A Case of Pseudoadenomatous Hyperplasia of Ciliary Body Epithelium Successfully Treated by Local Resection

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OHGURO, H., MARUYAMA, I. and NAKAZAWA, M. *A Case of Pseudoadenomatous Hyperplasia of Ciliary Body Epithelium Successfully Treated by Local Resection.* Tohoku J. Exp. Med., 2002, 197 (1), 41-45 — A case of pseudoadenomatous hyperplasia of ciliary body epithelium was reported in which malignant melanoma of ciliary body was suspected. Partial resection for histopathology was performed in conjunction with cataract extraction, anterior resection and photocoagulation. Histopathology of the tumor identified as pseudoadenomatous hyperplasia of ciliary body epithelium. Partial resection of ciliary body tumor may be an alternative method for its differential diagnosis rather than enucleation and iridocyclectomy. ——— Pseudoadenomatous hyperplasia; ciliary body; partial resection; malignant melanoma; differential diagnosis

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Pseudoadenomatous hyperplasia of ciliary body epithelium is rare tumor that may cause subluxation of the lens, segmental cataract, or secondary glaucoma (Shields and Shields 1992; Shields et al. 1996). Although this tumor is more likely to display an irregular surface, to transmit light well on transillumination, and to show high internal reflectivity on ultrasonography (Pavlin et al. 1991, 1992; Pavlin and Foster 1995) in contrast to malignant melanoma of ciliary body, it is very difficult to differentiate pseudoadenomatous hyperplasia, adenoma, malignant melanomas or metastatic tumors of the ciliary body (McLean et al. 1977;

Shammas and Blodi 1977; Shields and Shields 1992; Sahel et al. 1993; Shields et al. 1996). Therefore, enucleation or iridocyclectomy is usually performed for the differential diagnosis and treatment (Shields et al. 1991; Naumann and Rummelt 1996).

Herein, we describe a patient with pseudoadenomatous hyperplasia of ciliary body epithelium with segmental cataract, in which malignant melanoma was strongly suspected preoperatively due to the relatively rapid growth of tumor and rapid deterioration of the cataract. Histopathological examination of partially dissected tumor after lensectomy, ante-

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rior vitrectomy, and endophotocoagulation was effective for the differential diagnosis.

CASE REPORT

A 53-year-old woman was found to have a ciliary body tumor with segmental cataract in her left eye by a near ophthalmologist and then referred to our hospital for further evaluation and management of the tumor. She had no history of systemic disease including malignant tumors. Best-corrected visual acuities were 1.0 OD and 0.1 OS at the initial examination in our institute. Intraocular pressures were 18 mm Hg in both eyes. Ophthalmic examinations revealed no abnormalities in the right eye. Slit lamp examination of her left eye showed a pupil distortion with high peripheral anterior synechia superotemporally from 0 to 3 o'clock position. When the pupil was dilated, a tumor mass arised from ciliary body and segmental cataract adjacent to the tumor were observed. Gonioscopic examination revealed a opaque white solitary mass with vessels on the ciliary body between the 2- and 3-o'clock positions (Fig. 1). Indirect funduscopy examination revealed no abnormalities in the left eye. Fluorescein angiography revealed early filling

and late diffuse staining of the mass, but was not recognized leakage to the anterior chamber. Ultrasound biomicroscopy disclosed that a mass measured about 4 mm in diameter and was developing from the ciliary body region extending to the retrolental space and posterior chamber with iris invasion (Fig. 2). In the examinations by CT scan and ultrasound B-mode scan showed no calcification within the tumor and a regular reflectivity pattern at the base of the mass, respectively. Three months after the initial examination, her left visual acuity was worse to 0.01 and segmental cataract significantly developed. Based upon these clinical findings, a nonpigmented malignant melanoma of the ciliary body or adenoma of the non-pigmented ciliary epithelium was strongly suspected. The tumor was partially resected after cataract extraction (phacoemulsification), anterior vitrectomy and photocoagulation around the tumor. Histopathological examination revealed that the mass composed of small nodular lesions abundant of eosinophilic extracellular material, and pigmentation was mainly recognized between the nodular lesions (Fig. 3). Immunocytochemical study showed negative staining of melanoma or metastatic

Fig. 1. Gonioscopic examination shows a reflected image between 7 and 9 o'clock of the ciliary tumor, which is located 1 and 3 o'clock positions.

