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Retinal pigment epithelial tumor in a young Asian female

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PURPOSE. *To report a case of low-grade adenocarcinoma of the retinal pigment epithelium (RPE) in a young Asian woman.*
METHODS. *Interventional case report with histopathologic correlation.*
RESULTS. *A 28-year-old Asian woman with a year-long history of floaters presented with a dark pigmented lesion in the right eye. Partial lamellar sclerouvectomy was performed and histopathology revealed a retina pigment epithelial neoplasm consistent with a low-grade adenocarcinoma. The patient responded well to the surgery with no complications.*
CONCLUSIONS. *Neoplasms of RPE are rare but must be differentiated from choroidal melanoma, as they do not tend to metastasize. Local resection can be a good option for peripheral RPE tumors. (Eur J Ophthalmol 2009; 19: 487-9)*

KEY WORDS. *Eye, Retinal pigment epithelium, Tumor, Adenoma, Adenocarcinoma, Melanoma, Resection*

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INTRODUCTION

Retinal pigment epithelial (RPE) tumors (adenoma and adenocarcinoma) classically appear as round, abruptly elevated, heavily pigmented fundus tumors with retinal invasion (1-6). They can arise de novo or from congenital hypertrophy or hyperplasia of the RPE (5, 6). Several clinical features serve to distinguish this tumor from uveal malignant melanoma, particularly the presence of a dilated retinal artery. The latter feature can be found with adenoma/adenocarcinoma but not melanoma. We report a young Asian woman with a medium-sized pigmented fundus tumor who was referred for enucleation. Detection of a dilated artery contributed to the correct clinical classification of the lesion as an RPE tumor permitting local resection and salvage of the globe.

Case report

A 28-year-old Chinese woman had developed floaters in her right eye 1 year previously. She was diagnosed with choroidal melanoma and referred to us for treatment. Visual acuity was 20/20 in each eye. The left eye was unremarkable. The right anterior segment was quiet but there was moderate anterior vitreous cellularity. Funduscopy disclosed a darkly pigmented mass in the superonasal periphery of the right eye, measuring 8.0 x 8.0 mm in diameter and 6.0 mm in thickness. A prominent, dilated, beaded feeder artery to the mass was found (Fig. 1). Fluorescein angiography (FA) showed that the dilated artery entered the hypofluorescent mass (Fig. 1). Ultrasonography disclosed an echogenic, mushroom shaped mass with high internal reflectivity. Our clinical diagnosis was RPE adenoma/adenocarcinoma, ruling out choroidal melanoma with retinal invasion. The tumor was resected via partial lamellar sclerouvectomy (1).

Microscopic examination revealed that the heavily pigmented tumor was composed of bands of atypical retinal pigment epithelial cells that were separated by PAS-positive fibrous septa. The cytoplasm of the cells contained large round melanin granules including macromelanosomes. Bleached sections disclosed moderate nuclear pleomorphism and prominent nucleoli, but no mitoses were identified. The tumor was diagnosed as a

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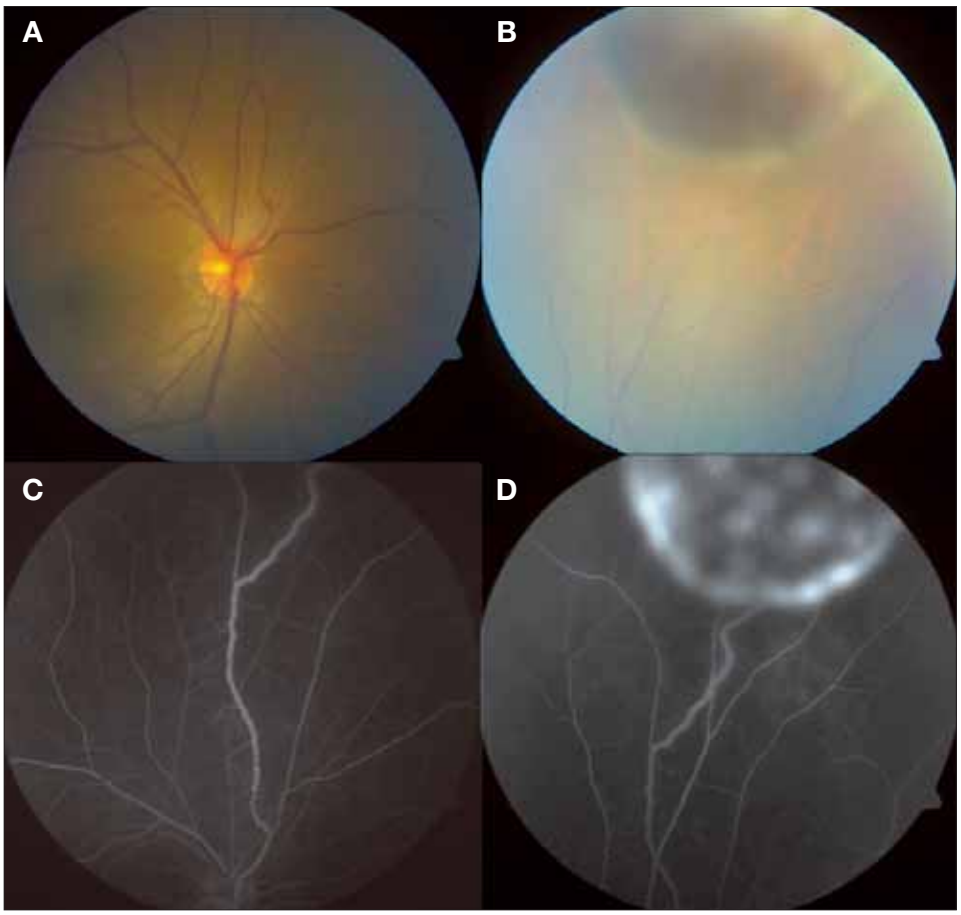

