Choroidal ischaemic plaques in sarcoidosis

EDITOR—Sarcoidosis is a multisystem granulomatous disorder of unknown aetiology. It may present with a variety of ocular inflammatory signs, including anterior or posterior uveitis, vitritis, periphlebitis, and choriorretinitis; choroidal lesions may manifest as hypop- or hyperpigmentation.1 We present a patient manifesting multiple placoid choroidal lesions with a previous history of sarcoidosis.

CASE REPORT

A 45-year-old white man presented in July 1994 with a severe bilateral acute anterior uveitis with raised intraocular pressure in the left eye. He also had bilateral punctate epithelial erosions and band keratopathy of his left cornea. Fundal examination revealed multiple placoid areas of choroidal pallor widespread throughout both fundi, but concentrated around the posterior poles (Fig 1). Past medical history included pulmonary consolidation in 1989 at which point bronchial biopsy revealed non-caseating granulomata. He had had an episode of left lacerate retinal ophthalmicus with corneal involvement in 1991. In April 1994 he presented with a recurring, itchy skin rash of oedematous papules on the shoulders, back, chest, and proximal limbs; biopsy again revealed non-caseating granulomata.

He was admitted for investigation and his uveitis treated with topical steroids, central visual acuity returning to normal as this subsided. Intravenous fundangiography was performed with fluorescein (IVF) and indocyanine green (ICG); IVF showed no early masking (Fig 2A) but areas of late staining hyperfluorescence corresponding to the placoid lesions (Fig 2B) and ICG revealed a marked, sustained hypofluorescence in the same areas (Fig 3).

Routine blood testing revealed a marked hypercalcaemia (corrected calcium 3·60 mmol/l) and renal impairment (urea 19·2 mmol/l, creatinine 334 umol/l). He was noted to have a benign paraproteinaemia (IgG type kappa 7 g/l). He was treated with intravenous hydration, diuretics, and corticosteroids and improved rapidly.

Four weeks after presentation his anterior uveitis had cleared and the fundal placoid lesions had begun to show signs of hyperpigmentation.

COMMENT

Ocular sarcoidosis may present with choroidal lesions in 29–50% of cases.1,2 Presentations may vary from patches of creamy yellow depigmentation to a serpiginous appearance extending out from the peripapillary region2,3; and have also mimicked birdshot chorioidopathy.4 The single reported case of serpiginous choriditis associated with sarcoidosis in the literature was marked by early masking and late staining on IVF. Lesions mimicking birdshot chorioidopathy have been described as hypopigmented, flat lesions which manifested late staining on IVF.4 However, fundal distribution was confined mainly to the peripapillary and nasal regions and active investigation of patients with birdshot chorioidopathy has revealed no link with sarcoidosis.

The appearance demonstrated in our patient is more reminiscent of acute posterior multifocal placoid pigment epiretinalopathy, and indeed this has been linked with areas of

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reached as most of the mutations reported to date have only been found in one family. Although ND is believed to be rare, it is likely that many cases remain undiagnosed. The predominant ocular feature of ND—retrolental mass—is not necessarily pathognomonic, and it is clinically difficult to exclude other diseases of defined aetiology, particularly in sporadic and non-syndromic cases. The ongoing molecular assessment of the ND gene will enable us to make definitive diagnoses in cases in which mutations are detected and will also provide useful information for genetic counselling. From the molecular studies to date, we are aware of the marked heterogeneity of pathogenic mutations in the ND gene and, hence, direct sequencing of the gene has to be performed to test for unknown mutations.

