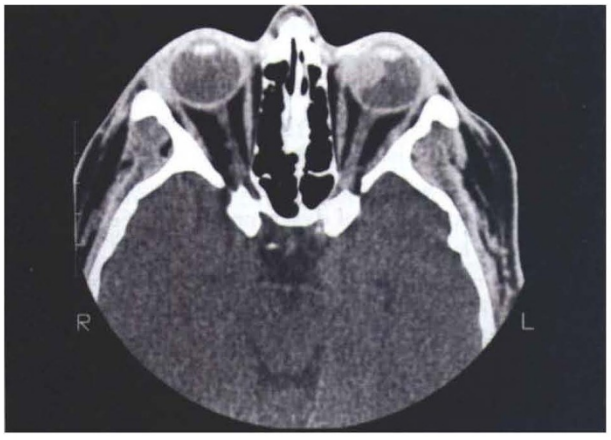

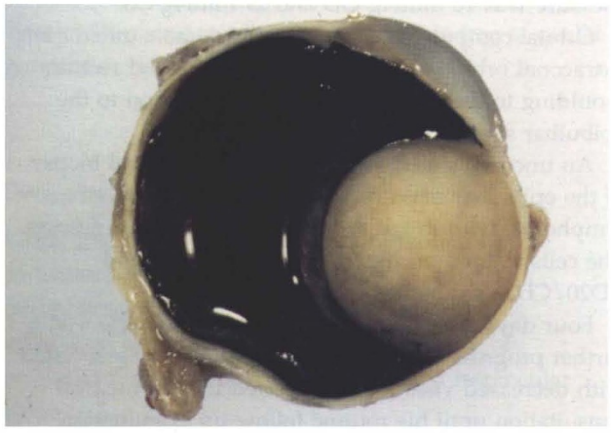

Fig. 1. *Above: Orbital CAT scan (axial section). A retrolental growth is clearly evident in the left eye. Below: Gross specimen after fixation. The solid nature of the mass is visible in section.*

show metatastic dissemination. A meticulous enucleation was carried out with an implant of hydroxyapatite coral prosthesis enveloped in donor sclera.

Macroscopic examination of the excised eye showed an encapsulated tumour of greyish appearance indenting the lens and a serous retinal detachment extending as far back as the optic disc (Fig. 1, below).

Microscopic examination proved that the tumour was a neurilemmoma with the characteristic pattern described by Antoni. It probably took origin from Schwann cells associated with the ciliary nerves and was histologically benign (Fig. 2).

Comment

Although the benignity of the tumour does not contraindicate enucleation in order to avoid progression to a painful blind eye, in this case enucleation was carried out in view of a possible malignant melanoma. It is known that this type of tumour may undergo accelerated growth during pregnancy and ciliary body melanomas have also been described as evolving in this way. Besides, at times they do transilluminate. Therefore, in this case the assumption of a melanoma until proven otherwise was mandatory. Even in the presence of a fine needle biopsy positive for schwannoma, the malignant potential of this tumour cannot be assessed without histological sections and the rate of growth as measured

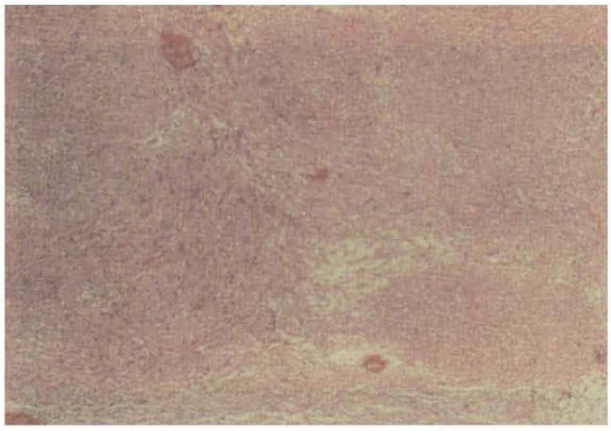

Fig. 2. *Haematoxylin–eosin section (×100). Interwoven bundles of bipolar cells, characteristic of schwannoma, are seen.*

by incorporation of radioactive phosphorus is not greatly different from that of a malignant melanoma.

In conclusion it seems that this condition will continue to be recognised retrospectively and treated as something different until a reliable diagnostic test is available.

