LETTERS TO THE JOURNAL

Sir,

Orbital Inflammatory Disease and Bone Destruction

'Orbital pseudotumour' or orbital inflammatory disease can present a diagnostic dilemma. We report the case of a 75-year-old woman who presented with pain, proptosis and diplopia 4 years following excision of a basal cell carcinoma of her right upper lid. Computed tomography of the orbits revealed a mass in the right inferior orbit and destruction of the superior lateral maxillary antral wall with extension to deep temporal fossa. Evidence of sinus involvement or bone erosion is generally not associated with 'orbital pseudotumour'.[1] We describe the case to highlight the need for tissue diagnosis in equivocal cases.

Case Report

A 71-year-old woman presented initially to eye outpatients in December 1984 with an ulcerating basal cell carcinoma of the right upper eyelid (10 × 4 mm). Following initial diagnostic biopsy a full-thickness wedge excision of upper eyelid had been performed. The upper lid defect was then repaired with a lower lid bridge flap (Cutler–Beard). The pathology report confirmed complete excision of the basal cell carcinoma. The adjacent tissue showed a marked chronic granulomatous inflammatory reaction.

She next presented in September 1989 with a 3-month history of right periorbital and temporal pain. There was diplopia on right gaze and visual blurring. Her visual acuity was 6/24 in the right eye, 6/9 in the left eye. There was 6 mm axial proptosis and a firm mass palpable in the inferolateral orbit. The conjunctiva was chemotic and she had limitation of ocular movements in abduction and adduction. Her intraocular pressure was normal and visual fields full. General medical history disclosed chronic asthma for which she was on treatment with prednisolone 10 mg daily. Systemic examination was unremarkable.

Initial investigations showed an elevated erythrocyte sedimentation rate of 97 mm/h, a hypochromic microcytic anaemia (Hb 10.7 g/dl), with slight neutrophilia. Chest radiograph was normal, as was renal function, and other serological investigations including anti-neutrophil cytoplasmic antibody were negative. Computed tomography showed a large mass in the region of the inferior orbit and adjacent maxillary antrum, with apparent destruction of part of the superior lateral antral wall and extension of the mass to the deep temporal fossa (Fig. 1).

An anterior orbital biopsy via an incision through the right lower eyelid was performed. Grossly the tissue appeared hard and 'fat-like'. Histological examination showed orbital fat infiltrated by a granulomatous inflammatory lesion (Fig. 2). Some of the granulomatous lesions showed central necrosis, with accumulation of nuclear fragments and infiltration by neutrophils, surrounded by macrophages and multinucleate giant cells. There was degeneration of adipose tissue and formation of lipophages. A low-grade non-necrotising vasculitis was present. There was no evidence of micro-organisms or fungi with specially stained sections (Gram, Ziehl-Neelsen and Grocott). There was no evidence of recurrent basal cell carcinoma in any of the sections.

Prednisolone therapy was increased to 80 mg daily on the day of biopsy and rapid symptomatic relief was attained. Nine days

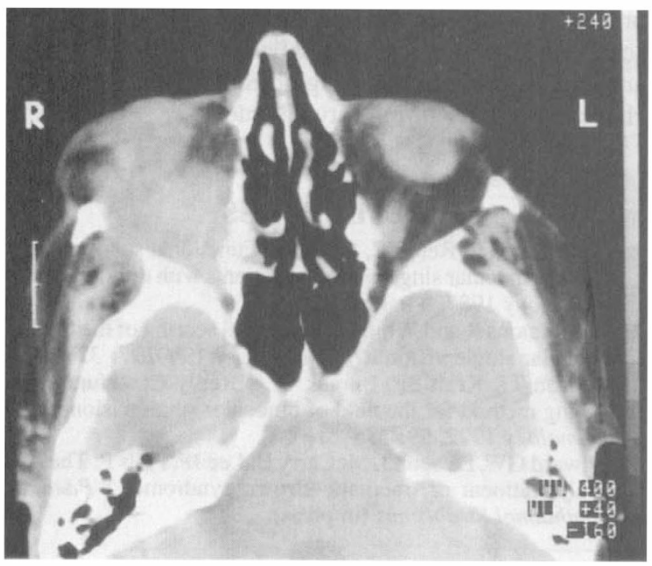

Fig. 1a.

