

## BENIGN AND MALIGNANT DISEASES OF THE ORBIT – A REVIEW

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### Summary

Although the orbit is affected by a broad spectrum of pathology, including structural, inflammatory, infectious, vascular, neoplastic and degenerative processes, the symptoms and signs of orbital disease are limited. To the astute clinician, however, a thorough history and examination for both ocular and systemic disease usually leads to a concise differential diagnosis and can guide further investigation. The clinical assessment of orbital disease, radiologic interpretation, and an approach to diagnosis, investigation, and management of commoner benign and malignant orbital diseases are presented in this review.

### Benign and Malignant Orbital Disease – Keypoints for the Clinician:

- Orbital examination includes a full assessment of optic nerve function (visual acuity, colour vision, visual fields, pupil reactions, and disc assessment), axial and non-axial globe position, ocular balance and motility, and intraocular and periorbital structures. Systemic examination is guided by the symptoms.
- The imaging of choice for orbital disease is CT. MRI may provide further detail of intrinsic optic nerve disease and orbital apical or intracranial pathology.
- Ultrasonography has higher resolution than CT and MRI and is valuable in assessing intraocular lesions and anterior orbital masses - in particular vascular lesions.
- Orbital inflammation is not a diagnosis but a tissue response to a wide range of pathologies, and immunosuppression should not be instituted until an adequate biopsy has been obtained. The exceptions to this general principle are typical scleritis, myositis, thyroid eye disease, and characteristic orbital apex syndrome, in which delay in suppression of apical inflammation may jeopardize visual outcome.
- Similarly, the term “orbital pseudotumour” is not a diagnosis, has often led to inappropriate

management, and is no longer in use.

- Thyroid eye disease is the most common cause of unilateral and bilateral proptosis. Management of aggressive disease consists of immunosuppression in the early, “active” phase, with nonresponsive patients requiring urgent decompression in the presence of optic neuropathy. Stable, inactive disease is managed by orbital decompression for exophthalmos, followed by correction of muscle imbalance and lid malposition.
- Subacute lacrimal gland inflammation, unresponsive to a few weeks of non-steroidal treatment, may be due to underlying carcinoma and a specialist opinion should be sought without delay.

## Introduction

Orbital disease may be limited to the soft tissue and bony structures of the orbit, or may be part of a more widespread systemic disease. A careful history and examination often points to the likely diagnosis, and modern imaging and histological techniques further define the extent and severity of disease. This review covers the more commonly encountered conditions, with summary of current best management practice. The spectrum of orbital disease differs between children and adults, as shown in Tables 1 and 2. Benign and malignant lacrimal gland lesions are covered in review, published in JBCR, Vol 3, N 2, 2010, but trauma and its management is not dealt with in this review.

**Table 1.** Common childhood orbital diseases

Childhood orbital diseases	
1.	Orbital infections
2.	Dermoid cyst
3.	Dermolipoma
4.	Vascular anomalies
5.	Malignant tumours: rhabdomyosarcoma and neuroblastoma
6.	Primary optic nerve tumours

**Table 2.** Common adult orbital diseases

Adult orbital diseases	
1.	Orbital infections
2.	Sinus mucocoeles
3.	Vascular lesions and haemorrhage
4.	Idiopathic inflammatory disease
5.	Benign orbital tumours
6.	Malignant orbital tumours

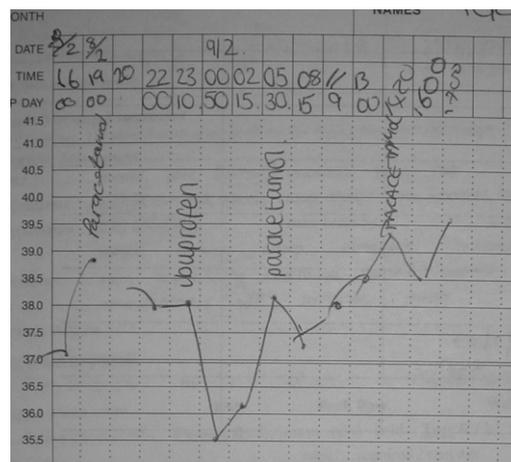
## Orbital Disease in Children

### Orbital infections

The average age for orbital infections in children is about seven years, although, for reasons unknown, it is almost twice as common in boys [1]. Presenting clinical features include pain, heat, redness, and swelling in the periorbital region, and a history of fever, sinusitis, upper respiratory tract infection and trauma is common (Figure 1 and 2). In *pre-septal* infection, lid oedema and redness are present, but ocular examination is otherwise normal. Early *post-septal* clinical features may be quite subtle, with signs of increasing severity including a red demarcation line (corresponding to the arcus marginalis), chemosis, proptosis, reduced eye movements, and loss of vision. More severe disease can lead to visual loss due to orbital congestion and ischaemia, orbital apex syndrome and cavernous sinus thrombosis. In the most severe infections, intracranial spread can cause meningitis and intracerebral abscess [2].



