Figure 1 (a) Subretinal aspirate. Large foamy macrophages containing lysed erythrocytes are seen against the background of intact red blood cells. (b) Subretinal aspirate. A clump of presumed RPE cells containing elongated neuromelanin granules. (c) A large disciform lesion at the posterior pole as seen after vitrectomy, 4 months after the initial presentation. (d) A peripheral disciform lesion superimposed over the area of retinal detachment observed at the same time of examination.

Figure 2 (a) Macrophotograph of the enucleated eye (horizontal section) demonstrates a large amelanotic tumour and the detached retina. A white fibrous band (arrowhead), which is a disciform reaction on histology, is seen adjacent to the tumour. (b) Microphotograph of the inferior part of the globe shows the edge of the melanoma (M) and a haemorrhagic exudative detachment of the retina (R). The site of the biopsy (arrowheads point to the sutures) is in the proximity of a disciform strand (D).

melanoma and an enucleation was performed. Histopathological examination revealed a predominantly spindle B cell malignant melanoma (18×12×18 mm), occupying the nasal calotte from the ciliary body to the optic nerve (Fig 2a). A remarkable feature was the presence of a prominent disciform scar at the posterior pole with an extension into the temporal and nasal mid periphery (Figs 2a and 2b). The site of the choroidal biopsy, identifiable by an intrascleral suture was in opposition to the disciform lesion, but at a considerable distance to the anterior edge of the melanoma (Fig 2b).

COMMENT

Despite the initial intention of local resection of the presumed melanoma, the intraoperative visualisation of an unaffected choroid left an aspiration biopsy the only reasonable mode of action. Negative cytological results in conjunction with an extensive clinical examination failed to establish the diagnosis of melanoma. Moreover, the vitrectomy which disclosed a central disciform and a similar peripheral lesion was even more misleading, since the presence of a central disciform lesion is more commonly associated with

disciform lesions elsewhere than choroidal melanomas.⁴

To our knowledge, comprehensive studies on the accuracy of subretinal fluid aspiration in presumed intraocular tumours do not exist. However, in fine needle aspiration biopsy a 100% sensitivity rate in cases with adequate cytological material decreased to 84% when insufficient material was included.³ Conceivably, this limitation also applies to the subretinal fluid aspiration. To our knowledge the implications of two different pathological processes within the same globe have not been stressed yet.

The following reasons might explain the sampling error in our case. Migration of RPE cells into the subretinal fluid was described in cases of choroidal melanoma,⁵ but their presence is unhelpful for the distinction between melanoma and benign processes with RPE cell proliferation. Similar observations were made by Augsburger *et al*¹ and by Reese.⁶ Our case also demonstrates that a vitreous aspirate is useless, unless the tumour has perforated the retina.

Histologically the biopsy site, identified by intrascleral sutures, was in close proximity to the disciform scar, but far from the tumour. Presumably, it was this disciform scar that misled the surgeon during the operation and

was the source of the RPE cells and haemomacrophages in the cytopathological specimen. Therefore, the statement of Augsburger *et al*,¹ that a negative cytological diagnosis is not unequivocal proof of the absence of a malignancy, is particularly important in cases where a combined pathology is anticipated.

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Bilateral sudden visual loss due to sphenoid mucocoele 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 craniofacial involvement, visual loss may be caused by compression of the optic nerves¹ or chiasm,² by dysplastic bone or haemorrhage into dysplastic bone.³ We report the first reported case of visual loss due to sphenoid mucocoele 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

Figure 1 Appearance of optic discs at presentation.

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 mucocoele.

Surgical decompression was undertaken the same day. A large cavity formed by the expanded sphenoid was entered via a trans-ethmoidal 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 mucocoele.

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 mucocoele 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.⁴ Differential diagnoses include pituitary tumour, cranio-pharyngioma, meningioma, sinus neoplasm, clivus chordoma, and internal carotid aneurysm. Computerised tomography shows a homogeneous isodense or hyperdense sellar or parasellar lesion, and magnetic resonance imaging a high intensity signal on T1 and T2 weighted images.

Sphenoid mucocoele has not previously been reported in Albright's syndrome. Visual loss was, however, ultimately attributed to sphenoid mucocoele 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 mucocoele 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.³

It is important that sphenoid mucocoele 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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Figure 2 Computerised tomography scan showing sphenoid sinus mucocoele and bony changes of fibrous dysplasia.

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Traumatic intraorbital aerocele with pneumocephalus

EDITOR,—We report the case of orbital aerocele with pneumocephalus due to orbital stab wound. Only two cases^{1 2} of orbital aerocele 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 periorbital 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 intraconal, 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

Figure 1 Plain x ray of orbits, showing a loculated collection of gas in the left orbit.

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