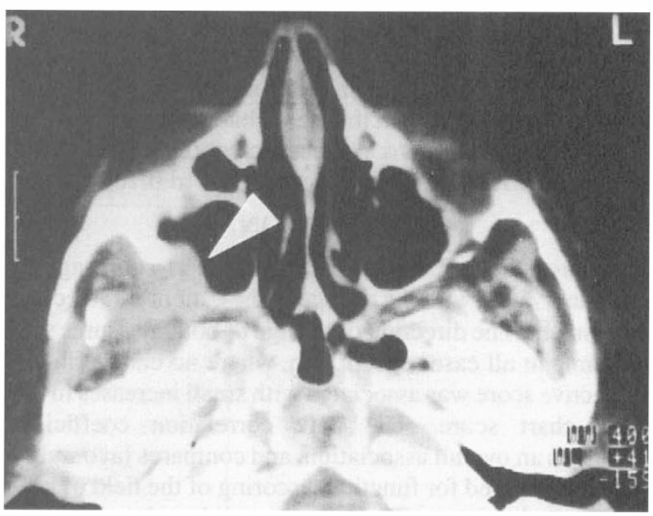

Fig. 1b.

Fig. 1. *Orbital CT scans demonstrating a large mass in the region of the inferior orbit* (a) *and adjacent maxillary antrum with destruction of part of the superior lateral antral wall and extension of the mass to the deep temporal fossa (arrow,* b).

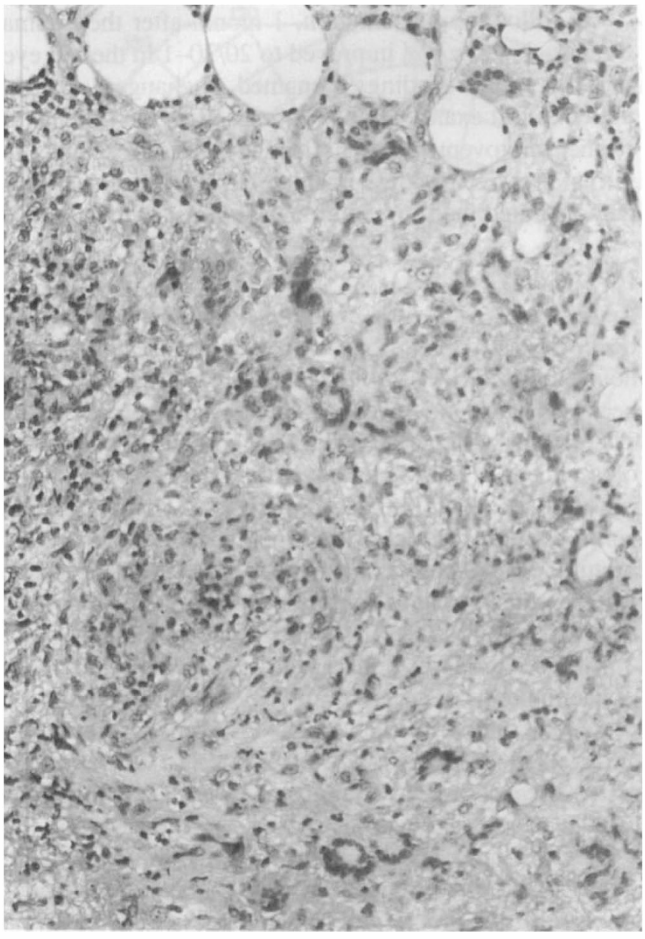

Fig. 2. *Mixed inflammatory 'pseudotumour': granulomatous lesion with areas of central necrosis, nuclear fragmentation, and infiltration with polymorphonuclear leucocytes, lymphocytes and macrophages. Multinucleate giant cells are also evident. Haematoxylin and eosin, ×100.*

later her right visual acuity had improved to 6/9, the proptosis had resolved and ocular movements were full with no restriction. Despite this initial response to steroid treatment her pain and proptosis recurred but then finally responded to a course of orbital radiotherapy. Following her relapse and radiotherapy her right visual acuity reduced to 6/36. Her steroid therapy was tapered and she is currently on prednisolone 10 mg daily with no proptosis, diplopia or pain.