**Figure 1.** Orbital cellulitis in a four year old child with associated ethmoid sinusitis



**Figure 2.** Observation chart of same patient demonstrating swinging pyrexia

Although a reliable history and examination are essential, investigations (which include haematology, microbiology, and orbital ultrasonography) are frequently non-contributory and tend to delay the initiation of potentially sight- and life-saving treatment. CT imaging, with contrast, defines the extent of sinus disease and the presence of a subperiosteal or orbital abscess (Figure 3), but organising imaging should not delay treatment for suspected orbital infection [3]. MRI is the secondary choice in orbital imaging and valuable when cavernous sinus, intracranial extension, or radio-lucent foreign bodies are suspected. Finally, lumbar puncture is reserved for children with features of meningitis.



**Figure 3.** Subperiosteal orbital collection in a child with deteriorating orbital cellulitis

The most commonly isolated organisms in children with post-septal infection are - or non-haemolytic streptococci, Group A - haemolytic streptococci, *Staphylococcus aureus* and *Haemophilus influenzae*; the latter organism is becoming rarer with programmes for immunization [1]. Among those requiring drainage, positive cultures from abscesses or the sinuses may be expected in two-thirds, with polymicrobial infection occurring in over a third. A variety of other aerobic organisms are implicated in orbital infections, including Group C -haemolytic streptococci, *Eikenella corrodens*, *Arcanobacterium haemolyticum*, and *Moraxella catarrhalis*. Anaerobes are encountered less frequently than aerobes, but carry a higher morbidity than aerobic organisms; these include *Peptostreptococcus*, *Bacteroides sp*, and *Fusobacterium necrophorum*.

Active and early involvement of both the paediatrician and the otorhinolaryngologist is essential in managing infective orbital disease. Daily out-patient review, and treatment with oral antibiotics, is appropriate for otherwise-well

children with mild pre-septal inflammation. In all other cases, the child should be admitted for intravenous antibiotics, with systemic and visual functions being monitored and recorded at least thrice daily, depending on the severity of the clinical signs.

Previously, the development of a subperiosteal abscess *per se* has been considered an indication for urgent drainage. Currently, indications for medical treatment include normal visual functions, age younger than nine years, medial location of the abscess, and no evidence for intracranial or frontal sinus involvement. Conversely, the indications for drainage include progression of disease despite medical management, visual failure or ophthalmoplegia, and a large and well-defined abscess [1].

### **Dermoid cyst**

Most dermoid or epidermoid cysts, the latter lacking dermal elements, are noted early in infancy [4]. They form from an invagination of ectoderm into the orbit occurring either after an injury (these occurring at any age) or during embryological development. Of the latter, two-thirds lie in the anterior orbit, associated with bony suture lines, and one-third occur deep within the orbit. Of the anterior cysts, two-thirds lie in a superolateral position ('external angular dermoid' Figure 4), and the others in the region of the fronto-maxillary suture. Dermoids are usually associated with bony scalloping, or may extend in a dumbbell fashion through the orbital wall. They enlarge gradually, and frequently become inflamed due to leakage of their contents, which include oil, proteinaceous material and epithelial debris (Figure 5). While lateral dermoids tend to be mobile and do not usually require imaging, patients with a medial dermoid



**Figure 4.** Dermoid cyst

should be investigated with CT imaging to exclude a rare orbito-cranial cleft with associated herniation of the meninges (meningocoele) or brain (encephalocoele). Dermoids should be excised intact before the child attends school – the majority of these being conveniently approached through an upper eyelid skin-crease incision.



**Figure 5.** Dermoid cyst – transillumination revealing oily content and small amount of proteinaceous supernatant

### ***Dermolipoma***

Formed from cutaneous adnexal tissues sequestered on the conjunctival surface (Figure 6), these choristomatous lesions may be associated with clefts of the outer canthus and Goldenhar's syndrome. Careful excision of the abnormal epithelium and underlying fat should be undertaken using the operating microscope, and care should be taken not to injure either the lacrimal gland ductules or lateral rectus Tenon's sheath [5]. Preoperative counseling should include the small, but real, risks of postoperative diplopia and dry eye.



