Adenoma of the Ciliary Pigment Epithelium with Diffuse Iris Pigment Dispersion

Ying Chang[1], Wen-Bin Wei[2], Xiao-Lin Xu[1], Xiu-Qian Yi[1], Hai-Xia Bai[1], Bin Li[1]

[1]Beijing Institute of Ophthalmology, Beijing Tongren Eye Center, Beijing Tongren Hospital, Capital Medical University; Beijing Ophthalmology and Visual Sciences Key Laboratory, Beijing 100005, China

[2]Beijing Tongren Eye Center, Beijing Tongren Hospital, Capital Medical University; Beijing Ophthalmology and Visual Sciences Key Laboratory, Beijing 100730, China

To the Editor: Adenoma of the ciliary pigment epithelium (CPE) is an exceedingly rare tumor that is difficult to clinically differentiate from malignant melanoma.[1] Herein, we report a case of an adenoma of the CPE in Chinese women and discuss the clinical features of this unusual tumor.

A 57-year-old woman complaining of obviously visual deterioration vision in her right eye for 1-year presented to Beijing Tongren Hospital in December 2013. There was no previous history of ocular trauma or intraocular inflammation. On examination, the visual acuity was 0.2 in the right eye and 1.2 in the left eye. Intraocular pressure readings were 23.8 mmHg and 14.6 mmHg in the right and left eyes, respectively. Slit-lamp examination revealed diffuse fine pigmentosus keratic precipitates on the posterior corneal surface and diffuse pigment dispersion on the iris in the right eye. There was no visible iris neovascularization and iridodonesis. After mydriasis, a dark brown mass occupied the superotemporal posterior chamber, and a segmental cataract was present with amber color [Figure 1a]. The left eye was entirely normal. Bilateral fundus was normal by indirect ophthalmoscopy.

Ultrasonography (US) showed an acoustically solid, abruptly elevated, hillock-shaped mass, measuring 9.6 mm × 7.6 mm (basal) × 5.4 mm (apical thickness) [Figure 1b]; ultrasound biomicroscopy demonstrated a moderately irregular acoustically solid in 10:30–13:30 o'clock meridians in the ciliary body, measuring 9.89 mm × 6.96 mm × 4.57 mm. The lesion had invaded the structure of chamber angle and the root of the iris [Figure 1c]; magnetic resonance imaging (MRI) showed that the mass was hyperintense in T1-weighted images and hypointense in T2-weighted images to the brain [Figure 1d]. The tumor showed enhancement after gadolinium.

The patient underwent local resection of the tumor in the form of iridocyclectomy. Histopathologically, the tumor showed heavy pigmentation and nests of pigmented epithelial cells forming gland-like structures that were separated by vascularized fibrous connective tissue [Figure 1e]. They contained numerous characteristic round or oval clear vacuoles, encompassed by cells whose cytoplasm was replete with large, spherical melanosomes [Figure 1f]. No mitoses were seen. The tumor was diagnosed as adenoma of the pigmented ciliary epithelium.

The CPE as a stretch from the retina pigment epithelium can sometimes undergo reactive proliferation and migration. However, true tumors of the pigment epithelia of the ciliay body are extremely rare.[1] A patient with an adenoma of the CPE may be asymptomatic, or impairment of visual acuity due to secondary cataract or subluxation of the lens may develop.[2] Angle closure was responsible for secondary glaucoma. Dinakaran *et al*.[3] reported that smaller tumors of adenoma of the CPE invaded the angle, and larger lesions caused extensive pigment dispersion although noninvasively. The tumor size in our patient was larger than the reported. It not only invaded the angle but also caused extensive pigment dispersion. Diffuse iris pigment dispersion and the dark pigmentation let us to be thought it could be a benign tumor, a necrotic melanocytoma or an adenoma of the pigment epithelium.

In general, adenoma of the CPE shows medium to high internal reflectivity and acoustic solidity with an abruptly elevated in US and displays hypointense in T2-weighted images to the brain and enhancement after gadolinium in MRI.[4] Adenoma of the CPE appears deeper color than melanoma. It lacks the surrounding pigmented base or collar-button configuration that is usually seen in melanomas. Prominent tumor blood vessels, frequently noted in melanoma, are rarely seen in adenoma of the CPE.[1] In addition, acoustic hollowness are more often seen

Address for correspondence: Prof. Bin Li, Beijing Institute of Ophthalmology, Beijing Tongren Hospital, Capital Medical University, 17 Hougou Street, Chong Wen Men, Beijing 100005, China
E-Mail: libin43_99@163.com

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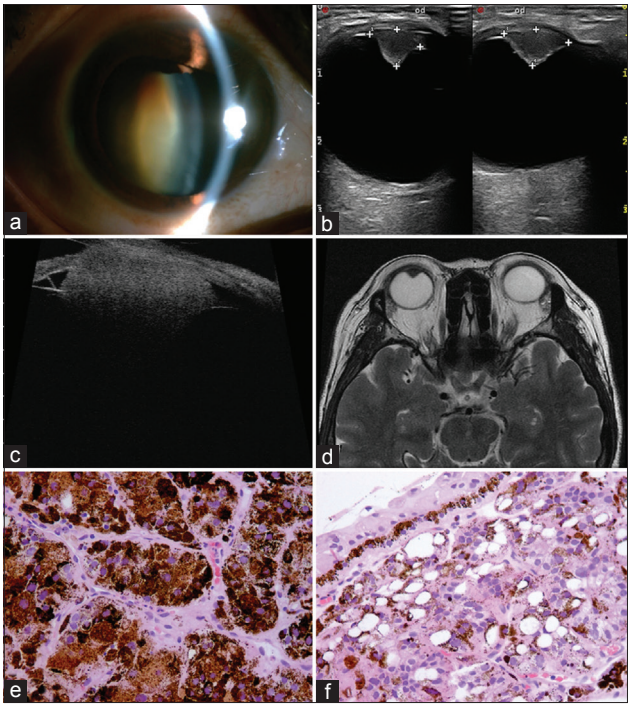

Figure 1: (a) A dark brown mass and a segmental cataract with amber color; (b) Ultrasonography showed an acoustically solid, abruptly elevated, and hillock-shaped mass; (c) Ultrasound biomicroscopy demonstrated a moderately irregular acoustically solid and the lesion invaded the structure of chamber angle and the root of the iris; (d) Magnetic resonance imaging showed that the mass was hypointense to the brain in T2-weighted images; (e) Histopathologically showed heavy pigmentation and nests of pigmented epithelial cells forming gland-like structures; (f) Numerous characteristic round or oval clear vacuoles (H and E, original magnification, ×200).

with melanomas since rapid growth forming cystoids in the tumor. Ciliary body melanocytoma is also a deeply pigmented solid mass and more likely to display pigment dispersion and secondary glaucoma due to undergo central necrosis. However, it arises from the uveal stroma with a rough or corrugated surface and no contrast enhancement in MRI with gadolinium. In some cases, the clinical differentiation adenoma of the CPE may be impossible. Histopathology is still the only standardization for a diagnosis. Tumors that arise from the CPE are usually characterized by prominent clear vacuoles and density of melanin granules.[5]

Recognition of the characteristic features of adenoma of the CPE should facilitate the diagnosis of this unusual tumor. This benign tumor should be observed periodically, and local tumor resection should be considered if there is progressive growth or secondary glaucoma.

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Conflicts of interest

There are no conflicts of interest.

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