

Disorders of the orbit

Introduction

Applied anatomy

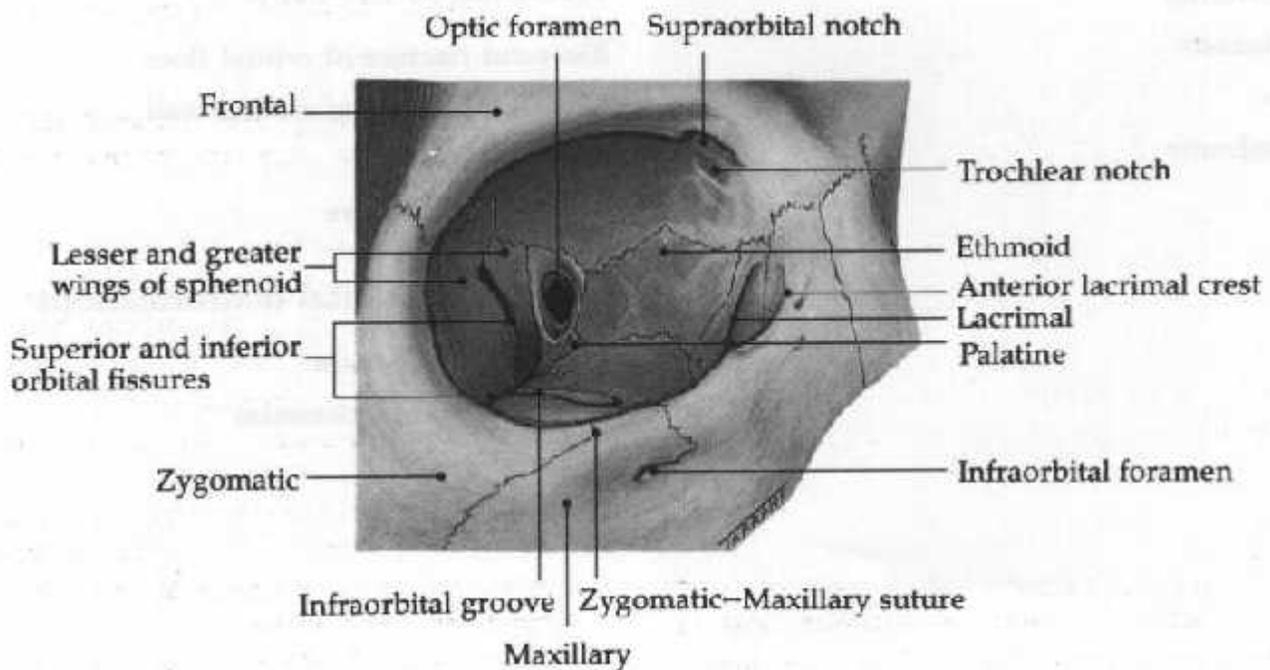
The orbit is a pear shaped cavity whose stalk is the optic canal .

The roof consists of **two** bones: the lesser wing of the sphenoid and frontal. It is located adjacent to the anterior cranial fossa and frontal sinus.

The lateral wall also consists of **two** bones: the greater wing of the sphenoid and zygomatic. The anterior half of the globe is vulnerable to lateral trauma because the lateral wall protects only the posterior half of the globe.

The floor consists of **three** bones: zygomatic, maxillary and palatine. The posteromedial portion of the maxillary bone is relatively weak and may be involved in a 'blow-out' fracture.

The medial wall consists of **four** bones: maxillary, lacrimal, ethmoid and sphenoid.



Dysthyroid ophthalmopathy

Association with thyroid dysfunction

Graves' disease is an autoimmune disorder caused by excess secretion of thyroid hormones by the entire thyroid gland. It most commonly presents in the fourth to fifth decades of life and affects women more frequently than man by an 8:1 ratio. It is the most common cause of thyrotoxicosis.

