Recurrent proptosis and thyroid eye disease

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A 58-year-old Caucasian woman presented to our hospital with a 2-week history of diplopia and proptosis of the left eye. Ten years earlier, she had been diagnosed as having ophthalmic Graves' disease and subsequently underwent successful left transantral orbital decompression for optic nerve compression. On clinical examination, there was bilateral proptosis, 5 mm greater on the left side, and hypoglobus of the left eye. Her visual acuities and colour vision were normal (Snellen visual acuity: left 6/6, right 6/6). There was significant restriction of ocular movements in the left eye. Routine haematological tests were performed, including thyroid function, which were all normal. A computed tomographic (CT) scan of the orbits was performed (figures 1 to 5).

Figure 1 CT scan of the orbits

Figure 4 CT scan of the orbits

Figure 2 CT scan of the orbits

Figure 5 CT scan of the orbits

Figure 3 CT scan of the orbits

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Questions

- 1 What are the differential diagnoses of unilateral proptosis in this case?
- 2 What are the symptoms of orbital disease?
- 3 What are the causes of visual impairment in orbital disease?
- 4 What do the CT scans show?
- 5 How common is recurrent proptosis after orbital decompression in thyroid eye disease?

Answers

QUESTION 1

The most likely causes are mucocoele or recurrent thyroid eye which are specific to this case. Other causes of proptosis include axial causes (orbital varices, carotico-cavernous fistulas, optic nerve gliomas, cavernous haemangioma, meningiomas and orbital pseudotumours, eg, lymphoma) and non-axial causes (dermoid cysts in children or young adults, neurofibromatosis in children, lacrimal tumours, mucocoele or carcinomas of sinuses (ethmoidal and frontal), meningoencephalocele, and rhabdomyosarcomas in children).

QUESTION 2

The symptoms of orbital disease are proptosis, globe displacement, lid retraction, diplopia, visual impairment, pain, reduced sensation of the ophthalmic and maxillary branches of the trigeminal nerve and bruit.

QUESTION 3

The causes of visual impairment in orbital disease are optic nerve compression, corneal exposure, pseudohypermotropia, visual field loss, and choroidal folds.

QUESTION 4

The CT scans demonstrate, on the left side, a mucocoele extending from the ethmoidal sinus through the bony defect formed by previous decompression, into the orbit causing exophthalmos (figure 1) and hypoglobus (figure 2). The mucocoele extended up to the frontal sinus (figure 3). The involvement of ethmoidal sinus (figure 4) and frontal sinus (figure 5) is extensive.

QUESTION 5

Recurrent proptosis after orbital decompression in patients with Graves' disease is extremely rare and is a diagnosis of exclusion (see Discussion).

Outcome

The patient was referred to an ear, nose and throat surgeon for further management with a view to removing the mucocoele endoscopically. The patient declined and the mucocoele has remained stable and has not increased in size to date.

Discussion

Orbital decompression is a well recognised treatment for severe dysthyroid eye disease. Its main indications are optic nerve compression, severe corneal exposure and cosmesis. Removal of the orbital wall during decompression allows excessive orbital fat to herniate through the opening thus relieving the intra-orbital pressure. Long-term results of decompression are encouraging and few complications are seen which include diplopia, meningitis, sinusitis and cerebrospinal fluid leak.¹⁻³

Graves' orbitopathy is thought to be a disease with a self-limiting time course of 1 to

5 months.⁵ True recurrence of orbitopathy has not been reported in the literature. The closest report of a 'recurrence' is the enlargement of extra-ocular muscle after orbital decompression.⁴ Any 'recurrence' should be treated with suspicion. In this case, a mucocoele originated from the frontoethmoidal sinus extended into the orbit through an iatrogenic opening following a transantral orbital decompression. To our knowledge this has not been previously reported.

Patients with frontoethmoidal mucocoele are often asymptomatic. Some of the presenting symptoms include sinusalgia, facial heaviness, epiphora (from lacrimal drainage impairment), swelling of medial orbitofrontal region, globe displacement, diplopia, infection of the mucocoele, and rarely nasal obstruction and rhinorrhea. The classical sign is 'egg shell cracking' or fluctuation on palpation over the frontal sinus due to thinning of bone over the mucocoele. Other signs include discharging fistula and proptosis. In terms of investigations, X-ray may show an overall loss of translucence of the affected side and a loss of the scalloped bony margin if the vertical portion of the frontal sinus is affected. CT scan may show the accompanying bone changes and the affected area often has a homogenous appearance which is isodense to the brain tissue. Magnetic resonance imaging is specific and shows a high signal obtained on the T₂-weighted spin-echo sequences (except in old lesions where these signals become dark) and a low signal on inversion recovery. Treatment for all forms of mucocoele is surgical. Current practice favours endoscopic surgical marsupialisation.⁶ In some cases of frontal mucocoele, an external approach (external frontoethmoidectomy) is required with an osteoplastic flap procedure or even sometimes bone grafts, depending on the degree of sinus expansion and bone destruction.⁷ The aim is to leave the sinus intact, remove the mucocoele lining but avoid stripping too much mucosa and thereby allowing the reconstitution of a well-drained sinus cavity. Occasionally, to improve drainage, the intersinus septum is perforated to unite the two frontal sinuses or the sinus is canalised with a fenestrated Silastic tube into the nasal cavity. When there is mucosal degeneration or the posterior wall of the frontal sinus is dehiscent on the dura, sinus obliteration or sinus cranialisation should be used.

In the light of this case, we recommend that all patients with a recurrence of orbital symptoms following orbital decompression in Graves' orbitopathy should undergo thorough investigation, including a repeat CT scan, to exclude such complications.

Final diagnosis

Frontoethmoidal mucocoele following orbital decompression.

Keywords: Graves' orbitopathy; orbital decompression; frontoethmoidal mucocoele

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A man presenting with limb weakness and electrolyte imbalance

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A 39-year-old man presented with lower limb muscle pains and weakness which had progressed rapidly over the preceding 2 weeks. Immediately prior to hospital admission he was unable to stand unaided. There had been no disturbance of bulbar, sensory or sphincter function and he reported no recent gastrointestinal symptoms. Medical history included colitis, which was quiescent. His only regular medication was olsalazine. On examination, he was normotensive (and remained so throughout his admission). There was no rash. Cardiovascular, respiratory and abdominal examinations were normal. Neurological examination revealed generalised weakness in all four limbs, most marked proximally. Tendon reflexes were preserved. Plantar response was flexor. Examination of cranial nerves was normal.

Laboratory investigations revealed a serum potassium of 1.8 mmol/l. Serum sodium, urea and creatinine were within normal limits. Creatine kinase was 10 719 IU/l. A quadriceps muscle biopsy was performed and the histology is shown in figure 1. Further investigations included an abdominal ultrasound scan, following which computed tomography (CT) of the abdomen was performed (figure 2). A 24-hour urine collection for catecholamines was undertaken (table).

Figure 1 Quadriceps muscle biopsy

Figure 2 CT scan

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Table Levels of free catecholamines (laboratory normal in brackets)

Catecholamine	
Noradrenaline (nmol/24 h)	3328 (<800)
Adrenaline (nmol/24 h)	725 (<100)
Dopamine (nmol/24 h)	>45 000 (<3100)

Questions

- 1 What does the CT scan show?
- 2 What is the likely underlying diagnosis?
- 3 What are the three most common presenting symptoms for this condition?
- 4 What caused the muscular signs and symptoms?