the bulbar conjunctiva was treated with cryotherapy, following which there have been no further episodes of bleeding from the eye over the last 2 years.

Comment

The patient presented had episodes of nasal and gastrointestinal bleeding as a child; she then presented in the third decade with bleeding from the eye. A diagnosis of BRBNS was established clinically, with the history of episodes of blood loss and multiple cavernous-type haemangiomas involving the skin of the face, the trunk and mucosal lesions affecting the eye, mouth and nose.

The episodes of pain and a sensation of fullness, together with the episode of horizontal diplopia, might represent intermittent haemorrhages into the closed tissue spaces of the orbit with spontaneous reabsorption. However, a contrast-enhanced CT scan failed to demonstrate any orbital lesions, in addition to the fact there was no proptosis. This is the first case as far as we are aware of BRBNS presenting with bleeding from the palpebral aperture. As there were no further gastrointestinal haemorrhages since the age of 5 years, endoscopy was not contemplated by the gastroenterologists; however, this does not rule out the presence of asymptomatic vascular malformation in the gut.

Though previous studies[3,5] have reported vascular malformations in the orbit,[6] iris and retina as a part of BRBNS, our case did not demonstrate these features. Three types of vascular lesions have been described: (1) large cavernous haemangiomas affecting vital structures, (2) the classical compressible blood sacs and (3) irregular blue-black spots within the skin. Other viscera such as the thyroid gland, the kidney and the heart may also be affected.[7] Though the central nervous system is only rarely involved,[8] hemispheric and cervical nerve involvement has been described.[9] Some histopathological studies have identified the vascular lesion in BRBNS as erectile cavernous tissue.[10] This might explain bleeding in our case.

Maffucci syndrome,[11] Klippel-Trenauny syndrome[11] and multiple glomus tumour[12] should be considered in the differential diagnosis of BRBNS.

In summary, an ophthalmologist should entertain a high index of suspicion of BRBNS when confronted with bleeding from cavernous conjunctival haemangiomas. This syndrome is associated with cutaneous and gastrointestinal vascular malformations and hence the onus is on the surgeon to make the appropriate referrals and order appropriate investigations to avoid potential life-threatening haemorrhages. As far as we are aware, this is the first case of bleeding from the conjunctival sac in BRBNS.

We are grateful to Mr E.G. Davies, FRCOphth, Consultant Ophthalmologist, for allowing us to use one of his patients to present this case.

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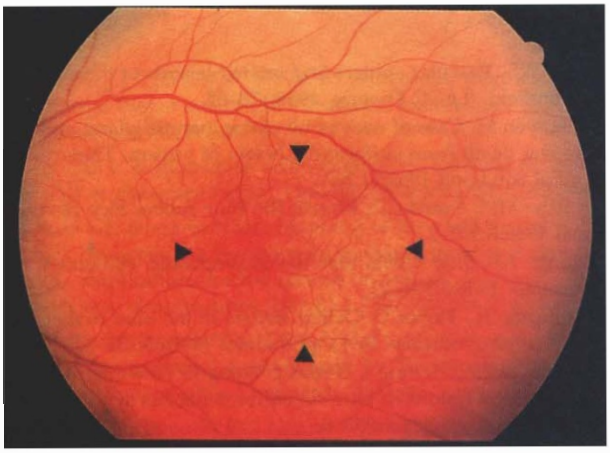
Sir,

Mantle cell lymphoma presenting as a choroidal mass: part of the spectrum of uveal lymphoid infiltration

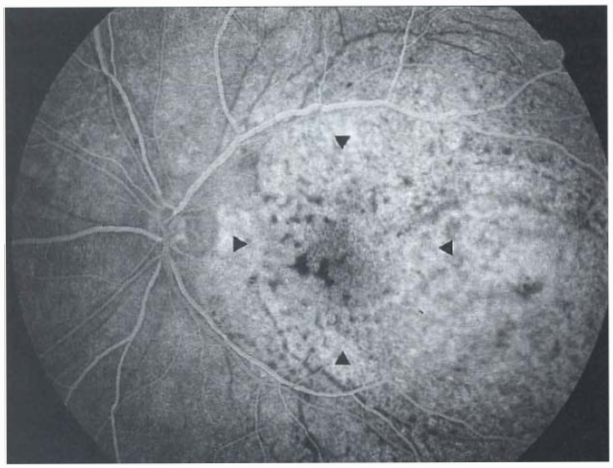
Although intraocular involvement with large cell high-grade B cell lymphoma is rare but well recognised, small B cell intraocular lymphoid infiltration is not well recognised and is extremely rare. We describe the clinical, radiological and pathological features of a mantle cell lymphoma presenting as a choroidal mass.