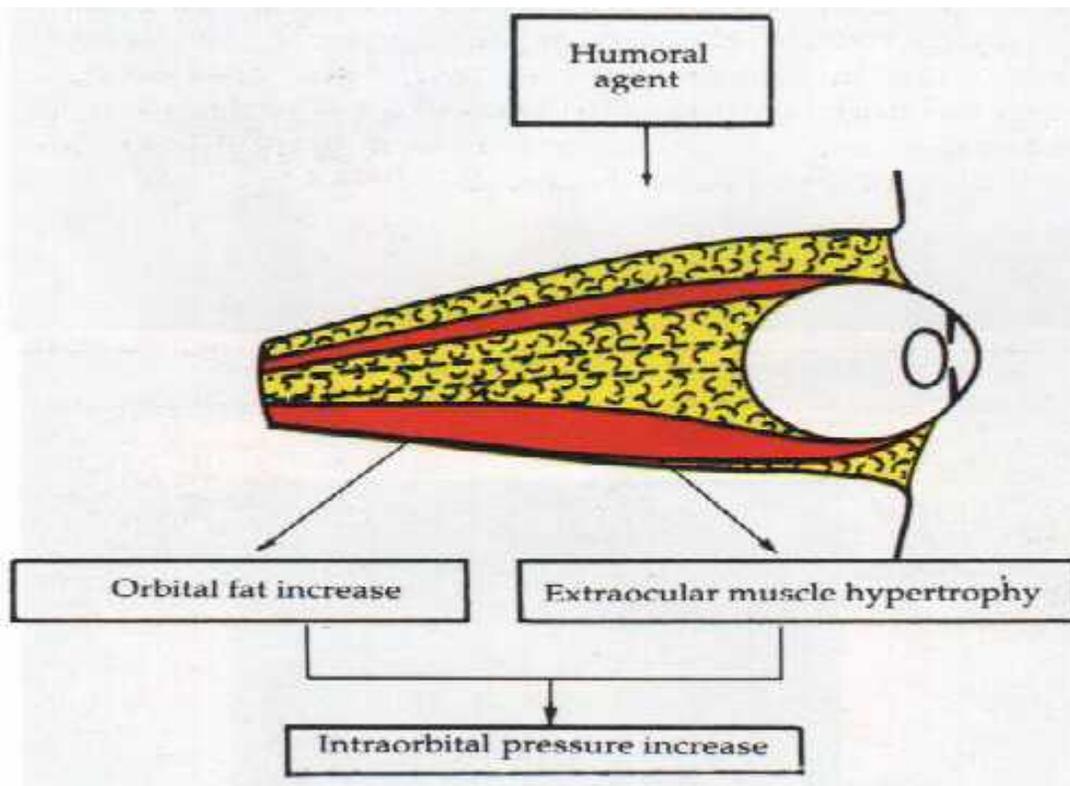
In 10-25% of cases, thyroid ophthalmopathy occurs in the absence of both clinical and biochemical evidence of thyroid dysfunction.

Pathogenesis

Dysthyroid ophthalmopathy appears to be an organ-specific autoimmune disorder in which a humoral agent (IgG antibody) is believed to be responsible for the following changes:

1. **Hypertrophy of extraocular muscles** caused mainly by an increase in glycosaminoglycans.
2. **Cellular infiltration** of interstitial tissues with lymphocytes, plasma cells, macrophages and mast cells occurs in the congestive stage. Subsequent degeneration of muscle fibres eventually leads to fibrosis resulting in restrictive myopathy and diplopia:
3. **Proliferation** of orbital fat, connective tissue and lacrimal glands occurs with retention of fluid and accumulation of glycosaminoglycans.

The above factors cause an increase in intraorbital contents and secondary elevation of intraorbital pressure.



Clinical features

There are two stages in the disease development of the disease:

1. The stage of active inflammation in which the eyes are red and painful.
2. The quiescent stage in which the eyes are white, although a painless motility defect is present.

1- Eyelid retraction

Retraction of both the upper and lower eyelids occurs in about 50% of patients with Graves' disease and is responsible for functional and cosmetic problems.

CLINICAL FEATURES

The margin of the upperlid normally rests about 2 mm below the limbus. Retraction is suspected when the lid margin is either level with or above the superior limbus, allowing sclera to be visible . Likewise, the lower eyelid normally rests at the inferior limbus and retraction is suspected when sclera shows above the lid.