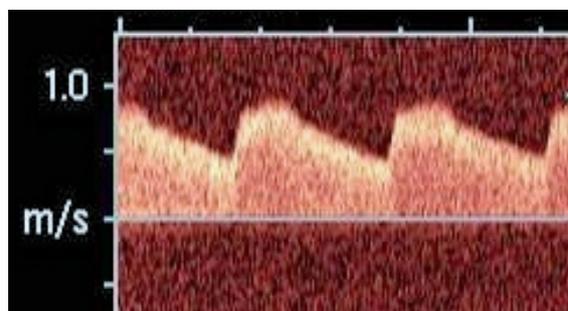
**Figure 6.** Dermolipoma

### ***Vascular anomalies***

Capillary haemangiomas may present within the first few months of life, frequent enlarging within the first year followed by cyclical proliferation and atrophy before eventual spontaneous involution (Figure 7) [6]. Due to their prolific early growth, this being disproportionate to the expansion of the bony orbit, they may distort or displace the globe, or cause a mechanical ptosis – all of which may lead to amblyopia. These children should be investigated with B-scan ultrasonography that shows a vascular lesion with high internal blood velocity (up to 1 m per second, Table 3, Figure 8), this differentiating a haemangioma from rhabdomyosarcoma, which is relatively avascular. Management involves regular assessment of visual development, with treatment offered for large lesions presenting a significant risk for the development of amblyopia [7].



**Figure 7.** Capillary haemangioma



**Figure 8.** Capillary haemangioma – Doppler ultrasonography demonstrating internal blood velocity up to 1 m per second

Until recently the first-line therapy for capillary haemangioma was systemic or periocular corticosteroid therapy; however, both have significant side effects and may need to be used for extended periods. Recently, it has been observed that propranolol, a non-selective beta-adrenergic antagonist, inhibits haemangioma

matous growth and this drug is now used as a first-line treatment in many units [8, 9]. As  $\beta$ -blockade carries systemic risks – including bradycardia, cardiac failure and conduction disorders, and hypotension – the administration of this medication should be supervised by a paediatrician. A protocol for propranolol treatment is given in Table 4.

**Table 4.** Details of propranolol therapy

Pre-treatment investigations	Dosage regime of propranolol
Full clinical examination	Week 1: 1mg/kg/day, divided into three doses
Routine blood tests to include thyroid function tests	Week 2: Dose increase to 2mg/kg/day divided into three doses, with a monthly dose adjustment according to the weight of the child up to 9 months of age if there is no clinical improvement
ECG	From month 9: Maintain the same dose, without weight adjustment, until a year of treatment – unless there is a need to continue beyond this time
Cardiac and abdominal ultrasonography	
Photography	

### **Malignant tumours of the childhood orbit**

These rare tumours include rhabdomyosarcoma (which may originate in the orbit or sinuses) and disseminated metastases from neuroblastoma, leukaemia, or histiocytic diseases (such as Langerhans cell histiocytosis) [10-12]. Typically enlarging rapidly, rhabdomyosarcoma may be associated with inflammation (Figure 9), while periorbital bruising may occur with neuroblastoma. Ultrasonography confirms the presence of a solid lesion, differentiating these neoplasms from capillary haemangiomas which also enlarge rapidly early in infancy. Characteristic features on CT imaging for rhabdomyosarcoma include a large mass displacing the normal orbital structures and, despite the misleading terminology, the mass is NOT associated with the extraocular muscles. In children with neuroblastoma, imaging often shows an osteolytic lesion within the orbital walls or an infiltrative orbital mass. All patients suspected of harbouring a malignancy should undergo an urgent biopsy and, in those with rhabdomyosarcoma, the mass can often be removed intact. The need for adjuvant systemic chemotherapy and radiotherapy depends on the tumour histology and presence of systemic disease.

**Table 3.** Doppler ultrasonography in capillary haemangioma

key features	High internal acoustic heterogeneity
	High spatial density of vessels throughout the tumour (typically more than 5/cm <sup>2</sup> )
	High internal blood flow velocity (as high as 100 cm/s), often with arterial wave-forms



**Figure 9.** Rapidly enlarging rhabdomyosarcoma with associated inflammatory signs

### **Primary optic nerve tumours**

Glioma and meningioma may present during childhood or early adult life, and are usually benign. Gliomas lead to gradual proptosis and mild visual loss; CT imaging identifies a fusiform expansion of the optic nerve, while MR studies may show more extensive changes within the intracanalicular and intracranial portions of the optic nerve (Figure 10) [13]. The management of gliomas includes neurosurgical excision or chemotherapy if tumour progression threatens the optic chiasm, and orbital resection in patients with significant proptosis and a blind eye.



**Figure 10.** MRI showing optic nerve glioma with expansion of the nerve sheath

Optic nerve meningiomas, in contrast, rarely cause significant exophthalmos but can impair optic nerve perfusion and cause slowly-progressive optic neuropathy [14]. Diffuse expansion of the optic nerve outline is shown on CT, with or without calcification in the nerve sheath (Figure 11). On MR imaging, a normal or small nerve is seen coursing through an enlarged sheath. Neurosurgical resection may be considered in younger people, in whom tumour growth can be quite active, and where there is a risk intracranial involvement. Amongst older patients, high-dose fractionated radiotherapy (~45Gy) is recognised to improve vision where the tumour is causing optic neuropathy [15].



