Aggressive orbital lymphoma in AIDS

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In recent years, the association of malignant non-Hodgkin's lymphoma (NHL) with AIDS has become well established.^{1,2} These tumours tend to be of high grade and aggressive. They are associated with a poor prognosis. Despite the clear predilection of AIDS-related NHL for extranodal sites, few cases with orbital involvement have been reported.^{1,2} Herein, we review the literature and report a case of a 40-year-old white male with AIDS with a highly aggressive unilateral orbital NHL which demonstrated extensive bony destruction.

Case report

A 40-year-old white male with a 2 year history of AIDS was admitted to the New York Hospital-Cornell Medical Center with a 4 week history of progressive left periorbital pain and swelling, as well as declining vision in the left eye over the preceding week. He also reported a 4 month history of a left frontal headache, with occasional fevers, chills, and night sweats. Five weeks before this admission, the patient had undergone an extensive neurological examination, including contrast computed tomography (CT) of the head and diagnostic lumbar puncture, which were all

within normal limits. A careful review of systems revealed chronic nasal congestion with recurrent rhinorrhoea and a bloody nasal discharge. The patient denied any history of weakness or paraesthesias and there was no history of trauma. The medical history was significant because of a 2 year history of AIDS, complicated by *Pneumocystis carinii* pneumonia, cryptococcal meningitis, and recurrent genital herpes. In contrast, there was no significant ocular history.

On admission, the patient had a fever of 39.1°C. Examination of the nose revealed a large, fleshy, erythematous mass eroding through the mucosa of the left naris. The remainder of the physical examination was unremarkable.

On ophthalmic examination, corrected visual acuity was noted to be 20/40 in the right eye and 20/400 with marked subjective red colour desaturation in the left eye. An afferent pupillary defect was detected in the left eye. Ductions were full on the right and markedly reduced on the left (Fig 1). Hertel exophthalmometry revealed 4 mm of left proptosis with relative resistance to retropulsion. While there was marked left sided lid swelling and erythema, there was neither conjunctival injection nor chemosis. Intraocular tension was 14 mm Hg in each eye. There was no evidence of intraocular inflammation. Ophthalmoscopy was significant only for left disc hyperaemia. There were no retinal haemorrhages, cotton wool spots, or striae present.

Laboratory test results included a white blood cell count of $4500 \times 10^9/l$ (78% polymorphonuclear leucocytes and 19% lymphocytes) and a T helper cell (CD4) count of $4 \times 10^9/l$ (normal range 800–1200). Blood cultures were negative. CT scan of the head and orbits revealed a large contiguous mass lesion of the left orbit, nasal cavity, paranasal sinuses, and cavernous sinus. Superior extension through the cribriform plate into the anterior inferior frontal lobe was noted (Fig 2). Obvious bony erosion as well as proptosis of the globe were evident (Fig 3). Apparent tumour impingement on the optic nerve was noted in the left orbital apex.

Biopsy of the mass was performed through the left naris. Pathological examination of the biopsy specimen revealed a high grade diffuse large cell lymphoma (Fig 4). Staging examination including CT scans of the chest, abdomen, and pelvis as well as repeated lumbar punctures and bone marrow biopsy were all negative for the presence of lymphoma.

The patient was treated with 250 cGy/day of radiation therapy to the orbit, sinuses, and anterior frontal lobe for 15 days. Treatment also included intravenous dexamethasone and one dose of intrathecal methotrexate. Subsequently, there was significant improvement of the proptosis and extraocular motility. The visual acuity of the left eye improved to 20/40 3 weeks after completion of therapy. Four months after dis-

Fig 1A

Fig 1B

Figure 1 Limitation of right (A) and left (B) lateral gaze only 4 weeks after onset of ocular symptoms.

Figure 2 Coronal computed tomogram demonstrating the presence of tumour in the maxillary and ethmoid sinuses, the nasal septum, as well as extension through the cribriform plate into the inferior frontal lobe.

Figure 3 Axial computed tomogram demonstrating bony erosion of the medial orbital wall, axial globe displacement, and tumour impingement on the optic nerve.

Figure 4 Haematoxylin and eosin staining of tumour biopsy consistent with diffuse large cell malignant lymphoma.

charge, however, the patient was readmitted with evidence of metastatic large cell lymphoma involving the lung retroperitoneum.