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non-perfusion in the choroid as demonstrated on ICG. The lack of early masking on IVP and the sustained hypofluorescence of the lesions on ICG (which is highly protein bound and therefore stays in the circulatory system), suggests that the lesions may represent areas of relative choroidal ischaemia. Previous authors have postulated that focal depigmented chorioretinal areas in sarcoidosis may correspond to subpigment epithelial granulomata similar to Dallen-Fuch's nodules (typically found in sympathetic ophthalmia). It has been suggested that these lesions may compress the choroidal vasculature rather than invade it. However, ocular postmortem examination of patients with fundal sarcoidosis has revealed non-caseating granulomata consisting mainly of epithelioid cells within the choroid, lymphocytic vascular cuffing was closely associated with these lesions. This may indicate a possible mechanism for the apparent vascular insufficiency of the choroid in the angiographic findings in our patient.

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Ossification in choroidal melanoma

EDITOR,—The correct diagnosis of uveal melanoma has improved in recent years following the introduction of superior methods of fundus examination, combined with modern ancillary tests such as fluorescein angiography and ultrasonography (echo-graphy). A percentage of misdiagnosis, however, is to be expected, even in preselected patients undergoing a rigorous screening programme as in the example of the Collaborative Ocular Melanoma Study (COMS) where a misdiagnosis rate of 0.48% has been reported. The diagnostic criteria of uveal melanoma with standardised echography are: (i) solid, dome-shaped or collarstud mass lesion; (ii) low to medium reflectivity; (iii) regular internal structure; and (iv) a variable degree of internal vascularity. Other features include choroidal excavation, associated exudative retinal detachment, extra-scleral/orbital spread, and, less commonly, vitreous haemorrhage. Calcification/ossification 'within' choroidal melanoma mass has not been previously reported, either echographically or histologically. We present such a case.

CASE REPORT
An 85-year-old white man presented with a 3 year history of painless, gradual loss of vision in the left eye. There was no history of eye disease apart from myopia. Systemic inquiry indicated that he was hypertensive receiving diuretic therapy. On examination, the vision was 6/12 partly in the right eye and hand movements in the left with a large inferior scotoma. There was a left afferent papillary defect. The anterior segments and intraocular pressures were normal. Fundus examination on the left showed pigmented cells in the vitreous, a dark, raised, collarstud mass in the superonasal quadrant, and associated retinal detachment involving the macula. Apart from mild atrophic macular changes the right fundus was normal.

B-scan ultrasonography confirmed the collarstud nature of the lesion (Fig 1). The tumour measured 13 mm × 10 mm at base, and 9 mm in height. Vascularity was noted during kinetic examination. An unusual feature was the presence of a highly reflective plaque, located within the tumour at the 'neck' of the collarstud, where the tumour had breached Bruch’s membrane. The plaque caused significant acoustic shadowing, indicating that it was calcified. No extrascleral spread was detected. Standardised A-scan showed the characteristic features of melanoma, except at the area of calcification where a single, high reflective, thick spike was located within the tumour echoes (Fig 2). Marked vascularity was seen appearing as fast short vertical oscillation of the tumour echoes. Systemic examination, haematological and biochemical screen, chest radiography, and liver ultrasonography were all normal. The eye was enucleated.

Pathological examination showed a heavily pigmented mushroom-shaped mass, 10 mm in base diameter and 7 mm in height. The tumour was predominantly of 'spindle B' type with a small amount of epithelioid cells. In the region of the perforated Bruch’s membrane there was an area of fibrosis containing bone, which showed calcification and the presence of osteocytes (Fig 3). This was surrounded by the tumour mass. Apart from Bruch’s membrane at the site of perforation, there was no calcification elsewhere in the eye. The retina was stretched and thinned in parts at the summit of the tumour but was not breached by it. There was no evidence of tumour necrosis or extrascleral spread.

One year after surgery, the patient was alive and well with no evidence of orbital or distant metastasis.

COMMENT
We believe this case to be the first in the literature demonstrating a choroidal melanoma containing a large plaque of calcified bone. Byrne and Green reported a small area of calcification on echography on a melanoma surface, and underlying an area of localised retinal detachment, but not within the melanoma mass itself. Calcification in choroidal melanoma has also been reported.