**Figure 11.** CT identifying meningioma of the optic nerve sheath

## Common Adult Orbital Disease

An accurate history and examination, with particular attention to the rapidity of onset of symptoms, frequently leads the clinician to the

correct diagnosis. In orbital myositis, for example, the patient typically describes a brief prodromal period followed by an acute orbital ache, redness and pain and diplopia looking away from the field of action of the affected muscle. In contrast, cavernous haemangioma may present without pain, a very gradual increase in proptosis, and slowly increasing hypermetropia. In all patients, the previous medical history is critical: an elderly lady presenting with globe displacement some years after treatment for breast carcinoma has orbital metastatic disease until proved otherwise. The young man with chronic sinus symptoms complaining of a month's history of a swollen, red upper eyelid may have developed a frontal mucocoele with orbital extension. Common among patients of African and West Indian descent, sarcoidosis may have been diagnosed a long time before the patient attends with painless upper lid fullness due to chronic dacryoadenitis.

## Orbital infections

Orbital infections in adults are typically caused by Gram-positive bacteria (particularly *Staphylococcus* and *Streptococcus* species), with sinus disease – usually of the ethmoid air cells – being present in up to three-quarters of cases (Table 5). The complications of orbital infection may be devastating (Table 6) and immediate management is therefore of paramount importance [1].

Ancillary investigations, such as a white blood cell count with differential, and microbial cultures, are typically unhelpful in managing orbital cellulitis. However, thin-slice CT imaging with contrast helps to define the extent of sinus and orbital involvement and, with appropriate soft tissue windows, might reveal a foreign body.

Patients with mild pre-septal cellulitis and without evidence for orbital or systemic disease can be managed as an outpatient with oral antibiotics. This is in direct contrast to those with suspected post-septal infection, who should be commenced *immediately* on a high-dose, broad-spectrum intravenous antibiotic (Figure 12). A combination of parenteral cefuroxime (1.5 g) and metronidazole (500 mg) each eight-hourly is a good empirical approach until culture results are known and the advice of a microbiologist obtained. Where coexistent sinus, ear or oral disease is present, these aspects should be managed by appropriate sub-specialists; a nasal decongestant can be useful in improving drainage

**Table 5.** Orbital infections – Aetiology

Infection from neighbouring structures	nose, sinuses, lacrimal sac
Direct injury	surface injury, penetrating injury, and surgery
Pre-existing orbital disease	e.g. dermoid with a fistula to the skin
Haematogenous spread	including opportunistic infections in an immunocompromised patient

**Table 6.** Complications of orbital infection

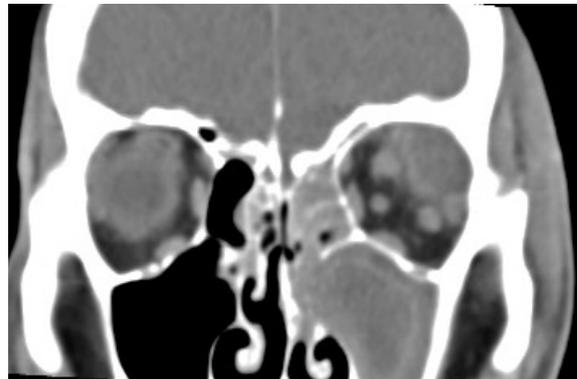
1.	Visual loss due to corneal exposure, raised intraocular pressure, endophthalmitis, vascular occlusion, mechanical compression of the optic nerve, and optic neuropathy
2.	Cavernous sinus thrombosis
3.	Superior orbital fissure syndrome
4.	Meningitis (2 %)
5.	Subdural empyema
6.	Intracranial abscess (3 %)
7.	Septicaemia
8.	Renal failure
9.	Toxic shock
10.	Death

of the paranasal sinuses.



**Figure 12.** Right orbital cellulitis with ptosis, chemosis, and restricted movements

The development of a subperiosteal abscess occurs either due to direct extension from an infected sinus, or by local haematogenous spread through the valveless veins between the sinuses and the orbit (Figure 13). Although *subperiosteal*, the penetration of antibiotics into this space is poor and visual loss, or expansion of the abscess into the cranial fossa, may rapidly ensue. If intracranial extension is suspected, thin-slice MRI, with fat suppression, should be performed to determine the extent of disease.