Case report

A generally fit 57-year-old man presented to the Ophthalmology Department at Leicester Royal Infirmary with a 2 month history of aching eyes and a 3 day history of gradual decreased visual acuity in the left eye. His left visual acuity was 6/24, with mild inflammatory activity in the anterior chamber and mild ocular injection. There was an area of choroidal infiltration and elevation at the posterior pole (Fig. 1a) but minimal vitreous cellular



(a)



(b)

Fig. 1. *(a) A fundus photograph showing a shallow choroidal elevation (arrowed) temporal to the macula with a mottled appearance. There is pigment epithelial clumping at the macula and more peripheral pigment epithelial streaks. (b) Fluorescein angiogram showing no leakage from the choroidal mass but highlights the masking defects from the pigment epithelial changes. The choroidal mass (arrowed) is slightly out of focus due to its elevation.*

infiltrate. The appearance of the left optic disc was normal. Examination of the right eye was completely normal with a visual acuity of 6/6.

Full blood count, plasma viscosity, autoantibody screen, ANCA and serum angiotensin converting enzyme level were normal. Fluorescein angiography revealed no leakage from the choroidal mass but highlighted the masking defects from the pigment epithelial changes (Fig. 1b). MRI scans of the brain and orbits were normal but detail in the orbital scans was limited at that stage. To exclude a possible extraocular primary neoplastic lesion serum electrophoresis, bone marrow trephine, chest radiograph, thoracic and abdominal CT were performed and found to be normal.

A provisional diagnosis of Harada's disease was made and the patient commenced on topical and systemic corticosteroids. No improvement occurred in either fundus appearance or visual acuity and so the steroids were stopped. Nine months after presentation, the patient was noted to have mild left proptosis (3 mm) and was referred to the orbital and oculoplastic service at Leicester Royal Infirmary. At this time visual acuity was 6/60 and there was a grade III left relative afferent pupillary defect. Fundal appearance, however, remained stable. MRI scan of the orbit revealed a homogeneous, non-enhancing intraconal 2.0 1.5 cm soft tissue mass adjacent to the left optic nerve and diffuse thickening of the choroid which was most marked around the macular region. There was no globe indentation (Fig. 2).

A left lateral orbitotomy and biopsy of the mass adjacent to the optic nerve was performed. Histology, immunohistochemistry and *in situ* hybridisation (Fig. 3) revealed this to be a mantle cell B cell lymphoma (REAL classification) with lambda light chain restriction.

The patient underwent orbital radiotherapy (total dose 34 Gy) resulting in disappearance of the mass, improvement of visual acuity to 6/12 and recovery of normal pupil function. At the last review (34 months after radiotherapy), pigment clumping at the posterior pole persisted but there was no evidence of tumour recurrence on the repeat orbital MRI scan.

Comment

Almost all ocular/orbital lymphomas are non-Hodgkin's lymphomas (NHL) of B cell lineage and a variety of types of B cell lymphoma may involve the periocular tissues.[1–3] Intraocular lymphoma is usually of the large cell NHL type having spread from the systemic or central nervous systems.[4]

We describe a very rare manifestation of a mantle cell lymphoma presenting as an isolated choroidal mass with uveitis. Positive reactions for B cells (CD5, 20, 79) and cyclin D1 with a reactive component of T cells (CD5, 45; RO3, 43) and negative reactions for CD10 and 23 confirmed the diagnosis of mantle cell lymphoma.[5] Our case can be considered part of the spectrum of the rare entity uveal lymphoid infiltration (ULI), which also may present with visual disturbance and a choroidal mass but is usually associated with a visible peribulbar mass and

