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Recurrent Mesectodermal Leiomyoma of the Ciliary Body : A Case Report

A 19-yr-old woman with a previous history of a mass of the right ciliary body presented with a decreased visual acuity of right eye. Clinicoradiologic examinations suggested a recurrent mass of the ciliary body. Enucleation of the right eye was performed under the impression of malignant tumor. On microscopic examination, the tumor was a mesectodermal leiomyoma of the ciliary body. On immunohistochemistry, the tumor cells were reactive to smooth muscle actin and vimentin, but not reactive to cytokeratin, S-100 protein, neurofilament, desmin, epithelial membrane antigen, HMB-45, glial fibrillary acidic protein, and synaptophysin. Electron microscopy revealed numerous thin longitudinally placed myofilaments and focal densities in the cytoplasms. In the review of the literature, only 27 cases of mesectodermal leiomyoma of the ciliary body were reported, however, there was no report of recurrent cases. Mesectodermal leiomyoma should be differentiated from other orbital spindle-cell tumors such as amelanotic melanomas and glial tumors. Immunohistochemical and electron microscopic studies may be useful for the correct diagnosis by showing smooth muscle differentiation in the tumor cells.

Key Words : *Leiomyoma; Ciliary Body*

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INTRODUCTION

Smooth muscle tumors of the ciliary body are extremely rare. Jakobiec et al. insisted that the smooth muscle tumor of the ciliary body constitutes a new nosologic entity of myogenic neoplasia in 1977 (1). Embryologically, the ciliary muscle originates from the neural crest (mesectoderm). So they proposed the new term, 'mesectodermal leiomyoma'. Until now, 27 cases have been reported in the literature. We report a case of recurrent mesectodermal leiomyoma of the ciliary body in a 19-yr-old Korean woman.

CASE REPORT

A 12-yr-old Korean girl was referred to the ophthalmology department due to gradual decrease of visual acuity of right eye for several months. She was born premature at 34 weeks of gestational age with 1.75 kg of birth weight. Her elder brother died of congenital heart disease. On ophthalmologic examination, a bullous lesion was noted in the right ciliary body. Funduscopic examination showed retinal detachment. Magnetic resonance imaging (MRI) revealed a $1.5 \times 1 \times 1$ cm-sized, ovoid intraocular mass on upper outer quadrant of the orbit. Radiologic impression included melanoma, choroidal hemangioma, medulloepithelioma, and retinoblastoma. Excisional biopsy of the mass was performed and we examined several fragmented tissues from mass. Histologically, the tumor was composed of relatively monotonous spindle-shaped cells and the fibrillary stroma with fine blood vessels. The tumor cells had round to oval nuclei with indistinct cellular border and vacuolar changes. There was no obvious nuclear atypia. Immunohistochemical study with antibodies for S-100 protein, synaptophysin, neurofilament, epithelial membrane antigen (EMA), glial fibrillary acidic protein (GFAP), and smooth muscle actin (SMA) was not contributory to the diagnosis due to nonspecific staining. The pathologist suggested the possibilities of glial tumor or smooth muscle tumor.

Seven years after the initial operation, the patient visited the ophthalmology department again due to progressive decrease of visual acuity of right eye. Ophthalmologic examination and computed tomography (CT) revealed regrowth of the tumor at the same region (Fig. 1). Enucleation of the right eyeball was performed under the clinical diagnosis of glioma or leiomyosarcoma. Grossly, a well demarcated, round to ovoid tumor was noted in the anterolateral portion of the ciliary body and measured $2.0 \times 1.5 \times 1.2$ cm in size. The retina was shifted upwardly by the tumor. The cut surface of the tumor showed homogeneous yellowish white, firm and solid appearance (Fig. 2). Microscopically, the tumor was well demarcated and covered by the ciliary epithelium. The transitional area from normal ciliary smooth muscle was noted (Fig. 3). The tumor

