Thyroid orbitopathy in a young woman

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A well co-ordinated and compassionate approach from the family physician, endocrinologist and ophthalmologist is essential in the management of thyroid orbitopathy.

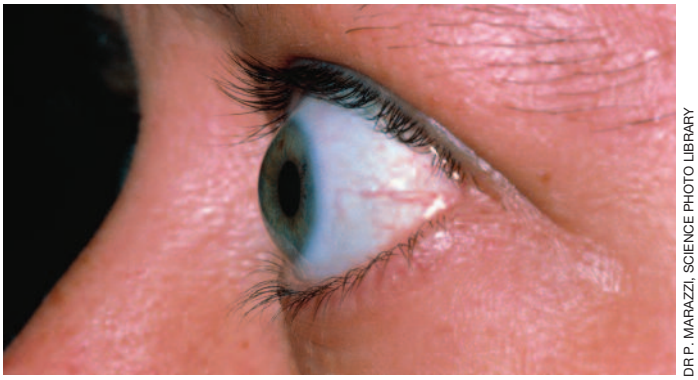

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Figure. Exophthalmos and tissue swelling in thyrotoxicosis. The changes are bilateral, and were highly symmetrical in the case described below.

Case presentation

A 36-year-old teacher presented who developed hyperthyroidism two years ago. She was treated with oral medication, which brought her symptoms under control. However, this relief was rudely disturbed three months ago when she developed swollen red eyes and eyelids. She was referred for ophthalmic evaluation at this stage.

Two weeks before her initial eye consultation, the patient noticed double vision while looking up at the surtitles at the opera. Her self-confidence was being eroded by the change in her facial appearance, and she felt that her job performance was suffering as a result. Her eyes were also dry and gritty, especially in the mornings.

Comment

The terms thyroid eye disease, Graves' eye disease, thyroid-related orbitopathy and thyroid orbitopathy all refer to the same unpleasant clinical entity, but the last two provide the most accurate description. The condition, in which the tissues around the eyeball become congested and swollen, occurs in patients who have a form of thyroid dysfunction, which may be subclinical. The orbitopathy can occur many years before or after the thyroid trouble.

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Thyroid orbitopathy can affect people of all ages but is more common in those in their 30s and 40s. Women are affected more often than men. It can have a disastrous effect on quality of life – the condition is disfiguring, sometimes painful (particularly if corneal drying and ulceration occur), and potentially blinding.

Examination

This patient had the typical stare of thyroid orbitopathy and the symmetry with which her eyes were affected left little doubt as to the diagnosis. Her visual acuity was normal (6/6 bilaterally), but she had diplopia in upgaze, with the left eye being unable to elevate beyond 35°. The diplopia was experienced only in a fairly extreme position and did not interfere with her daily activities. She had sclera showing between the upper lid margin and the limbus (superior scleral show) in both eyes (2 mm in the left and 1 mm in the right).

Pupil reactions were normal, and a slit lamp examination showed only mild redness and oedema of the conjunctiva on both sides, as well as mild punctate fluorescein staining of the cornea. Her colour vision was normal, and a fundus examination showed healthy optic discs. A computer visual field test was normal.

Comment

The changes in thyroid orbitopathy are bilateral. These changes may be highly asymmetrical, which makes the diagnosis more difficult, but reasonable symmetry is more common. The most obvious feature is prominence of the eyes, which is usually due to true proptosis (exophthalmos) but sometimes to eyelid retraction. Both the upper and lower eyelids can be retracted – this can be quantified by measuring the scleral show. The upper lid may exhibit lid lag, which is best demonstrated by asking the patient to follow a target such as the examiner's fingertip moving slowly downwards. Lid lag is sometimes present without retraction.

Exophthalmos and lid retraction lead to increased drying of the ocular surface because the exposed area is increased. This can result in exposure keratopathy and corneal ulceration, which in severe cases can lead to visual loss. The surface can be directly affected by the inflammatory elements of thyroid orbitopathy, with swelling of the conjunctiva (chemosis) and hyperaemia.

Eye movement can be restricted by changes in the extraocular muscles. Vertical movement, especially upgaze, is most commonly affected, but excursion in all directions can be reduced. Restriction in upward rolling of the eyes that normally occurs in eye closure can exacerbate exposure keratopathy. Orbital changes and congestion lead to proptosis and puffiness of the eyelids. Sometimes the lower lid swelling can be so severe that it creates a bag-like effect which hangs below the level of the orbital rim in festoons.