MANAGEMENT OF LID RETRACTION

Many patients with mild lid retraction do not require treatment and, in about 50% of cases, it improves spontaneously. Treatment of associated hyperthyroidism may also improve lid retraction. Surgery to decrease the vertical lid fissures should be considered in patients with significant but stable lid retraction

2-Soft tissue involvement CLINICAL

FEATURES

1. **Conjunctival injection** is a sensitive sign of disease activity. 2. **Chemosis** refers to oedema of the conjunctiva and caruncle. 3. **Oedema and fullness of the eyelids** 4. **Superior limbic keratoconjunctivitis**

The above signs are usually associated with variable lacrimation, photophobia, grittiness and retrobulbar discomfort

MANAGEMENT

1. **Topical therapy** with lubricants. Artificial tears can be used during the day and ointment at bedtime. Patients with superior limbic keratoconjunctivitis may require topical adrenaline and acetylcysteine.
2. **Head elevation** through use of three pillows during sleep may be useful in reducing periorbital oedema. Taping of the eyelids during sleep may be useful in patients with mild exposure keratopathy.

3. **Diuretics** given at night may reduce the morning accumulation of periorbital oedema.

3- Proptosis

The proptosis in thyroid ophthalmopathy is typically axial. Thyroid ophthalmopathy is the most common cause of both bilateral and unilateral proptosis in adults. The proptosis is uninfluenced by treatment of hyperthyroidism and it is permanent in about 70% of patients. Severe proptosis prevents adequate lid closure and, unless treated, may lead to severe exposure keratopathy, corneal ulceration and endophthalmitis.

MANAGEMENT *Systemic*

steroids

Systemic steroids may be used in patients with rapidly progressive and painful proptosis during the early course of the disease provided there are no contraindications such as tuberculosis or peptic ulceration:

1. **Oral prednisolone**

2. **Intravenous methylprednisolone** (0.5 g in 200 ml isotonic saline over 30 min), which can be repeated after 48 hours, may also be effective but requires careful monitoring because of potential cardiovascular risks.

Radiotherapy

Radiotherapy may be an alternative to systemic steroid therapy in patients who have contraindications to systemic steroids or are unresponsive to steroids despite an **adequate** dose.

Surgery

Surgical decompression may be considered either as primary treatment or when noninvasive methods are ineffective. Surgery may also be considered when non-invasive methods are inappropriate such as during the inactive phase of the disease in patients with cosmetically unacceptable proptosis.

4- Dysthyroid optic neuropathy

CLINICAL FEATURES

Symptoms

The main symptom of optic nerve involvement is a slowly progressive impairment of central vision associated with defective red-green colour appreciation. In order to detect early involvement, patients should be advised to monitor their own vision at home by occluding each eye alternately and reading small print, as well as by assessing the intensity of colours on a television screen.

Signs

1. **Ophthalmoscopy** may be normal or it may show disc oedema and chorioretinal folds. Optic atrophy may be present in advanced cases.

2. **Visual field defects**

3. **An afferent pupillary conduction defect**

MANAGEMENT

The management of dysthyroid optic neuropathy is essentially similar to that of severe proptosis.

5- Restrictive thyroid myopathy

Between 30% and 50% of hyperthyroid patients develop ophthalmoplegia. The diplopia may be transient but, in 50% of patients, it is permanent. Ocular motility is restricted by oedema during the infiltrative phase and by fibrosis during the fibrotic phase..

CLINICAL FEATURES

In order of frequency the four ocular motility defects are:

1. Defective **elevation** caused by a fibrotic contraction of the inferior rectus muscle..
2. Defective **abduction** caused by fibrotic contraction of the medial rectus
3. Defective **depression** caused by involvement of the superior rectus.
4. Defective **adduction** caused by involvement of the lateral rectus which is the least common.

MANAGEMENT

Indications for surgery are diplopia in the primary or reading positions of gaze or in both. In addition the angle of deviation must be stable for at least 6 months, and at the time of surgery there must also be no evidence of congestive ophthalmopathy which is indicative of active disease. Until these criteria are met the diplopia should be alleviated, if possible, with prisms.

The goals of surgery are to achieve binocular single vision in the primary position of gaze and when reading..