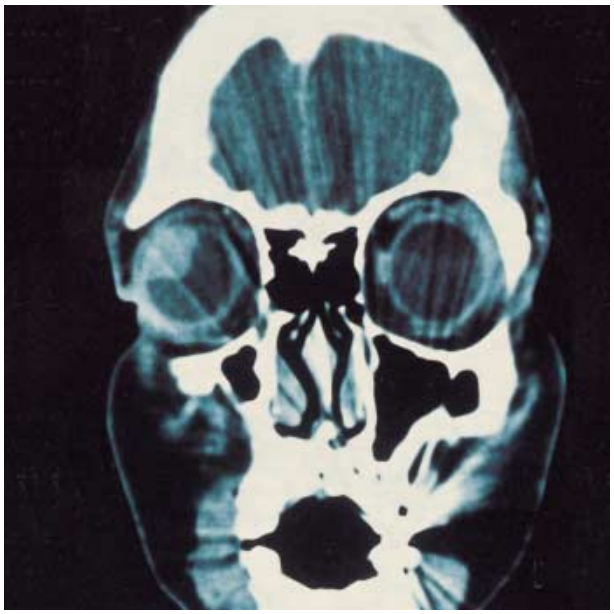

Fig. 1. Facial computed tomography, coronal view: a well demarcated ovoid mass is noted in the right eye.

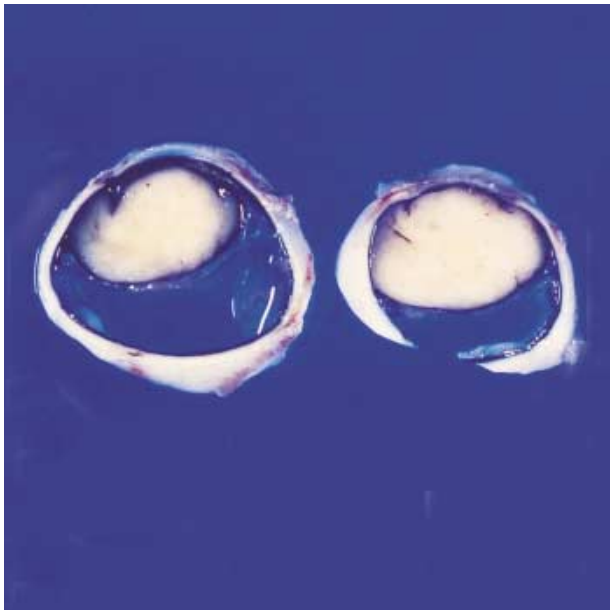

Fig. 2. A pale gray solid firm mass is located in the anterolateral portion of the ciliary body.

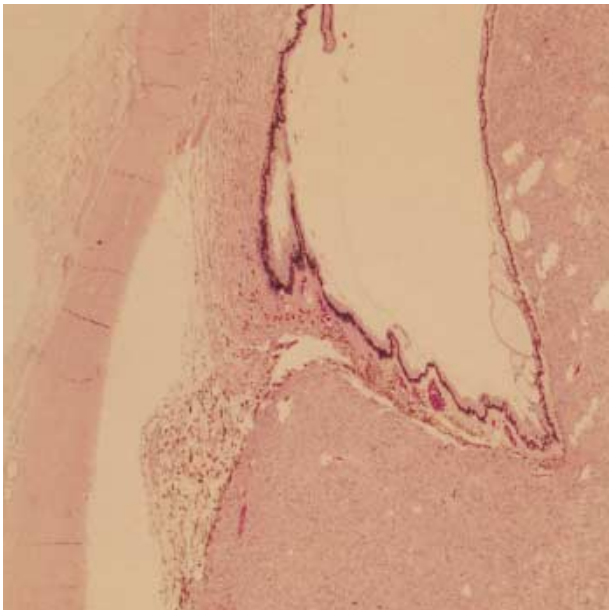

Fig. 3. The tumor is covered by the ciliary epithelium and mixed with normal ciliary smooth muscle (H&E stain, ×40).

