Bilateral sudden visual loss due to sphenoid mucocele in Albright's syndrome

EDITOR—Fibrous dysplasia, the replacement of bone by abnormal fibrous tissue, may occur in association with endocrine abnormalities and cutaneous pigmented patches as Albright's syndrome or without such manifestations as isolated fibrous dysplasia. With any involvement, visual loss may be caused by compression of the optic nerves or chiasm, by dysplastic bone or haemorrhage into dysplastic bone. We report the first reported case of visual loss due to sphenoid mucocele in Albright's syndrome.

CASE REPORT
Three weeks before presentation a 20-year-old woman experienced pain around the right eye followed by deterioration of vision in the right eye. A week later, painless visual loss occurred in the left eye. Direct questioning elicited a history of spontaneous right sided long bone fractures and precocious puberty. Albright's syndrome had been diagnosed at the age of 8 years.

On examination, she was of short stature and had a flat pigmented lesion on her buttock; no other abnormality was found. The vision in the right eye was perception of light and in the left eye, counting fingers. Using a 30/1000 white target the visual field in the right eye was unrecordable and in the left eye was restricted to an island of 15°. There was minimal temporal pallor of both
optic discs but the nerve fibre layer appeared normal and the discs were flat (Fig 1). Serum haematology and biochemistry were normal with the exception of a raised alkaline phosphatase.

Computerised tomography (Fig 2) showed a midline cystic tumour with irregular edges and fluid levels which involved sphenoid, ethmoid, and maxillae. This was felt to be consistent with fibrous dysplasia and sphenoid mucocele.

Surgical decompression was undertaken the same day. A large cavity formed by the expanded sphenoid was entered via a transethmoidal approach, and an ostium fashioned from it into the nasal cavity. Histological examination of the wall of the cavity showed no evidence of malignancy and was characteristic of mucocele.

Ten days after surgery, her vision improved to 3/60 right, 6/12 left, with a centrocaecal scotoma in the right eye and a paracentral scotoma in the left eye. One month after surgery her vision was 6/9 in each eye with small paracentral scotomas, and the appearance of the optic discs was unaltered. One year after surgery, vision was 6/9 in each eye.

COMMENT

Sphenoid mucocele may produce clinical syndromes by compression of the optic nerves, chiasm, oculomotor nerves, or pituitary gland. Pain, visual loss, and diplopia are the commonest symptoms.6 Differential diagnoses include pituitary tumour, cranio-ophthalmic dysplasia, meningioma, sinus neoplasm, clivus chordoma, and internal carotid aneurysm. Computerised tomography shows a homogenous isodense or hyperdense sellar or parasellar lesion, and magnetic resonance imaging a high intensity signal on T1 and T2 weighted images.

Sphenoid mucocele has not previously been reported in Albright’s syndrome. Visual loss was, however, ultimately attributed to sphenoid mucocele in one woman age 20 with isolated fibrous dysplasia, in whom an intracranial tumour had initially been diagnosed, but had failed to respond to steroids and radiotherapy.5 Bone biopsy indicated the presence of fibrous dysplasia, and a haemorrhagic sphenoid mucocele found to encircle both optic nerves at craniotomy; however, vision had been no light perception in both eyes for some time before surgery and did not improve following it. This is in marked contrast with the present case in which, despite severe visual loss before surgery, a good visual outcome was obtained because of prompt diagnosis and surgical intervention. This parallels findings in optic nerve compression by dysplastic bone, in which a better visual prognosis is associated with a short duration of visual loss.

It is important that sphenoid mucocele be recognised as a cause of visual loss in patients with Albright’s syndrome. Patients are typically young, visual loss may be profound and bilateral, and prompt diagnosis and surgical decompression sight saving. A history of spontaneous fractures, endocrine dysfunction, and characteristic radiological appearances may provide diagnostic clues.

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Letters

Traumatic intraorbital aerocoele with pneumocephalus

EDITOR,—We report the case of orbital aerocoele with pneumocephalus due to orbital stab wound. Only two cases1 of orbital aerocoele have been described previously, both caused by surgical trauma. The case we present is unique in several aspects; it was caused by non-surgical trauma, was acute, resolved spontaneously, and was associated with pneumocephalus. The absence of any ocular injury is also unusual.

CASE REPORT

A 25-year-old man attended the accident and emergency department. He had sustained a stab wound with a kitchen knife to his left orbit and complained of pain and blurred vision in his left eye.

Examination 1 hour after the incident revealed left corrected visual acuity 6/36, right 6/6, marked left peribulbar swelling and a stab wound entry on the lateral aspect of the left lower lid, 1 cm below the lid margin, 2.5 cm long, 0.5 cm wide. There was proptosis which was not reducible, marked restriction of eye movements, chemosis, but no sign of conjunctival or scleral laceration visible externally. The cornea was oedematous and intraocular pressure was 40 mm Hg. The pupil reacted sluggishly with no definite relative afferent pupillary defect and there was no sign of intraocular injury.

Orbital x ray (Fig 1) showed a radiolucent area within the left orbit. Computed tomography scan (Fig 2) showed a large collection of intraorbital, mainly intracanal, air with proptosis. Intracranial air was demonstrated in the left parasellar subarachnoid space.

Over the next 9 hours the degree of proptosis reduced spontaneously. Eye movements, intraocular pressure, and corneal appearance returned to normal. Visual acuity improved to 6/9. In view of the intracranial air the patient was observed closely for signs of infection but developed no complications. Prophylactic
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