**Figure 13.** Left subperiosteal orbital abscess and ipsilateral sinusitis

Most patients improve on medical measures alone, with constitutional symptoms generally responding before the orbital signs, and the need and timing for surgical intervention depend on the history and clinical signs. Most orbital collections are subperiosteal; intraorbital abscesses are rare and usually result from ocular surgery, penetrating orbital injury, or infected haematogenous emboli. A collection which does not respond to antibiotic therapy is a clear indication for drainage, with the approach determined by the location of the abscess (Table 7). Elevating the periorbita until the anterior extent of the abscess is reached, the contents are released, the cavity irrigated with saline, and a corrugated or vacuum drain left in place until fluid drainage ceases -- typically within 48 hours (Figure 14).



**Figure 14.** Drainage of bilateral superior orbital subperiosteal abscesses via the skin crease

### **Necrotising fasciitis (NF)**

This aggressive, rapidly-progressive disease is typically caused by group A - haemolytic *Streptococcus* [16]. Spreading along subcutaneous fascial planes, periocular NF is a true ophthalmic emergency that can lead to septicaemia, multi-organ failure and death in up to a quarter of patients (Figure 15). NF requires urgent treatment with high-dose intravenous penicillin and clindamycin, with debridement of the affected necrotic subcutaneous tissues.



**Figure 15.** Moribund patient with periocular necrotising fasciitis

### **Sinus mucocoeles**

Sinus mucocoeles that affect the orbit arise most commonly from the fronto-ethmoidal recess [17]. They cause a gradual displacement of the globe and inflammation due to the leakage of necrotic debris (Figure 16). A more acute compressive optic neuropathy may rarely arise from infective orbital cellulitis and from chronic

sphenoidal mucocoeles. Infrequently, a fronto-ethmoidal mucocoele can discharge spontaneously through the upper lid, resulting in a fistula that resembles an ulcerated periocular epithelial malignancy. Rarely, mucocoeles extend intracranially to form an extra-dural collection of mucus or pus.

All such retention mucocoeles should be drained, frequently possible through an endonasal route [18], although superolateral orbital collections can require simultaneous direct orbital drainage through a local skin-crease incision. In cases of intracranial extension of an abscess, neurosurgical intervention is likely to be required.



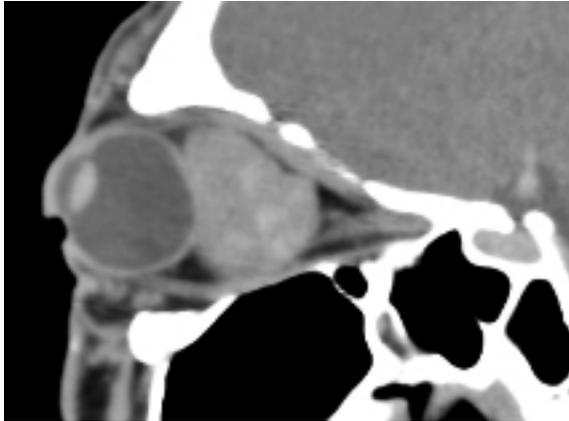
**Figure 16.** Huge frontal mucocoele depressing the contents of the orbit

### **Vascular lesions and haemorrhage**

Low-flow vascular anomalies such as varices, lymphangiomas and cavernous haemangiomas may originate in childhood, but only present in adult life [19]. The most common adult orbital lesion is cavernous haemangioma, which typically presents with a slowly-progressive and painless proptosis in the 4<sup>th</sup>-5<sup>th</sup> decades (Figures 17 and 18). Compression of the globe can lead to premature monocular presbyopia or to choroidal folds that can persist after surgery. CT often reveals a well-defined retrobulbar lesion with patchy enhancement due to extremely slow perfusion. Although asymptomatic lesions can be managed with observation alone, those causing optic neuropathy, choroidal folds or significant proptosis should be excised. The patient should be counselled that surgery carries a small risk of visual loss due to proximity of the haemangioma to the optic nerve or the orbital apex [20].