Discussion

Orbital pseudotumour can be difficult to diagnose with certainty as its presentation is similar to that of other conditions causing orbital inflammation; for example thyroid orbitopathy, orbital cellulitis, lymphoma, primary or secondary orbital malignancies. It is also important to recognise that in a proportion of cases of inflammatory pseudotumour there will be progression from orbital to systemic disease, e.g. Wegener's granulomatosis.[2] Orbital inflammation is typically painful, with proptosis, chemosis, limitation of ocular movements and visual impairment. These features, as shown by our patient, give no clue, however, to the definitive diagnosis. Indeed our patient's previous history raised doubts of recurrence of her basal cell carcinoma, possibly from transplantation of neoplastic cells with the lower lid bridge flap reconstruction 4 years previously. A recurring basal cell carcinoma will have infiltrative signs which fix the globe, restricting its movement. Contracting tissue results often in enophthalmos or dystopia. This case exhibited some degree of infiltration resulting in restriction, but mainly exhibited a mass effect which produced proptosis. The clinical signs therefore suited the diagnosis of a tumour with a differing cell type from the original basal cell carcinoma and the two diseases are coincidental.

Computed tomography has been reported as the most useful study to rule out other conditions within the orbit.[3] It is suggested that the presence of bony erosion or sinus involvement goes against the diagnosis of orbital inflammatory disease and is more suggestive of a malignant process[1]. Others have found evidence of bone destruction in patients with orbital pseudotumour, albeit rarely.[4–6] Our patient falls within the latter group, with destruction of part of the superior lateral maxillary antral wall and extension of the mass to the deep temporal fossa, and highlights the need for an expeditious biopsy in equivocal cases before empirical treatment with steroids or radiotherapy.

Orbital pseudotumours can be divided into two main groups: polymorphic inflammatory diseases and predominantly lymphoid infiltrations.[2] The first group comprises non-granulomatous or granulomatous reactions, which occur as part of a systemic disease or are confined to the orbit. Pathological examination of our patient's biopsy revealed an infiltrate of neutrophils, lymphocytes and macrophages in the adipose and fibrous connective tissue. There was breakdown of orbital fat, with a lipogranulomatous reaction predominating. All these are features of mixed inflammatory (polymorphic) infiltrations. As the disease progresses there have been reports of bone destruction with extension to the intracranial cavity.[6]

The patient's initial rapid response to systemic steroids is in keeping with other reports of granulomatous orbital inflammatory disease.[7] Despite the lack of any features of systemic involvement in our patient continued long-term review is advised, as patients with a diagnosis of polymorphic orbital inflammatory disease may manifest later with progression from orbital to systemic disease.[2]

This case is presented to highlight the need for tissue diagnosis in equivocal cases of orbital inflammation where computed tomography may be unable to give a firm conclusion. Bony destruction can occur in orbital pseudotumour as well as other conditions (orbital malignancies, Wegener's granulomatosis, fungal infections, etc.).

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Sir,

A Case of Unique Altitudinal Distribution of a Retinal Contusion (Berlin's Oedema) in a Healthy Teenager