Fig. 1 - *A pigmented fundus tumor was found superonasally in a young Asian woman.* **(A)** *Dilated, tortuous, beaded feeder artery is noted superiorly.* **(B)** *The abruptly elevated, darkly pigmented mass is seen. There was full thickness retinal involvement and the tumor supplied by dilated retinal artery and vein.* **(C)** *Retinal feeding artery in venous phase on fluorescein angiography.* **(D)** *Mild hyperfluorescence of the mass in late phase fluorescein angiography.*

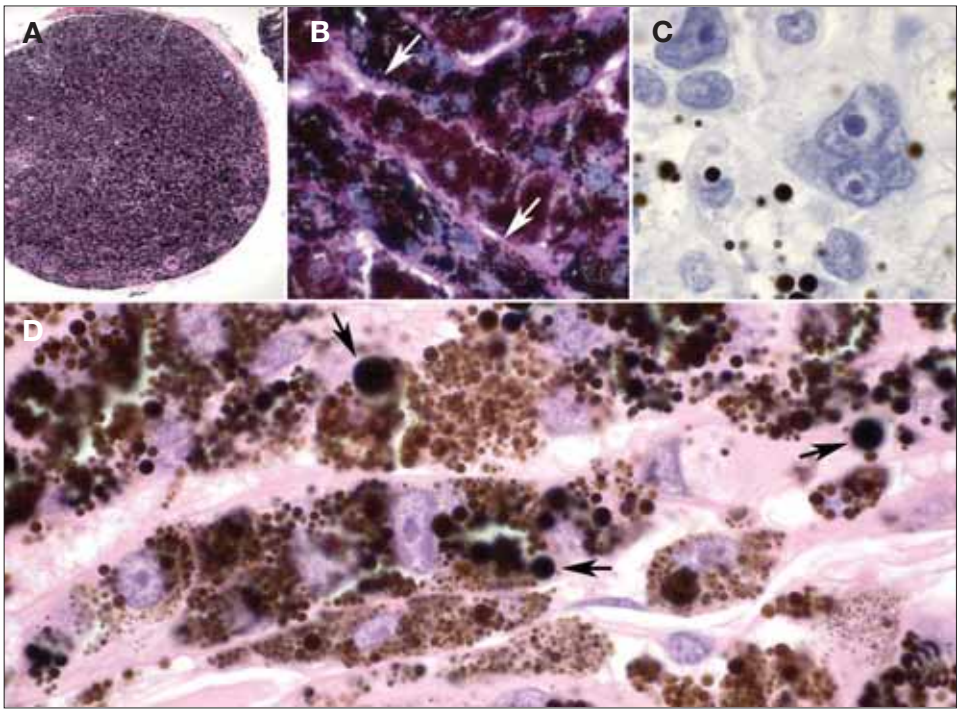

Fig. 2 - *Histopathology, retinal pigment epithelium (RPE) adenocarcinoma.* **(A)** *Heavily pigmented RPE tumor with rounded contours (seen at low magnification in inset* **(B)** *is composed of atypical epithelial cells filled with round pigment granules including macromelanosomes (black arrows). Bands of cells rest on fibrous septa (white arrows) highlighted by PAS stain in inset* **C**. *Atypical nuclei with prominent nucleoli are evident in depigmented section (inset* **D**). *Some of the macromelasomes are incompletely bleached. Main figure* **(A)**, *H&E x400, inset* **B** *H&E x10, inset* **C** *PAS x100, inset* **D** *bleach x400.*

low-grade adenocarcinoma of the RPE based on nuclear characteristics (Fig. 2). One month following surgery, visual acuity was 20/60 and the retina was flat with an atrophic chorioretinal scar at the site of the previous tumor.

DISCUSSION

Tumors of the RPE are rare and comprise benign and malignant lesions including massive RPE hyperplasia, congenital simple hamartoma, combined hamartoma, and adenoma and adenocarcinoma. RPE adenoma/adenocarcinoma is exquisitely rare and classically appears as a heavily pigmented fundus mass, occasionally with surrounding subretinal fluid and exudative retinopathy. In the analysis of 13 cases by Shields and coworkers, the mean patient age was 53 years and the tumor was located most often in the peripheral fundus (n=6, 46%) (2). Associated clinical features included subretinal fluid (n=4, 30.7%), epifoveal membrane (n=2, 15.3%), macular scar (n=1, 7.6%), and macular hole (n=1, 7.6%). Retinal exudation, epifoveal membrane, and macular hole are features rarely associated with uveal melanoma. A particularly important feature of RPE adenoma/adenocarcinoma is the occasional presence of a dilated retinal feeder vessel, a feature very rarely found with melanoma. Shields and associates found that 8 of 13 (61.5%) RPE tumors had dilated retinal vessels. Dilated retinal veins are observed rarely when melanoma invades the retina, but, to our knowledge, melanoma has not been found to manifest a dilated retinal artery. Uveal melanoma's blood supply arises from the luxuriant choroidal vasculature, whereas RPE tumors are supplied by the retinal vasculature.

Fine needle aspiration biopsy can be used to differentiate RPE adenoma/adenocarcinoma from melanoma (1), but this determination can be difficult even for experienced cytopathologists. In our case, tumor resection was an option so we avoided needle biopsy and obtained histopathology. Most RPE tumors require therapy because progressive growth can produce visual loss and destroy the eye (1-6). Radiotherapy has been employed in cases when there are no other suitable therapeutic options, but it does not appear to be highly effective (2, 6). The peripheral location of the tumor in the case reported here was quite amenable to local resection, which led to the avoidance of long-term radiation complications.

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