Orbital infections

Preseptal cellulitis

Preseptal cellulitis typically affects children and is usually secondary to lid infection such as severe acute hordeolum, skin laceration or an insect bite. The infection does not penetrate the orbital septum which separates the anterior structures from the orbit.

Examination shows periorbital swelling and tenderness without proptosis Ocular motility, visual acuity and pupillary reactions are all normal.

Treatment is with oral antibiotics on an outpatient basis.

Bacterial orbital cellulitis

Bacterial orbital cellulitis is an infection of the soft tissues behind the orbital septum. It is much less common but potentially more serious than preseptal cellulitis. The following are the main types:

1. **Sinus-related** is by far the most common and is most frequently secondary to ethmoidal sinusitis. It typically affects children and young adults.
2. **From adjacent structures** such as dacryocystitis, mid-facial infection or dental infection.
3. **Post-traumatic** most commonly develops within 48-72 hours of an injury that penetrates the orbital septum.
4. **Post-surgical** may complicate retinal detachment surgery, strabismus surgery, dacryocystorhinostomy and orbital surgery.

Polymicrobial infection, which may include anaerobic bacteria, is the rule. The most common causative

organisms are *Streptococcus pneumoniae*, *Staphylococcus aureus* and *Streptococcus pyogenes*. In children under the age of 5 years, the offending organism is frequently *Haemophilus influenzae*.

CLINICAL FEATURES

Presentation is with a rapid onset of unilateral chemosis, proptosis and painful diplopia.

Examination shows an unwell and pyrexial patient. The proptosis is most frequently lateral and downwards. The eyelids are swollen, erythematous, warm and tender to palpation. Ocular movements are restricted and painful. In advanced cases visual acuity may be diminished and there may be an afferent pupillary conduction defect.

POTENTIAL COMPLICATIONS

1. Intracranial complications, include meningitis, brain abscess and cavernous sinus thrombosis. The last is a rare but extremely serious complication which should be suspected when there is evidence of bilateral involvement, rapidly progressive proptosis, and congestion of the facial, conjunctival and retinal veins. Additional features include an abrupt progression of all clinical signs associated with prostration, severe headache, nausea and vomiting.

2. Subperiosteal abscess is most frequently located along the medial wall of the orbit. 3. Ocular complications are exposure keratopathy, raised intraocular pressure, occlusion of the central retinal artery or vein and contiguous inflammation of the optic nerve.

MANAGEMENT

Orbital cellulitis is an emergency requiring hospital admission. The patient should be evaluated by an ophthalmologist and an otolaryngologist. Occasionally, a neurosurgeon is required for intracranial abscess drainage. Because orbital cellulitis is potentially vision-threatening, and occasionally life-threatening, the patient should be re-evaluated at frequent intervals. Optic nerve function should be monitored every 4 hours by testing pupillary reactions, measuring visual acuity and assessing colour vision.

Investigations

Investigations, where appropriate, include the following:

1. White cell count.
2. CT of the orbit, sinuses and brain. Sinus radiographs are obtained only if CT is not available.
3. Blood and nasal cultures may be taken but are unlikely to have a positive yield. Conjunctival and oropharyngeal cultures are invariably unhelpful.
4. Lumbar puncture is indicated if meningeal or cerebral signs develop.

Antibiotic therapy

In children under the age of 5 Years, the antibiotics should cover *Haemophilus influenzae*. The usual combination is ampicillin 200 mg/kg per day in divided doses and a penicillinase-resistant penicillin (100 mg/kg per day), administered parenterally.

In adults antibiotic therapy is with parenteral broad-spectrum therapy with third or fourth generation cephalosporins and metronidazole to cover anaerobes. In the case of previous anaphylactic reaction to penicillin or its derivatives, the patient should be treated with clindamycin or vancomycin. Antibiotic therapy should be continued until the patient has been fever-free for 4 days.

Surgery

Surgical intervention should be considered in the following circumstances: unresponsiveness to antibiotics, decreasing vision, orbital abscess, subperiosteal abscess and the need for a diagnostic biopsy in atypical cases. In most cases, it is necessary to drain the orbit as well as the infected sinuses.