A 16-year-old caucasian male presented complaining of diffuse headaches, lightheadedness and blurred vision in his left eye, 48 hours after suffering a blunt trauma by a fist to his left lateral orbit and temple. In the previous 12 hours before ophthalmological examination the patient had experienced loss of the entire inferior half of the left field of vision, accompanied by scintillations and nausea. Past history was unremarkable except that the patient had been smoking 20 cigarettes per day for the previous 2 years. Best corrected visual acuity was 20/20 in the right eye and 20/50 improved with pinhole to 20/40 in the left eye. There was +2 relative afferent pupillary defect in the left eye. Slit lamp examination of both eyes was within normal limits. Intraocular pressure was 18 mmHg in both eyes. Fundoscopic examination through a dilated pupil was normal in the right eye; in the left eye substantial retinal whitening was noted in the entire superior half of the posterior pole with minimal extension beyond the superior temporal vessels. The retinal pathological area transected the fovea in a perfect horizontal line, creating a 'hemicherry red spot' (Fig. 1). The left optic nerve head was normal in appearance with no signs of oedema, haemorrhages or vasculopathy. No evidence of occlusion or thromboembolism was found. Goldmann's visual field testing confirmed the clinical picture, demonstrating an inferior altitudinal defect in the left eye (Fig. 2). Ishihara tests for colour-blindness were normal in both eyes. Fluorescein angiography failed to show any abnormality. There was no evidence of vascular constriction, occlusion or perivascular leakage. The fluorescein transit time to the superior retina was not slowed in any way compared with the distribution to other areas of the retina. Heart and lung auscultation was normal. No bruits were heard along either carotid artery, and the rest of the physical examination was within normal limits. Computed tomography of the head and orbits did not reveal any pathological findings.

On follow-up examination, 1 month after the trauma, the visual acuity had improved to 20/20–1 in the left eye, but the retinal findings remained unchanged. On the patient's last examination, 3 months after the trauma, no further improvement was noticed in the visual acuity. The retinal area seemed less opaque, but the left visual field demonstrated the same inferior altitudinal defect.

Discussion

Retinal contusion is a countercoup injury which may occur centrally (Berlin's oedema) or peripherally. A few hours after such trauma the affected area of the outer retina becomes white and opaque, due to tissue disorganisation. The swollen layer blocks the background choroidal fluorescence in angiography but with no leakage. Prognosis is usually excellent, except for cases with severe pigment epithelial damage, subfoveal haemorrhage or choroidal rupture. When photoreceptors are destroyed, localised visual field defects are expected, but arcuate field defects are not found due to an intact overlying nerve fibre layer.[1]

Altitudinal hemianopsia has been reported with ischaemic optic neuropathy, optic neuritis, meningiomas, congenital nerve head anomalies, advanced glaucoma, enlargement of the internal carotid arteries, and paranasal sinus disease. Traumatic injuries have also been implicated, causing damage to the blood vessels supplying the optic nerve head via the relatively narrow subarachnoid space. Torsion and oedema compromise tissue oxygenation, resulting in a unilateral altitudinal defect having a horizontal border, great density and steep edges.[2] In a recent case report altitudinal hemianopsia developed after severe facial injuries during a motor vehicle accident.[3] But in contrast to our case, bone fragments were found to apply pressure to the optic nerve, and no retinal contusion was noticed. A history of ocular trauma was present in 5 of 27 patients under the age of 30 years reviewed at Wills Eye

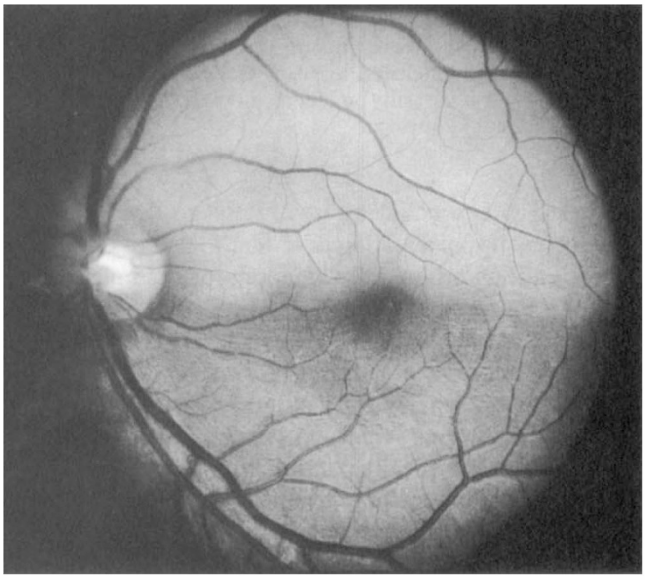

Fig. 1. *The left posterior pole. Note the opaque superior retina and the horizontal line, traversing through the fovea, separating healthy and diseased retina.*