**Figure 17.** Gradual onset of proptosis due to intraconal cavernous haemangioma

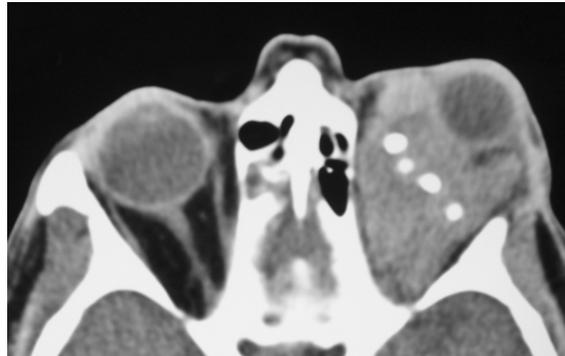


**Figure 18.** CT showing cavernous haemangioma compressing the globe

Orbital varices and lymphangiomas are congenital *low-pressure* venous-type malformations that are usually unilateral and may also involve other parts of the head and neck [21]. Varices are generally a primary low-pressure anomaly, presenting in young adulthood as intermittent proptosis on bending or straining (Figure 19). Orbital enlargement is often visible on CT, together with multiple serpiginous opacities and calcified thrombi (Figure 20). Whilst varices are blood-filled and in communication with normal orbital or cranial veins, 'lymphangiomas' are largely isolated from the venous system and have a greater inflammatory infiltration. The latter typically present with spontaneous orbital haemorrhage in the early school years, often at night, or as variable proptosis which is worse during upper respiratory tract infections. Management of these hamartomas is difficult and rarely curative; surgical excision carries a major risk of visual loss, as the normal orbital structures often pass through the anomaly. Nevertheless, haemorrhagic cysts often require drainage if they cause persistent optic neuropathy, and larger lesions may require excision for aesthetic reasons.



**Figure 19.** Huge orbital varix causing proptosis and hypoglobus



**Figure 20.** CT identifying phleboliths within orbital varix

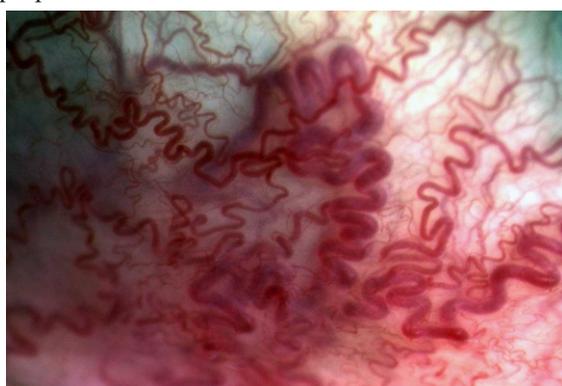
High pressure arterio-venous anomalies which affect the orbit can be divided into low-flow and high-flow shunts. *High-pressure, high-flow* anomalies (spontaneous in atheromatous individuals, or post-traumatic) (Figure 21) are rare and may result from shunting within the orbit or the anterior part of the intracranial [22]. These present with pulsatile proptosis, severe chemosis, a global reduction in eye movements, dilated 'corkscrew' episcleral veins (Figure 22), raised intraocular pressure, and retinal haemorrhages and ischaemia. CT imaging shows mild enlargement of all extraocular muscles, and tortuous engorgement of the orbital vessels, while orbital Doppler ultrasonography identifies reversal of flow within an 'arterialised' superior ophthalmic vein. In contrast, *high-pressure, low-flow* dural shunts, caused by a spontaneous fistula between a minor dural vessel and the cavernous sinus, are commoner and present with a chronic "red eye" (Figure 23). High pressure, *high-flow* shunts usually require radiological intervention to close the fistula<sup>1</sup> [23]. This contrasts with *low-flow* shunts which typically occlude spontaneously after a few years, although in cases where there is progressive impairment of optic nerve function, or significant congestion in the superior ophthalmic vein and cavernous sinus,

<sup>1</sup>Modern intervention entails highly-selective arteriography and transvenous thrombogenic coiling

occlusion using modern interventional radiology should be considered.



**Figure 21.** Carotido-cavernous sinus fistula (high pressure high flow vascular shunt) leading to proptosis and severe chemosis



**Figure 22.** Dilated episcleral 'cork-screw' vessels seen in high flow orbital vascular shunts, and occasionally mistaken for episcleritis



**Figure 23.** A patient with a high-pressure low-flow dural shunt presenting with proptosis and engorged episcleral vessels, but initially misdiagnosed as having thyroid eye disease

### **Orbital inflammatory disease**

In many cases of orbital inflammation the aetiology remains obscure, despite extensive laboratory tests and histological analysis. Infection, structural causes and thyroid eye disease should be excluded first, and investigations to exclude a systemic cause, such as Wegener's granulomatosis or sarcoidosis, are

indicated. Ancillary tests include a chest X-ray and screening for peripheral markers of systemic disease, including serum angiotensin converting enzyme (sACE), anti-nuclear antibodies (ANA) and anti-nuclear cytoplasmic antibodies (ANCA).

### **Thyroid eye disease**

Thyroid eye disease (TED) is the commonest orbital inflammation and may present before, after, or without the emergence of biochemical evidence for thyroid gland dysfunction [24]. In most patients the disease is mild, being limited to mild ocular surface inflammation, lid retraction, and minimal proptosis. More severe orbital inflammation occurs in a small minority of patients, and can lead to a major expansion of orbital fat and extraocular muscles. In turn, this leads to orbital congestion with a rise in the intra-orbital and intraocular pressures, impaired globe movement, and varying degrees of proptosis. Corneal exposure and ulceration result from severe proptosis and lagophthalmos, and posterior orbital congestion can cause optic neuropathy (Figure 24) [25].



