in autoimmune thyroid disease react against orbital tissues which share similar antigens. While the majority of patients with thyroid eye disease are hyperthyroid a proportion are euthyroid or hypothyroid at presentation. Thyroid eye disease remains the commonest cause of unilateral and bilateral, acute and chronic proptosis.	Enlargement of the extraocular muscles which spares the muscle tendons.	Exposure keratopathy of which this lady had signs and/or optic neuropathy necessitates treatment with intravenous methylprednisolone 1g daily for at least three days. Orbital decompression surgery should be considered thereafter if there is no sustained improvement.
Orbital metastases breast carcinoma	Metastases to the orbit are infrequent but breast carcinoma is the cause in 70% of cases.	Non-capsulated mass within the orbit.	Treatment should be directed initially at the underlying malignancy. Radiotherapy is the mainstay of local therapy. Systemic chemotherapy may also be of benefit in some cases. Surgical excision of the metastatic focus is also occasionally carried out.

Carotid-cavernous fistula	Low-flow or indirect carotid-cavernous fistula are communications between branches of the internal and/or external carotid artery and the cavernous sinus. They usually develop in older females like the lady described here with hypertension and atherosclerosis. There is subacute onset of painful proptosis associated with haemorrhagic chemosis ssuch as that described here.	Prominence of the superior ophthalmic vein and diffuse enlargement of the extraocular muscles. Angiography may be required to definitively demonstrate the fistula.	Many low-flow or indirect fistulae do not need treatment but rather close spontaneously. The artery may be repaired or the sinus occluded e.g. with a balloon or coil via a trans-arterial approach in other cases.
Idiopathic orbital inflammatory disease	Non-neoplastic, non-infective, space occupying inflammation of the orbit. Unilaterality is usual in adults.	Ill-defined opacification of the contents of the orbit.	Idiopathic orbital inflammatory disease is a diagnosis of exclusion and so biopsy may be required to exclude other diagnoses. Treatment is with local i.e. orbital injection of corticosteroid and/or systemic corticosteroid.
Orbital cellulitis	Infection of the tissues behind the orbital septum. A combination of reduced vision, conjunctival injection and chemosis, a limitation of eye movements and optic nerve compromise differentiate orbital cellulitis from periorbital cellulitis, infection of the skin and subcutaneous tissues anterior to the orbital septum.	Infiltration of the orbital tissues often with sinusitis from which the infection frequently originates.	Intravenous antibiotics. A typical initial regime is ceftazidime, metronidazole +/- flucloxacillen.

Cavernous sinus thrombosis	Infection of the cavernous sinus from the paranasal sinuses, ear or orbit and periorbital tissues. The individual is usually acutely unwell.	Magnetic resonance venography demonstrates incomplete filling of the cavernous sinus.	Intravenous broad spectrum antibiotics. Occasionally surgical drainage of the sinus is warranted.

Table 2 Causes for visual loss in orbital disease

Optic neuropathy as the optic nerve is stretched and/or compressed within the orbit

Exposure keratopathy resulting from the combination of proptosis and lagophthalmos

Glaucoma resulting from acute elevations in orbital and so intraocular pressure

Retinal vascular, usually venous, occlusion as a consequence of raised episcleral venous pressure

Globe subluxation

Table 3 Assessment of optic nerve function

Measurement of best corrected visual acuity.

Pupil examination. A relative afferent pupillary defect is an important sign of optic nerve compromise.

Colour vision. Optic nerve dysfunction affects red/green colour vision relatively early.

Brightness perception.

Optic nerve head appearance. Optic nerve head oedema and atrophy suggest acute and chronic optic nerve dysfunction respectively.

Visual field assessment either by confrontation or by formal visual field testing if this is available and the patient is able to undertake the test.

Table 4 Means of addressing corneal exposure

Moisture chamber
Taping eyelids completely closed
Temporary tarsorrhaphy
Botox induced ptosis

Orbital disease is a relatively uncommon ophthalmological presentation but, as we hope this case illustrates, its prompt and appropriate management can be sight and in such cases as those of orbital cellulitis or cavernous sinus thrombosis, even life-saving.