

Thyroid eye disease

Eye series – 10

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A 50 year old woman presents complaining of intermittent dry eye and puffiness under her lids, particularly in the morning. The patient has also noticed that her eyes have an increased 'starey' look to them (Figure 1). About 10 months ago she experienced irritability, fatigue, increased appetite, weight loss, palpitations, heat intolerance and difficulty sleeping. She was diagnosed with Graves disease (GD) and has been on neomercazole.

Question 1

What is the mechanism of the eye problems in GD?

Question 2

Discuss the common ocular manifestations of thyroid disease.

Question 3

Name the differential diagnoses.

Question 4

What is the long term prognosis for both general appearance and vision?

Question 5

Discuss the available treatment options.

Question 6

When is surgery indicated?

Answers

Answer 1

Graves disease (GD) is an autoimmune disease that involves both orbital tissues and the thyroid gland. It is also referred to as thyroid eye disease (TED) or thyroid orbitopathy. It is not caused by an overactive thyroid gland. Immune cells 'attack' both the eye muscles and the thyroid leading to dysfunction of both. Although most patients with GD have an overactive thyroid, patients with normal and even subnormal thyroid function may also have GD. It is important to realise that destructive procedures of the thyroid (eg. radioactive iodine) and even surgical removal of the thyroid, do not alter the course of the orbital (eye) disease.

Answer 2

Thyroid eye disease is the commonest cause of proptosis (protruding eye) in adults. Women are affected nine times more than men. Most patients present between 20–45 years of age. One or both eyes may be proptotic (protruding) and the eyelids retracted, creating a staring



Figure 1. Patient with Graves disease



Figure 2. CT scan of orbit showing swelling of extraocular muscles

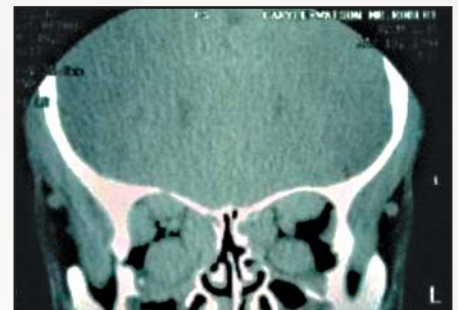


Figure 3. CT scan showing compression of orbit

appearance. The upper eyelids tend to lag behind the eye as the patient looks down. Because of the greater exposure of the eye, dry eye symptoms (eg. burning, stinging, etc.) may be present. There is often redness of the eyes and swelling of the ocular surface (conjunctiva). Because the eye (extraocular) muscles enlarge in this condition, double vision may occur due to limited motility of the eye (Figure 2). Also, because there is inflammation of orbital (eye socket) fat and enlargement of the eye muscles, there may be com-

pression of the optic nerve resulting in optic neuropathy (Figure 3). This latter condition may lead to permanent visual loss. A further manifestation of the disease is puffiness of the lids, which usually appears worse in the morning.

Answer 3

Thyroid eye disease represents the commonest cause of both unilateral and bilateral proptosis. Differential diagnosis includes:

- orbital cellulitis (usually rapid onset and associated with fever)
- orbital tumours and metastases
- orbital pseudotumour (an idiopathic, nonspecific inflammatory orbital condition that is usually painful, progresses quickly compared to TED and commonly causes ptosis rather than the classic lid retraction of GD. It is almost always unilateral in presentation)
- carotid cavernous fistulas (patients present with proptosis, chemosis, diplopia and visual loss usually associated with a bruit)
- lesions or vascular accidents of the mid brain (can lead to restriction or paralysis of upgaze and pupil defects). CT and MRI scans can help lead to differentiation of these conditions.

Thyroid eye disease can also appear concurrently with other autoimmune conditions such as myasthenia gravis. This may confuse ocular symptoms and may require further investigation by a specialist.

Answer 4

Thyroid eye disease is a self limited process. The condition has two main phases. The first stage is the acute stage characterised by the onset of the inflammation and subsequent ocular symptoms. This is followed by a more stable quiescent phase. Although inflammation has usually subsided by this stage, fibrosis of the tissue can occur. The process can last between 12–24 months. If the eyes are moderately affected, it is possible they

may return to normal. If inflammation has been severe, it is unlikely that changes will completely disappear as the initial swelling has turned into scarring. Surgery may be indicated for cosmetic purposes. With appropriate treatment it is rare that vision is severely affected.

Answer 5

Although most patients with TED are hyperthyroid, medication to return thyroid function to normal has not been shown to affect the course of TED. Medical treatment may be employed to reduce the possibility of long term damage. Steroid therapy is occasionally used to reduce swelling during the active phase. This has limited use during the later stage of TED. As the process is self limiting, most treatment is aimed at reducing various symptoms of the condition. Artificial tears and ointments can provide relief from irritable, red eyes. Nonpreservative tears should be used during the day with ointment at night to provide greater protection for the corneal surface. Sleeping with the head elevated may help reduce lid swelling or puffiness. Diuretics may also help to improve the appearance by reducing swelling. If the patient has noticed double vision, prisms can be added to glasses to help maintain a single image. If the condition is deteriorating, more aggressive treatment may be required (eg. steroid therapy). Only when the condition has stabilised should surgery be indicated to realign the eyes. Visual field loss or decrease in colour vision may indicate optic nerve compression. Immediate referral to an ophthalmologist is required. Several factors can contribute to a deterioration of the condition and therefore should be avoided. If hyperthyroidism is over treated and becomes hypothyroid the ophthalmopathy can deteriorate. Several studies have confirmed a link between the progression of the disease and smoking. Smoking is associated with a more prolonged and severe course of GD. Reducing or ceasing smoking may help

decrease the risk of congestive orbitopathy and prevent further change.

Answer 6

Unless optic nerve compression or severe corneal disease has occurred, surgery should be delayed until the later quiescent phase of TED. In cases of optic nerve compression, if no response has been noted within 24–48 hours of steroid treatment, surgery is indicated. Orbital decompression surgery may serve to relieve the pressure caused by swollen tissue. During surgery at least two orbital walls are decompressed. As the muscles more commonly affected are the inferior and medial rectus muscles, the medial wall and orbital floor are typically chosen and provide the greatest release of pressure. Possible side effects of surgery include postoperative double vision and further lid retraction. Decompressing the lateral wall can help the appearance of proptosis. Persistent double vision as a result of strabismus can be corrected once the condition has stabilised. The main goal of strabismus surgery is to increase single vision in primary and reading positions. Postsurgery, the patient may still notice some diplopia in extreme gaze. Although some of the effects of TED will diminish over time, some changes will persist. Simple measures such as growing a fringe or wearing tinted glasses may help camouflage these changes, however, cosmetic surgery may be indicated to 'normalise' the patient's appearance. Preoperative counselling should be undertaken to ensure realistic expectations of surgery. Lid lengthening procedures can improve existing lid retraction and may help reduce corneal exposure. Blepharoplasty of both upper and lower lids to reduce eye puffiness can also help in some circumstances.

Conflict of interest: none declared.

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