**Figure 24.** Severe thyroid eye disease presenting with right exposure keratopathy and bilateral optic neuropathy

The objectives in managing thyroid eye disease include reducing the risk to vision in the active, or 'wet', phase and correction of residual structural defects once the active inflammatory phase has subsided. In all patients, adequate thyroid control and cessation of smoking are important. In the patient with mild surface inflammation, topical immunosuppression and lubricants may suffice. Systemic immunosuppression (oral or parenteral glucocorticoid) and low-dose radiotherapy are reserved for those with significant soft-tissue inflammation, including those with muscle involvement early in the disease [26]. Orbital decompression during

the active, inflammatory phase is reserved for patients with a congested 'hydraulic' orbit, very severe proptosis, or optic neuropathy unresponsive to high-dose corticosteroid therapy.

Once the active inflammatory phase of TED has resolved, rehabilitative measures aim to redress the effects of residual fibrotic changes. These include orbital decompression<sup>2</sup>, treatment of squint and diplopia, and finally eyelid surgery. The latter can include lowering and/or debulking of the upper lid, or elevation of the lower lid (although decompression tends to reverse lower eyelid displacement).

### Idiopathic orbital inflammatory disease

Idiopathic inflammation may involve specific orbital structures (for example, trochleitis, scleritis, myositis, and dacryoadenitis), or diffusely involve all tissues. Occasionally such chronic inflammation causes severe fibrosis, also known as sclerosing orbital inflammation [27]. Orbital biopsy, to exclude malignancy and other infiltrative diseases, is essential in all cases because the clinical features and orbital imaging may not differentiate the two. Similarly, the diagnosis of 'pseudotumour', frequently managed empirically with a 'trial of steroids', has no place in contemporary clinical management: inflammation due to both malignancy and idiopathic causes will respond to systemic steroids, and such treatment without prior biopsy typically leads to diagnostic uncertainty and delay – potentially with disastrous sequelae for the patient.

Orbital myositis presents with acute orbital ache, ocular redness and diplopia (Figure 25) [28]. Occurring most often in young women, the pain and diplopia are worse on attempted ductions *away* from the field of action of the affected muscle. Imaging identifies diffusely enlarged muscle(s) in the acute phase, with subsequent fibrosis once the inflammatory phase has subsided. The primary management of myositis is with oral non-steroidal anti-inflammatory drugs, which usually lead to a rapid relief of symptoms. In resistant cases, a biopsy should be performed before considering systemic steroids or low-dose orbital radiotherapy.



**Figure 25.** Myositis with MRI identifying enlarged medial rectus

Lacrimal gland inflammation may be due to an acute neutrophilic inflammation, occurring in bacterial infections, or present as a more indolent, predominantly lymphocytic, inflammation. The latter can be related to an underlying systemic disease, such as sarcoidosis or Wegener's granulomatosis [29]. Features of acute dacryoadenitis include a prodromal ache, and a swollen, tender and red upper lid with an 'S'-shaped ptosis; management with oral non-steroidal anti-inflammatory drugs and antibiotics is usually effective. Chronic inflammation of the lacrimal gland usually presents as a relatively painless, non-tender swelling which may be bilateral. CT imaging identifies diffusely enlarged glands which mould around the globe; biopsy and systemic investigations are essential before immunosuppression is initiated.

### Benign orbital tumours

Neurilemmoma, also termed Schwannoma, generally presents in a similar fashion to cavernous haemangioma and has a similar CT appearance (Figure 26) [30]. Solitary neurofibromas, in contrast to plexiform neurofibromas, usually develop as a slowly-progressive mass in the supraorbital nerve territory, leading to proptosis and hypoglobus [31]. Resection of localised tumours should be considered if they cause significant impairment of orbital function. Removal of frontal nerve lesions result in a predictable sensory loss on the forehead, and resection of intraconal lesions carries a small risk of visual loss and motility disturbance. Plexiform neurofibroma, in contrast, diffusely infiltrates the anterior orbital tissues and surgical resection, although not curative, can improve the appearance [32].

<sup>2</sup>Lateral wall decompression frequently suffices, with the medial wall reserved for more significant proptosis. Floor decompression, carrying the risk of hypoglobus and postoperative diplopia, is reserved for severe cases.