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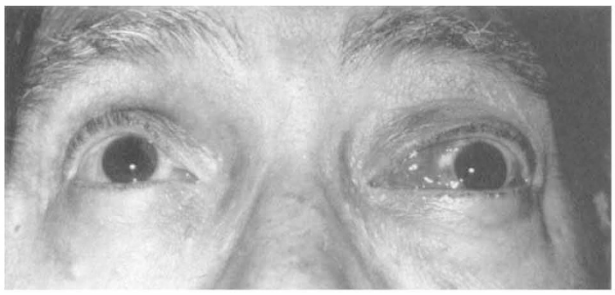
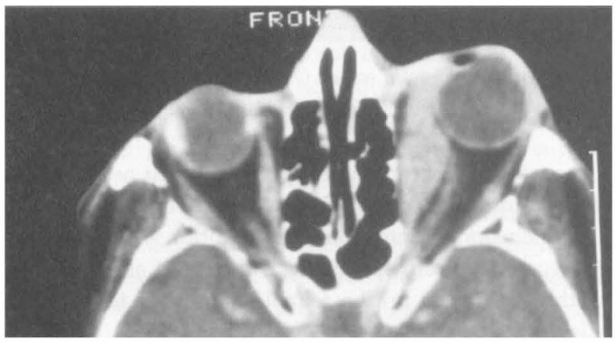
Sir,

Central retinal artery occlusion due to rapidly expanding orbital lymphoma

The presentation of lymphoproliferative disorders affecting the orbit is usually insidious and painless.[1] The common manifestations are proptosis, blepharoptosis, diplopia, limited ocular motility and rarely decreased visual acuity. To our knowledge, this is the first report of a rapidly progressive orbital lymphoma resulting in an



(a)



(b)

Fig. 1. *(a) Clinical photograph demonstrating left exotropia, proptosis and a sub-conjunctival mass. (b) Orbital axial CT scan. An extraconal mass displacing the left medial rectus muscle medially and extending onto the epibulbar surface is demonstrated. The lesion has distinct borders and uniform density.*

orbital compartment syndrome leading to a central retinal artery occlusion.

Case report

An 85-year-old man presented with a 6 day history of sudden painful swelling of his left eye and horizontal diplopia. Six months prior to presentation the patient experienced bilateral sequential seventh cranial nerve palsies associated with headaches. His past ocular history was notable for an intracapsular cataract extraction in both eyes and a subsequent retinal detachment repair of the right eye 20 years earlier.

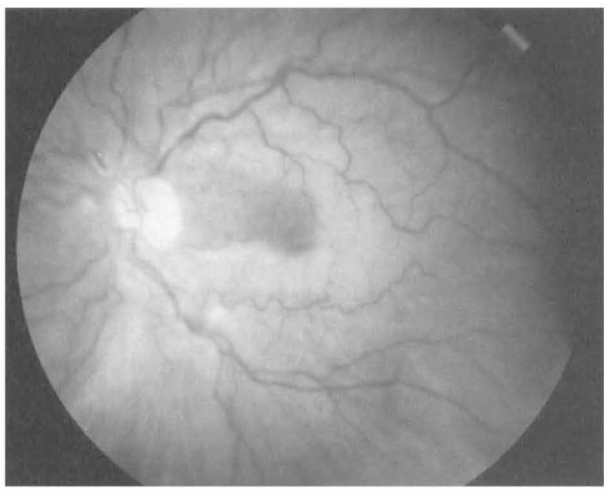
At presentation, visual acuity was 20/200 OD and 20/60 OS with aphakic spectacle correction. There was no afferent pupillary defect of dyschromatopsia. Seven millimetres of left proptosis (Fig. 1a), associated with generalised limitation of ductions and an exotropia in primary position, was present. A non-tender rubbery subcutaneous mass was palpable near the medial canthal angle, and an epibulbar subconjunctival mass was noted at the medial aspect of the left globe. The intraocular pressure was 10 mmHg OD and 27 mmHg OS.

Orbital computerised tomography demonstrated a left extraconal orbital mass, displacing the medial rectus, moulding to the globe with anterior extension to the epibulbar space (Fig. 1b).

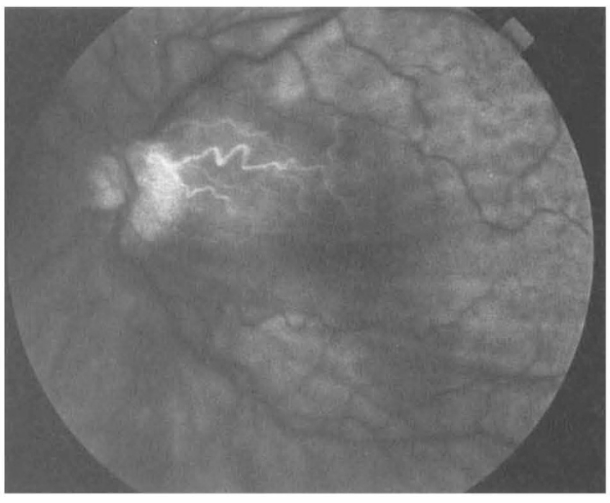
An uncomplicated subconjunctival incisional biopsy of the epibulbar mass revealed a high-grade B-cell lymphoma, with irregular nuclei and numerous mitoses. The cells stained positively with IgG Kappa and CD20/CD22 antibodies.

