CLINICAL AND EXPERIMENTAL
OPTOMETRY

CLINICAL PICTURE

Adenoma of the retinal pigment epithelium mimicking ciliochoroidal melanoma

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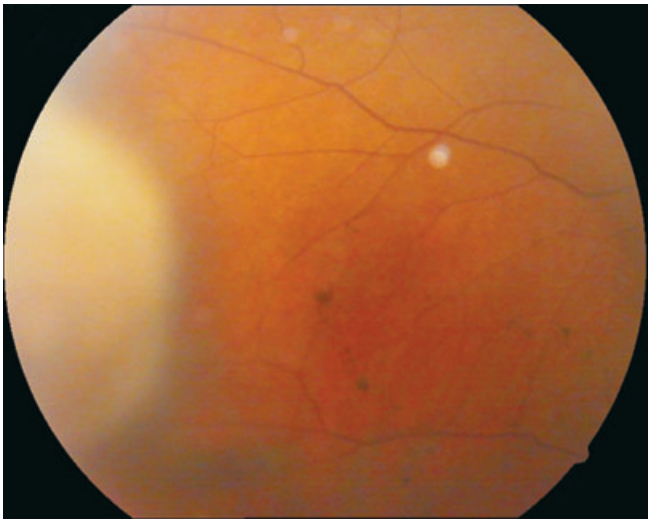

Figure 1. The appearance of the mass and associated scattered pigmentation

Adenoma of the retinal pigment epithelium (RPE) is a rare pigmented intraocular tumour. Without a biopsy it may be difficult to differentiate it from pigmented tumours such as choroidal melanoma for which treatment may include enucleation.

A 44-year-old otherwise healthy man had had visual deterioration in his right eye for six months. Visual acuity was 6/9 OD and 6/6 OS. Applanation intraocular tension and the anterior segment were unremarkable OU. While the left fundus was normal, there was a pigmented mass located in the far temporal periphery of the right fundus with clumps of pigment scattered between the macula and mass (Figure 1). Good quality fluorescein angiographic pictures could not be obtained due to the peripheral location of the mass. Orbital magnetic resonance imaging (MRI) is shown in Figures 2A and 2B. A and B scan ultrasonography showed a solid mass with medium internal reflectivity with a 'stuck on' appearance but without choroidal excavation (Figure 2C). Systemic evaluation and laboratory tests were normal. After receiving a second opinion, which concurred with our presumptive diagnosis of right ciliochoroidal melanoma, the eye was enucleated. Histopathologic examination revealed that the mass was an adenoma of the RPE (Figures 3A and 3B).

A RPE adenoma is a unilateral, solitary dark black tumour that often occurs in adults with no history of prior ocular trauma or inflammation. They often acquire a dilated tortuous retinal feeding artery and a vein during their evolution.[1,2] As an RPE adenoma is initially managed conservatively, differential diagnosis from malignant melanoma is crucial. After re-evaluating the case, we suggested that scattered retinal pigmentation posterior to the mass was relevant to the type of

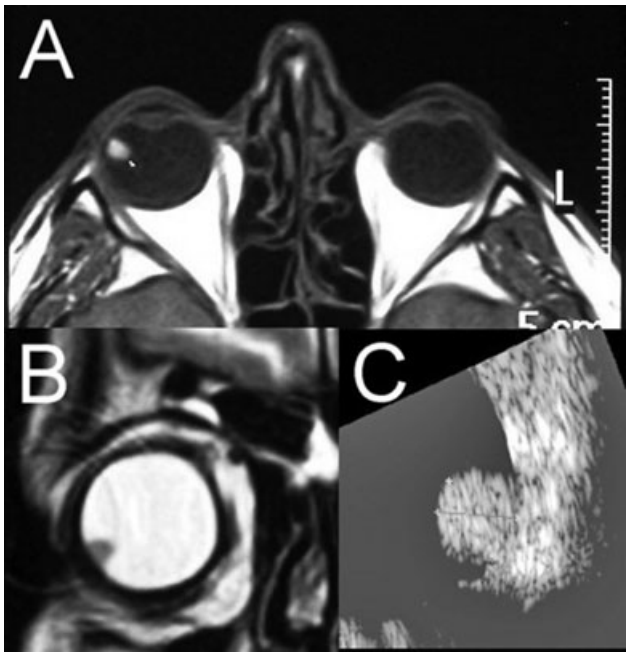

Figure 2. A: Axial cranio-orbital T1-weighted MRI scan showing the hyper-intense tumour. B: The tumour is hypointense on coronal T2-weighted view. C: B-mode ultrasonogram demonstrating the dimensions of tumour.

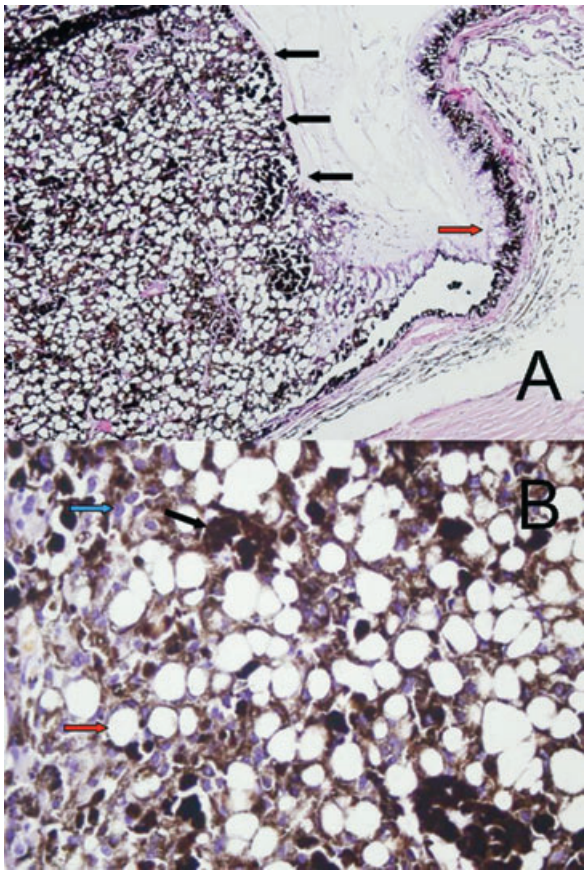

Figure 3. A: Heavily pigmented tumour (black arrows) has a flat base towards the sclera and is continuous with the retinal pigment epithelium (red arrow) (H&E staining, original magnification X10). B: Note pigmented cells (black arrow) and large polyhedral cells with round vesiculated nuclei (blue arrow) and large intracytoplasmic vacuoles (red arrow) (H&E staining, original magnification X40).

tumour. The pigmentation may represent RPE hypertrophy and Shields, Shields and Singh[3] reported five cases with acquired RPE tumours arising from solitary congenital RPE hypertrophy. Ultrasonographic findings were not entirely consistent with melanoma (collar-button appearance or overlying retinal detachment). Fluorescein angiography might have elucidated clinically undetectable feeding vessels and the absence of typical fluorescence pattern might rule out melanoma. Unfortunately, MRI was not very helpful as the imaging properties are similar in malignant melanoma, choroidal haemangioma and RPE tumours.[4]

Our case clearly demonstrates the difficulty of accurate diagnosis of intraocular masses. Every case of presumed choroidal melanoma should be thoroughly evaluated for subtle clues that may prevent misdiagnosis, or unnecessary or even irreversible treatment such as enucleation.

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