The most serious complication of orbital congestion is compressive optic neuropathy. This can result in visual loss, but the following may suggest less severe compression:

- colour desaturation, especially red (bright red objects appear slightly pinker)
- visual field defects (which are sometimes subtle and detected only by computerised or Goldmann perimetry)
- a relative afferent pupil defect (Marcus Gunn pupil)
- swelling of the optic disc (a sign of quite serious compression).

The relative afferent pupil defect is caused by a difference in the responses of the two optic nerves to the same light stimulus. It can be demonstrated by swinging the stimulus from one eye to the other – the pupil of the weaker eye will dilate slightly as the light source moves away from the better eye and towards it because the consensual pupillary light reaction is stronger than the direct one.

Investigations

The patient's thyroid status was monitored by her family physician and endocrinologist, and the computer visual field test mentioned earlier was performed to look for subtle evidence of compressive optic neuropathy. Her clinical presentation was entirely typical of thyroid orbitopathy, so further diagnostic tests were not required at that time.

Comment

Thyroid orbitopathy is essentially a clinical diagnosis. However, if a patient presents with eye findings that are not entirely typical or if a thorough history and endocrine evaluation do not reveal any thyroid dysfunction then it may be necessary to look for diagnostic findings on imaging, although the presence of anti-TSH receptor antibodies would be supportive.

CT scanning of the orbits is the chief investigation used to diagnose and evaluate thyroid orbitopathy. The hallmark finding is extraocular muscle enlargement with sparing of the tendons. Proptosis is readily shown on axial scans, and the absence of other intraorbital pathology such as a space occupying lesion is important in borderline or highly asymmetrical cases. The lacrimal glands may be mildly enlarged. In cases of marked crowding of the apical portion of the orbit, the normal fat plane around the optic nerve can appear obliterated by the closeness of the muscles to the optic nerve, raising suspicions of compressive optic neuropathy.

Management

In this case, no immediately sight-threatening complications were present. Attention was focused on educating her about the natural course of the active disease and therapeutic options.

The patient was assisted in an attempt to quit smoking by her family physician, and she found that regular use of artificial tears increased her stamina in visual tasks such as computer work. She purchased a pair of lightly tinted sunglasses that helped to hide her deformity, and she became accustomed to sleeping on two to three pillows. Definitive surgical repairs could not be performed at this stage (see 'Comment' below).

The patient remains stable. She elected to have botulinum toxin (Botox) injected into her upper lids as a temporising measure for her upper lid retraction; this improved her appearance as well as her exposure symptoms. She is not yet out of danger of visually threatening compressive optic neuropathy, and she will remain under review for many months to come.

Comment

General supportive measures are important. Artificial tears or gel, generally used at least four times a day and in conjunction with ointment at bedtime, are a starting point, but much higher dosages are sometimes required. Surgical temporising measures such as tarsorrhaphies or lacrimal punctal occlusion are occasionally needed. Sleeping with the head elevated can alleviate the worsening of congestive symptoms in the mornings. Prisms incorporated into spectacles can alleviate diplopia.

Controlling hyperthyroidism (if present) can improve ocular symptoms, even though it does not directly affect tissue changes in the orbit. For unknown reasons, hyperthyroid patients treated with radioactive iodine have increased risk of thyroid orbitopathy compared with those treated with thyroid surgery or other medical agents. Cigarette smoking has been shown definitely to worsen thyroid orbitopathy – an attempt to quit must be recommended.

The active stage of thyroid orbitopathy, in which there are signs of hyperaemia and more marked congestion, can last up to around two years, and physical changes in the orbits can occur during this time. It is therefore important to reserve surgical correction until the signs remain stable for several months – unless an urgent indication such as compressive optic neuropathy arises. Oral corticosteroids have a role in the active phase of the disease, but close monitoring for adverse effects is required. More recently, botulinum toxin has been used with some success to weaken the upper lid retractor (the average duration of action is three months).

Traditionally, radiotherapy to the orbit has been considered an adjunct to medical and surgical treatment, but some recent evidence suggests that it may not be as effective as previously thought. A recent controlled clinical trial at the Mayo Clinic showed no benefit of radiotherapy over placebo in the group of patients with moderate thyroid orbitopathy.

For stable problems suited to surgical correction, deeper repairs are conducted before superficial ones. First, orbital bony decompression is performed to create more room for the expanded tissues (if necessary), followed by any required extraocular muscle surgery for eye movement restriction and misalignment, and finally any procedures needed on the eyelids to correct lid retraction and puffiness. Of course, patients with milder disease will need only superficial repairs, if any. MT