Four days after the biopsy the patient experienced a further progression of proptosis of the left eye associated with decreased vision, but neglected to seek medical consultation until his routine follow-up examination 1 week post-operatively. At that time, the left proptosis was more prominent and the vision in the left eye had decreased to 20/400. Diffuse retinal oedema sparing the macula (Fig. 2a) and choroidal folds were present in the left eye. Fluorescein angiography demonstrated filling only of the cilioretinal artery, indicating a central retinal artery occlusion (Fig. 2b).

Anterior chamber paracentesis was performed with no immediate response. However, prompt treatment with oral corticosteroids (prednisone 80 mg/day) resulted in dramatic improvement of the vision in the left eye to 20/40, reduction in proptosis and resolution of retinal oedema within 2 days.



(a)



(b)

Fig. 2. *(a) Fundus photograph of the left eye after the acute visual loss. Diffuse retinal oedema sparing the macula is evident. (b) Fluorescein angiography reveals filling of the cilioretinal artery and circumlinear veins. No filling of the retinal vasculature is evident.*

Due to the high-grade nature of the lymphoma and the progressive symptoms, the investigation for possible central nervous system involvement was expedited. A lumbar puncture demonstrated malignant lymphocytes in the cerebrospinal fluid. The patient was treated with 2000 cGy whole-brain irradiation and 3500 cGy to the left orbit. In the ensuing months he sequentially developed a complete third nerve palsy of the right eye and choroidal effusions of the right eye, all in the setting of lymphomatous meningitis. The patient was then treated with five cycles of intrathecal and one dose of intravenous methotrexate. Recovery of the ocular motility was achieved after 4 months. Thirty-three months following treatment the patient is free of ocular symptoms.

Discussion

Visual loss complicating orbital lymphoma may be permanent due to optic neuropathy,[2,3] which is a consequence of direct external compression or infiltration of the optic nerve. Another cause of permanent visual loss is chronic elevated intraocular pressure.[4]

Central retinal artery occlusion has been described as an early manifestation of ocular–central nervous system (CNS) lymphoma.[5] In these cases there was no orbital involvement. To our knowledge an orbital compartment syndrome, as observed in this patient from a rapidly expanding orbital lymphoma resulting in central retinal artery occlusion, has not been reported. In this case we postulate that the rapid tumour growth caused increased orbital pressure and compromised perfusion of the optic nerve. Such a rapid clinical progression is unusual for orbital lymphoma. In a large series, the mean duration of symptoms attributable to orbital lymphoma prior to presentation was 4 months.[6] In this case, symptoms were reported for only 10 days prior to the central retinal artery occlusion. The biopsy sampled only the anterior-most portion of the orbital tumour, and was not associated with any intraoperative or post-operative haemorrhage. It did not appear to adversely affect the inherent clinical course of this aggressive tumour. Therefore, patients with a rapidly progressive orbital lymphoma should be monitored very closely after biopsy, even in the absence of an orbital haemorrhage. In rare cases demonstrating rapid progression, systemic steroids may be warranted to decrease the risk of developing an orbital compartment syndrome, prior to definitive treatment. The systemic investigation should be accelerated because prolonged steroid use may mask cerebrospinal fluid signs of CNS lymphoma. Combined systemic steroids, cranial irradiation and intrathecal methotrexate resulted in prolonged control of disease and survival in this patient.

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Sir,

Carbon deposits in the conjunctival fornices

We describe a patient who had long-term exposure to airborne graphite, and presented with ocular discomfort associated with deposits of graphite in the subepithelial connective tissue of the conjunctival fornices.

Case report

A 30-year-old woman presented with discomfort and a foreign body sensation in the right eye. Her past medical history was unremarkable; however, her occupation for the previous 12 years involved sawing and cutting graphite blocks using a stationary power tool with built-in eyeguard, therefore without protective goggles. The sawing produced a large quantity of graphite dust. General physical examination was normal, with specifically no clinical or radiological evidence of interstitial lung disease. Visual acuities were 6/6 right and left. Deep in all four conjunctival fornices were black granular deposits with some lace-like subconjunctival scarring. The cornea, anterior chamber, lens, intraocular pressures and fundi were all normal. The tear film was not disturbed in either eye. A conjunctival biopsy was taken under local anaesthetic.

Histology showed conjunctiva with deposits of black particulate material both within the macrophages of the subepithelial connective tissue, and extracellular, consistent with graphite. EXAX analysis (energy dispersive analysis of X-rays of the wax-embedded tissue