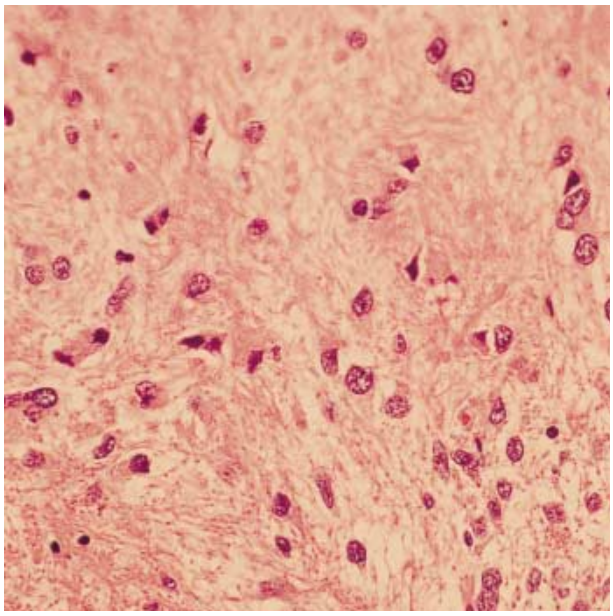

Fig. 4. Polygonal tumor cells in the fibrillary stroma are noted (H&E stain, ×200).

cells were polygonal to spindle in shape and had abundant eosinophilic, fibrillary cytoplasms (Fig. 4). The cellularity was moderate. There was no nuclear atypia or mitosis. Immunohistochemically, the tumor cells were reactive to vimentin and SMA (Fig. 5). They were negative for cytokeratin, S-100 protein, neurofilament, desmin, EMA, HMB-45, GFAP, and synaptophysin. Electron microscopy (EM) revealed abundant mitochondria, numerous longitudinally placed thin myofilaments, and focal densities in the cytoplasms (Fig. 6). The nuclear membrane showed mild degree of irregularity. The nucleus showed inconspicuous nucleoli and chromatin clumping.

DISCUSSION

The mesectodermal leiomyoma is a rare variant of the benign smooth muscle tumor, which microscopically resembles a neurogenic rather than a myogenic tumor. The term mesec-

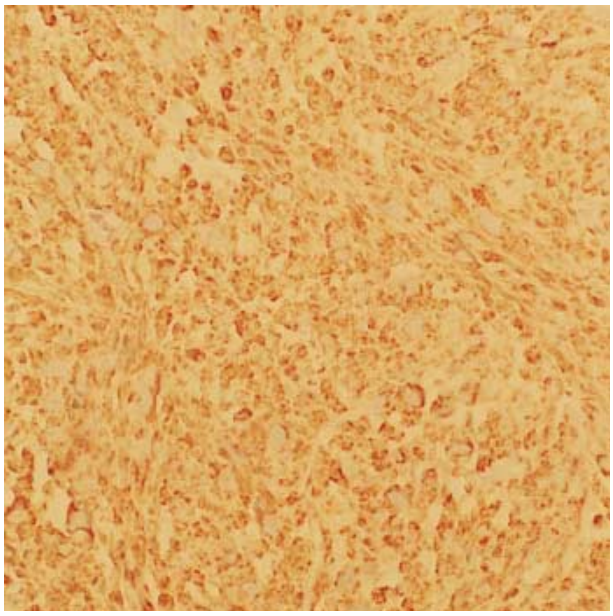

Fig. 5. Immunohistochemical stain for smooth muscle actin reveals positive reaction (× 100).

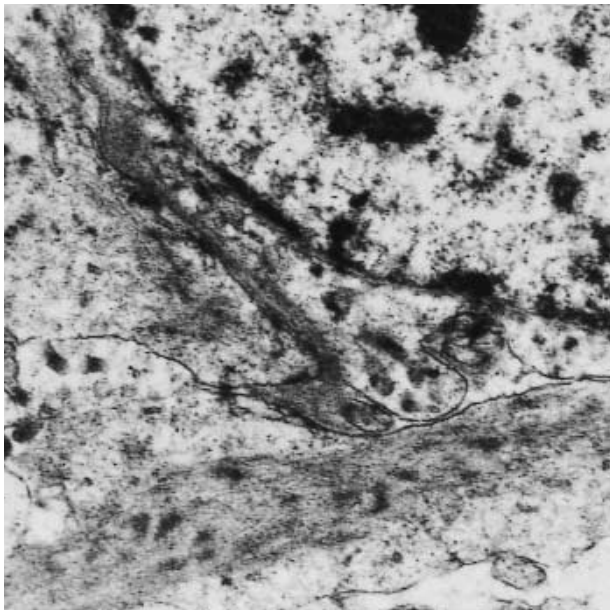

Fig. 6. Electron microscopy reveals longitudinally placed thin filaments and focal densities in the cytoplasm (× 10,000).

