Malignant melanoma and massive retinal gliosis in phthisis bulbi

Sir,

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Uveal melanoma is the commonest primary intraocular malignancy with an incidence of approximately 5-7 per million per year in the United States and Western Europe.^{1,2} The tumour may be asymptomatic, remaining undetected unless ophthalmoscopy is performed or it may give rise to symptoms due to its location or secondary retinal effects like exudative detachment. It has been long known that a phthisical eye may harbour an occult malignant melanoma, though very few such cases have been reported. Similarly, massive retinal gliosis is an uncommon finding in such eyes. We present a case in which a small uveal melanoma and extensive retinal gliosis were discovered in an eye enucleated for longstanding phthisis. This case re-emphasises the importance of subjecting all enucleated eyes to histopathological

examination, the result of which may influence the further clinical management.

Case report

A 45-year-old woman was referred with a painful and phthisical right eye. She had undergone bilateral surgery for congenital cataracts at the age of about 3 months. Subsequently, she developed phthisis in the right eye and secondary glaucoma in the left eye. She underwent enucleation of the right eye, which was subjected to histopathological examination.

Macroscopic examination showed a collapsed, disorganised eye 12 mm \times 17 mm \times 13 mm in dimension. Microscopic examination revealed a disorganised anterior segment with corneal fibrosis and extensive retro corneal and iridocyclitic membranes. There was supraciliary and suprachoroidal haemorrhage. The vitreous cavity was largely filled by a mass of eosinophilic spindle cells with prominent nuclei. GFAP staining confirmed that these cells were



Figure 1 Histological sections through the malignant melanoma (asterisk) and part of retina in the enucleated eye: (a) stained with haematoxylin and eosin; (b)–(d) stained immunohistochemically for glial fibrillary acidic protein (GFAP—a glial marker), HMB45 (melanoma marker) and a control antibody respectively (red reaction product, haematoxylin counterstain). In (a), the malignant melanoma can be seen as a lightly pigmented plaque-like expansion of the anterior choroid beneath a disorganised, more heavily pigmented, pigment epithelium. The retina is also disorganised and grossly thickened, and contains engorged, partly hyalinised, blood vessels. In (b), the retina can be seen to be replaced by glial tissue. In (c), infiltrating HMB45-positive tumour cells extend anteriorly from the main plaque as well as around pigment epithelial clusters. Calcification (white c) is clearly seen in the adjacent area of massive retinal gliosis. (d) No reaction product is seen in the control section. (All \times 40).

glial in origin, indicating massive retinal gliosis. There was calcification in the retina with some osseous metaplasia at the RPE level and abnormal retinal blood vessels (Figure 1).

The anterior choroid contained a plaque-like proliferation of variably pigmented spindle cell melanocytes. Although the lesion was generally well demarcated, there was local infiltration of adjacent structures (Figure 1). The cells were immunoreactive for HMB45. The melanocytes showed anisonucleosis and occasional prominent nucleoli. These features were consistent with a spindle cell malignant melanoma. No evidence of extraocular extension was seen.

Comments

It has been long known that a phthisical eye may harbour an occult malignant melanoma, though very few such cases have been reported. Sarma *et al*³ reported a case of malignant melanoma in the left eye of a 62-year-old man who had been blind due to trauma for 35 years. Perry *et al*⁴ reported a case of occult choroidal malignant melanoma in an eye with spontaneous expulsive choroidal haemorrhage and suggested that this association may be more than a chance occurrence, inasmuch as both are associated with necrosis of the posterior ciliary arteries.

Undiagnosed malignant melanoma of choroid is uncommon in phthisical eyes. Massive retinal gliosis is also an uncommon finding seen in eyes enucleated for phthisis bulbi. The case reported here shows both these rare features. It has been reported earlier that retinal gliosis may simulate a choroidal malignant melanoma^{5,6} and also that ultrasound reflectivity and magnetic resonance imaging findings of massive retinal gliosis may resemble those of choroidal malignant melanoma. However, to the best of the authors' knowledge, the presented case is the first one to exhibit histological evidence of both malignant melanoma of choroid and massive retinal gliosis. It is possible that factors that lead to massive retinal gliosis in this phthisical eye, also lead to atypical proliferation of melanocytes giving rise to malignant melanoma. One may speculate that necrosis of posterior ciliary arteries was a precipitating factor for suprachoroidal haemorrhage, retinal gliosis and malignant melanoma formation in our case. Whatever the cause, this case once again emphasises the importance of subjecting enucleated eyes to a histopathological examination since the discovery of a malignant melanoma would influence the clinical management of such patients.

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Sir,

Cataract surgery in Senior–Loken syndrome is beneficial despite severe retinopathy *Eye* (2002) **16**, 782–785. doi:10.1038/sj.eye.6700171

A common problem in patients with retinopathy is whether they will benefit from surgical correction of co-existent cataracts.¹ The timing of surgery is dependent upon a clinical assessment of the degree of cataract compared to severity of retinopathy. The case presented here illustrates such a dilemma where cataract surgery exceeded all expectations of predicted benefit in the context of severe, widespread retinopathy.

Case report

A 28-year-old female was referred to our ophthalmic department in 1995. Her optometrist had noted reduced acuity and cataracts. Other medical history of