Comment

Non-Hodgkin's lymphoma (NHL) of the orbit is uncommon in the general population and is usually a low grade, small cell lymphoma occurring in the sixth or seventh decades. Lymphomas accounted for only 7.5% of primary orbital tumours in one large series.³ Orbital involvement in systemic lymphoma is also quite rare.⁴

Malignant lymphomas of the orbit have a characteristic CT appearance, with irregular margins, lack of encapsulation, and moulding to the globe and orbital bones being typical. Erosion of the bones of the orbit or sinuses is rarely seen.⁵

Recently, three case reports^{6,7,8} and one autopsy investigation⁹ have described features of orbital lymphoma in young adult patients with AIDS. Common to all three cases was that the lymphomas were all of the large cell variety; they primarily involved the medial orbit; and they caused radiographic opacification of the ethmoid sinus. Antle and associates reported a case of a 28-year-old man with large B cell lymphoma which did not show radiographic evidence of erosion or expansion of the orbit although 'thickening' of the ethmoidal bony complex was noted.⁷ Similarly, Mansour reported a case of a 33-year-old patient with AIDS which demonstrated the characteristic CT appearance of moulding of the lymphoma to the orbital wall seen in elderly patients without AIDS.⁶

Most recently, Turok and Meyer reported a case of a 34-year-old man which demonstrates some of the atypical features of orbital lymphoma in patients with AIDS.⁸ In this case, there was rapid clinical progression and the CT scan demonstrated both indentation and displacement of the globe. Immunohistochemical staining confirmed a large cell lymphoma of the B cell lineage.

This case of unilateral orbital NHL in a patient with AIDS demonstrates atypical radiographic features not previously reported. The presence of significant bony erosion, including erosion through the cribriform plate into the inferior aspect of the frontal lobe is consistent with an extremely aggressive tumour, as is the rapid 4 week clinical progression of periorbital swelling and pain. Lymphomas in patients with AIDS have frequently been described as aggressive and have been shown to be 'high-grade' in 78% of cases.¹ High grade lymphomas have a poor prognosis with only 10% 2 year survival in a study of 90 homosexual men, many of whom had documented AIDS.¹⁰ Our patient returned only 4 months following treatment with evidence of systemic disease.

As in previously reported cases, our patient had a lymphoma of the large cell variety which primarily involved the medial orbit. The radiographic features, including erosion of bone, however, distinguish it from the others. It is clear that extranodal malignant lymphoma, specifically of the orbit, is becoming more common with the rapid expansion of the AIDS epidemic.

This case underscores the fact that in this patient population, lymphoma may have atypical clinical or radiographic features. NHL should be considered in the differential diagnosis of any patient with AIDS who presents with unilateral orbital signs, such as proptosis or motility disturbances. With improvements in diagnosis and treatment of this disease, short and long term prognosis may improve.

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Ocular pulse measurements to assess pulsatile blood flow in carotid artery disease

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Patients with carotid artery stenosis frequently present with symptoms associated with embolic episodes or, more rarely, with chronic ocular ischaemia, the varied signs of which include hypotensive retinopathy, uveitis, rubeosis, corneal decompensation, and cataract.¹ Carotid Doppler ultrasound scanning is the preferred test to detect those patients with severe stenosis who might benefit from vascular surgery and, in the postoperative period, to monitor increased carotid perfusion.² Assessment of ocular perfusion is not routinely performed. We report non-invasive measurement of pulsatile ocular blood flow (POBF) using the Langham pneumotonometer^{3,4} in two patients undergoing carotid artery surgery.

Case reports

CASE 1

A 72-year-old man presented with a 4 month history of left sided amaurosis fugax relieved by stooping. He had intermittent claudication and took oxpentifylline. His acuity was 6/9 in the right eye and counting fingers in the left. The right eye was normal apart from a low central retinal artery perfusion pressure revealed by digital compression of the globe. The left eye showed low grade anterior uveitis with rubeosis, an unreactive dilated pupil, widespread peripheral fundal haemorrhages, and a very low central retinal artery perfusion pressure, pulsating with

the slightest pressure on the globe. After intravenous acetazolamide the left vision improved to 6/18. There were bilateral carotid bruits, and absent pulses in the right arm and distal to both femoral arteries.

Fluorescein angiography of the left eye confirmed slow flow retinopathy, with a prolonged arm to retina time of 19 seconds (normal <15 seconds) and delayed arteriovenous transit, some retinal veins never filling completely. Doppler ultrasound demonstrated reduced flow velocity in the right common, external, and internal carotids; no flow in the left carotid system; reverse flow in the right vertebral; and increased velocity in the left vertebral.

Digital subtraction angiography (DSA) was consistent with these findings demonstrating an occlusion of the right innominate and left common carotid arteries at their origins. Intracranial flow was solely from the left vertebral artery. Surgery revealed a tight atheromatous stenosis in the left internal carotid artery extending from the bifurcation. Following left subclavian to right common and internal carotid Dacron grafting there were no further episodes of amaurosis fugax though the clinical signs were unchanged.

Ocular pulse measurements and systemic blood pressure were recorded before, and 5 days after surgery, in the supine position (Fig 1). The POBF increased in the postoperative period by more than four times in the right eye and rose from being unrecordable to almost 200 µl/min in the left eye (Table 1).

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