Fig. 2. Ultrasound biomicroscopy shows that the tumor was developing from the ciliary body region extending to the retrolental space and posterior chamber with iris invasion.

CB, ciliary body; I, iris; T, tumor.

Fig. 3. Histopathology of the tumor.

Hematoxylin-Eosin staining and immunofluorescence labeling by anti-vimentin and anti-actin antibodies (1:100 dil) of the tumor are shown. Scale bar=50 μ m. Immunofluorescence staining using paraffine embedded section of the tumor was performed as described by Maruyama et al. (2000).

tumor specific antigen (HMB-45, S-100, cytokeratin, p 53), but positive of vimentin and smooth muscle actin (Fig. 3). Based upon these observations, this tumor was identified as pseudoadenomatous hyperplasia of ciliary body epithelium.

DISCUSSION

In terms of differential diagnosis of ciliary body tumors, including melanoma, pseudoadenomatous hyperplasia, adenocarcinoma, and metastatic tumors, some clinical

characteristics may help distinguish a primary melanoma from an adenoma including yellow tan color, confinement of the tumor to the ciliary body, failure of the tumor to block transillumination, and high internal reflectivity demonstrated by ultrasound (Pavlin et al. 1991, 1992; Pavlin and Foster 1995) in the latter. However, it may be very difficult to differentiate a metastasis from primary ciliary tumors, although multifocality, an associated inflammatory reaction, and dilated episcleral blood vessels may be more apparent in the metastasis (Shields and Shields 1992; Shields et al. 1996). It is also clinically indistinguishable from amelanotic malignant melanomas of the ciliary body or metastatic carcinomas (McLean et al. 1977; Shammus and Blodi 1977; Shields and Shields 1992; Sahel et al. 1993; Shields et al. 1996).

In our present case, relatively rapid development of size of the tumor and related segmental cataract suggested that her tumor was a malignant melanoma of ciliary body (McLean et al. 1977; Shammus and Blodi 1977; Sahel et al. 1993). Histopathological observations of the partially resected tumor including immunohistochemical characteristics showed that her tumor was diagnosed as pseudoadenomatous hyperplasia of ciliary body epithelium (Fuchs adenoma). Pseudoadenomatous hyperplasia of ciliary body epithelium occurs in approximately 20% of eyes examined after death (Ilf and Green 1972) and consists of small, usually less than 2 mm in diameter, pedunculated, counter-sunk or buried nodules of ciliary body non-pigmented epithelium entrenched in a considerable amount of basement membrane material, which stains positive with periodic acid-Schiff (Shields and Shields 1992; Shields et al. 1996).

It was reported that most patients with acquired neoplasms of the non-pigmented ciliary body epithelium were treated by enucleation, because they were suspected clinically to have malignant melanoma, and local resection of the tumor was used infrequently (Shields et

al. 1991; Naumann et al. 1996). Shields et al. (1991, 1996; Shields and Shields 1992) reported nine patients with non-pigmented ciliary body epithelium tumors treated by local resection (partial lamellar sclerouvectomy), and good prognosis and good vision in the affected eyes. In the present case, because she had dense cataract at the operation, we planned to first remove cataract and anterior vitrectomy, then tumor was partially removed after photocoagulation of basement of the tumor due to avoid massive bleeding. Then if the tumor was malignant, we thought perform partial lamellar sclerouvectomy or enucleation, but her tumor was benign tumor. Therefore, our method may be an alternative option to treat non-pigmented ciliary body tumors rather than more extended surgical methods, partial lamellar sclerouvectomy or enucleation.

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