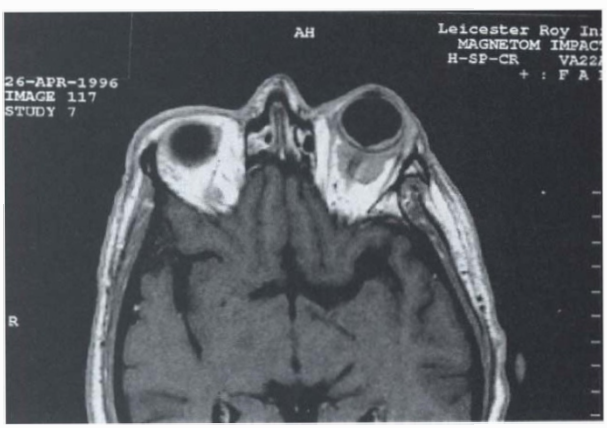
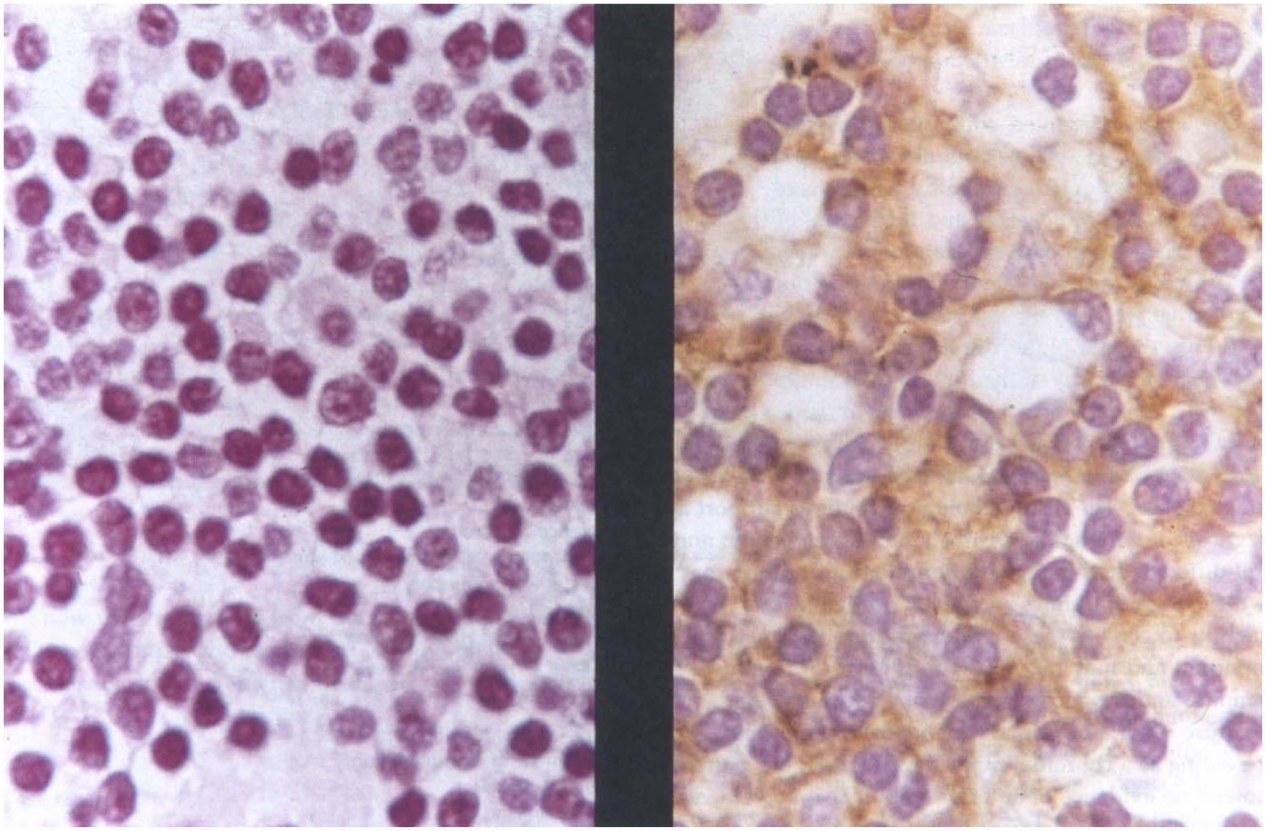