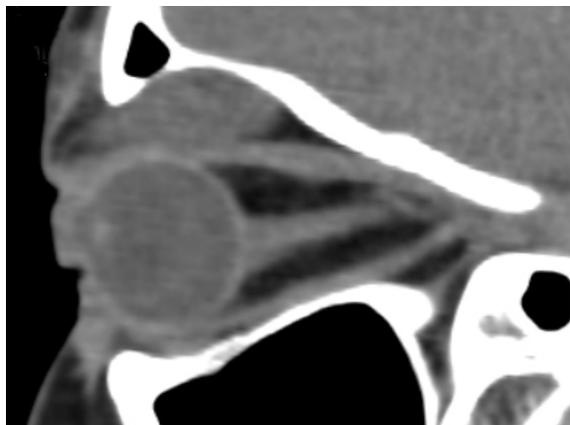


Figure 26. CT showing benign nerve sheath tumour

Sphenoid wing meningiomas usually present in women during the fifth and sixth decade of life [33]. Clinical features include chronic and variable lid swelling, chemosis and mild proptosis. CT imaging identifies hyperostosis of the greater wing of the sphenoid, with *en-plaque* soft-tissue on the lateral wall of the orbit, which may also be present in the temporalis fossa and the middle cranial fossa (Figure 27). Currently, sphenoid wing meningiomas are managed conservatively; since tumour proliferation is stimulated by progestogen, treatments containing this hormone should be avoided.

Pleomorphic adenoma of the lacrimal gland typically presents as a slowly-progressive, often painless, mass that compresses the globe and indents the orbital roof and lacrimal fossa on imaging. It is imperative to recognize these lesions prior to surgery, as a putative diagnosis of pleomorphic adenoma necessitates intact excision of the mass; incomplete or piecemeal resection of an orbital pleomorphic adenoma carries a significant risk of pervasive recurrence – often necessitating orbital exenteration – or a risk of malignant recurrence [34].

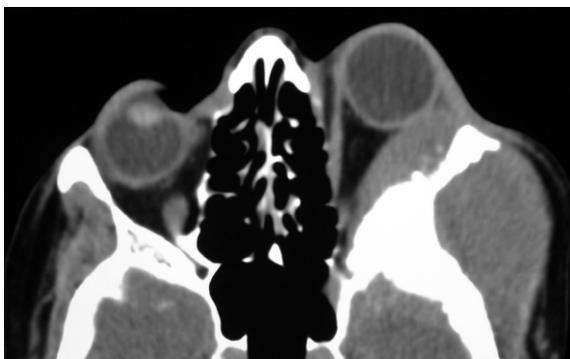


Figure 27. CT of patient with sphenoid wing meningioma identifying hyperostosis and *en-plaque* soft tissue on the lateral wall of the orbit

### **Malignant orbital tumours**

Although primary and secondary orbital malignancy is rare, it can occur at any age, and should always be considered in patients where there is progressive disease, inflammation, or an atypical behaviour for assumed non-malignant disease.

Lymphocytic lesions of the orbit are common, varying from benign “reactive” lymphoid hyperplasia to frankly malignant lymphoma, these typically being B cell in origin [35]. In general, these lesions present as a pink fleshy mass under the conjunctiva, or a painless, soft swelling of the eyelids. Rarely, lymphomas may present with pain, and these tend to progress more rapidly and carry a worse prognosis (Figure 28). CT scan typically shows a moderately well-defined soft-tissue mass moulding around the globe and other orbital structures (Figure 29); imaging is, however, never diagnostic for lymphoma and biopsy is always required. Low-grade lymphomas (for example marginal zone lymphoma) have a low risk of systemic involvement at the time of orbital diagnosis, the converse being true for high-grade lymphomas (for example, diffuse large B-cell lymphoma) [36]. Treatment is dependent on the grade of lymphoma and extent of systemic involvement, and can involve radiotherapy and/or chemotherapy. In principle, intervention is designed to contain low-grade lymphomas and ablate high-grade lymphomas.



Figure 28. Aggressive diffuse large B cell lymphoma of the orbit presenting with inflammatory signs

Finally, a large number of very rare malignancies can involve the orbit, either as primary tumours, such as lacrimal gland carcinomas [37], or as metastatic disease, for example, from breast and prostatic carcinoma. Secondary tumours which spread directly to the orbit may arise in the paranasal sinuses (for



**Figure 29.** CT scan of patient shown in figure 28 showing pervasive lymphoma moulding around the globe

example, carcinomas and aggressive lymphomas (Figure 30), the globe (melanoma or retinoblastoma), or the eyelids (squamous cell, basal cell and sebaceous carcinomas) [38]. Perineural invasion, usually along frontal nerve branches, is commonest with squamous carcinomas arising on the forehead, these causing severe pain without any evidence of an orbital mass [39]. Finally, adenoidcystic carcinoma of the lacrimal gland has a propensity to perineural invasion that carries a dismal prognosis.



**Figure 30.** CT of patient with aggressive sinus lymphoma invading the orbital structures

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