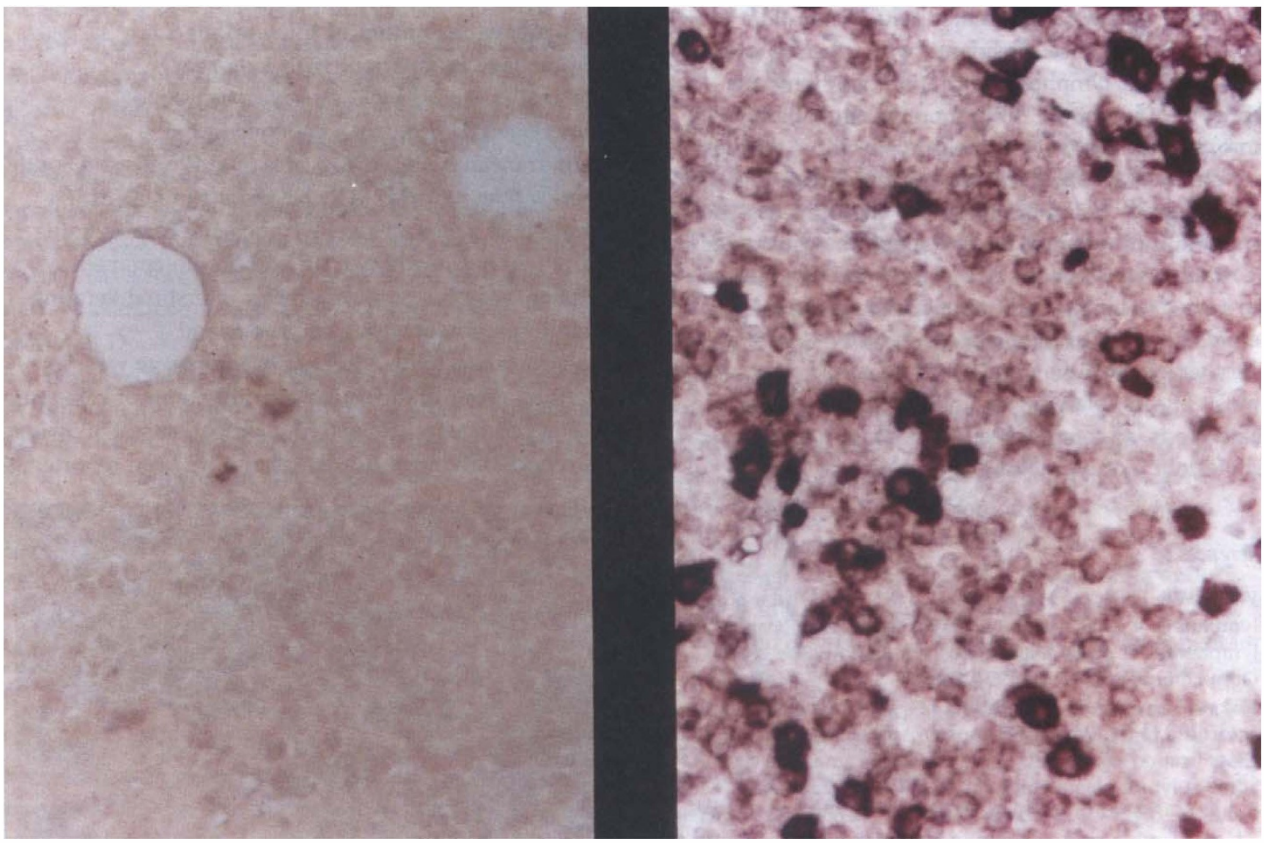


Fig. 2. *MRI scan (axial view) showing a homogeneous mass surrounding the optic nerve but with no globe indentation. There is also diffuse choroidal thickening most marked in the macular region.*



(a)



(b)

Fig. 3. *(a) Histopathology (haematoxylin and eosin) showing monomorphic infiltrate (left) consisting of small lymphocytes with convoluted nuclei with a single nucleolus. Pan B lymphocyte marker CD20 (Dako M0755) is positive (right). (b) Photomicrograph showing negative* in situ *hybridisation for kappa light chains (left) and positive for lambda light chains (right).*

polyclonality, lymphoplasmacytoid differentiation or incomplete light chain restriction of the lymphoid infiltrate.[6–9]

Our patient presented with anterior uveitis, a choroidal mass, no visible epibulbar mass, and subsequently an optic neuropathy with a mass surrounding the optic nerve and thickening of the choroid. Histolopathological examination revealed small monoclonal B cells with slightly angulated nuclear profiles that were light chain restricted. Although we did not biopsy the choroidal component of the mass it is likely that our biopsy of the tumour adjacent to the optic nerve was representative and our patient had a mantle cell lymphoma involving the choroid at presentation. Our patient's tumour resembles the picture of ULI in some respects. The histopathological findings support a diagnosis of lymphoma. Distinction between benign and well-differentiated malignant lymphoid infiltration may be difficult and at times somewhat academic, but attempts to classify these lymphoid tumours are important and may prove useful in terms of treatment planning and prognosis. We are not aware of other literature reports of primary mantle cell lymphoma presenting as a choroidal mass. Interestingly, our patient showed no evidence of anterior epibulbar involvement, which is a common feature of ULI, and did have uveitis, which is not common in the early presentation of ULI but common in large B cell NHL involving the eye.

In summary, this patient with biopsy-proven mantle cell orbital lymphoma presented with a choroidal mass and uveitis. Our case considered with other reported cases of ocular lymphoid infiltration highlights the spectrum of lymphoid infiltration that may involve the choroid from reactive lymphoid hyperplasia, through well-differentiated and mantle cell lymphoma to large cell high-grade lymphoid tumours. The full spectrum of lymphoid infiltration of the eye should be considered in the differential diagnosis of a yellow choroidal lesion. Our patient's tumour did not respond to systemic steroids but proved to be very responsive to orbital radiotherapy, and the patient has remained disease-free almost 3 years following radiotherapy. Long-term follow-up is, of course, essential because a further manifestation of lymphoma may present.

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Sir,

Spontaneous resolution of bilateral macular haemorrhage in a patient with kala-azar

Kala-azar is caused by *Leishmania donovani*, a haemoflagellate protozoan endemic in the Mediterranean basin, Asia, Africa (Old World) and South America (New World).[1] Ocular lesions in kala-azar include post kala-azar uveitis,[2] subacute focal retinitis and retinal haemorrhages,[3] in addition to cutaneous leishmaniasis causing ulcerative or interstitial keratitis, ulcerating granulomatous lid lesions and blepharoconjunctivitis.[1] Retinal haemorrhage was first described as one of the ocular manifestations of kala-azar by Ling and Lee in 1924 in 6 Chinese patients.[3] Following this report, only three further cases have been reported.[4]

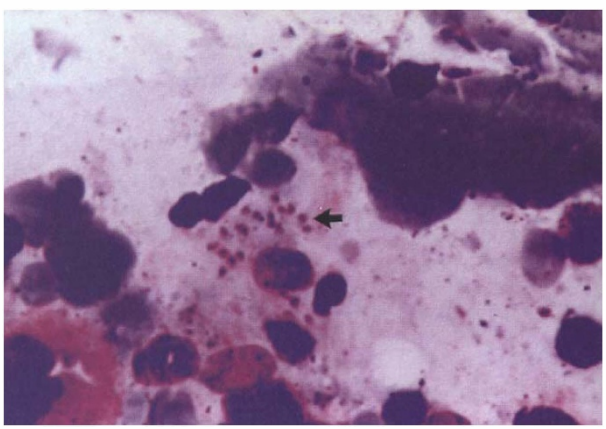

Fig. 1. *Microphotograph showing Leishman–Donovan bodies in bone marrow aspirate (arrow).*