Table 1. Summary of leiomyomas of the ciliary body reported in the literature

Author (Reference)	Year	Age (yr)	Sex	Race	Side	Size (mm)	Operation	Clinical Impression	Follow up
Blodi (2)	1950	40	F	NA*	Rt	NA	Enucleation	NA	NA
Dunbar (3)	1956	49	F	NA	Lt	7×5	Enucleation	NA	NA
Bonameur et al. (4)	1957	39	F	NA	NC	NA	NA	NA	NA
Meyer et al. (5)	1968	50	F	NA	Rt	9×7	Enucleation	NA	NA
Lowe & Greer (6)	1970	24	F	NA	Rt	8×6×5	Resection	NA	NA
Calmettes et al. (7)	1971	25	F	NA	Lt	NA	Enucleation	NA	NA
Jakobiec et al. (1)	1977	37	F	NA	Lt	6×5×5	Resection	metastasis, melanoma	Disease free 6 yrs
Jakobiec et al. (1)	1977	20	F	NA	Rt	9×7×2	Enucleation	melanoma	NA
Jakobiec & Iwamoto (8)	1978	28	F	NA	Rt	NA	Enucleation	melanoma	NA
Vogel et al. (9)	1978	55	F	NA	Rt	8×8×3	Resection	NA	NA
Gloor et al. (10)	1979	12	F	NA	Rt	NA	Resection	NA	NA
Sautter et al. (11)	1979	23	F	NA	Lt	6×6	Resection	NA	NA
Croxatto & Malbran (12)	1982	23	F	White	Lt	7×5×5	Resection	melanoma, cyst	NA
Orsoni et al. (13)	1985	18	F	NA	Rt	8×5	Enucleation	uncertain	NA
Takagi et al. (14)	1985	38	F	Japanese	Lt	13×7×5	Enucleation	malignant tumor	NA
Burk et al. (15)	1989	63	F	NA	NA	NA	NA	NA	NA
Ishigooka et al. (16)	1989	28	F	Japanese	Rt	9×8×3	Resection	neurogenic or glial tumor	NA
White et al. (17)	1989	38	M	White	Rt	8×7×5	Resection	melanoma	NA
Yu et al. (18)	1990	8	M	White	Lt	NC	Resection	NA	NA
Shields et al. (19)	1994	80	F	White	Lt	4×4×3	Resection	melanoma, leiomyoma	Death by another cause (2 yrs)
Shields et al. (19)	1994	11	F	White	Rt	14×12×9	Resection	melanoma	Disease free (5 yrs)
Shields et al. (19)	1994	29	F	NC	Rt	NA	Resection	atypical staphyloma	Disease free (4 yrs)
Shields et al. (19)	1994	20	F	NC	Rt	16×14	Resection	leiomyoma	Disease free (4 yrs)
Shields et al. (19)	1994	68	M	White	Lt	15×10	Enucleation	melanoma	Disease free (3 yrs)
Shields et al. (19)	1994	54	F	White	Lt	9×9×4	Resection	melanoma, leiomyoma, neurilemmoma	NA
Shields et al. (19)	1994	24	M	White	Lt	13×12×8	Resection	melanoma, leiomyoma, neurilemmoma	NA
Campbell et al. (20)	1997	47	F	NA	NA	NA	NA	NA	NA
Present case	2003	19	F	Korean	Rt	20×15×12	Enucleation after resection	melanoma, glioma	Recurrent 7 yrs after resection

*NA: not available.

todermal leiomyoma was suggested by Jakobiec et al. in 1977 (1). Embryologically, the cells of the neural crest that contribute to the formation of bone, cartilage, connective tissue, and smooth muscle in the regions of the head and neck have been called mesectoderm (1). They proposed that the unusual neural appearance of ciliary leiomyoma was a reflection of their probable origin from the mesectodermal smooth muscle of the ciliary body.

Mesectodermal leiomyomas resemble ganglionic, astrocytic, and peripheral nerve tumors because of their fibrillary neurogenic appearance. Therefore, histologically, the differential diagnosis of this unusual tumor includes melanoma, glioma, peripheral nerve tumor, or paraganglioma. We initially diagnosed this tumor as glial tumor or smooth muscle tumor. The final diagnosis of a mesectodermal leiomyoma was supported by the electron microscopic demonstration of thin filaments with focal densities and smooth muscle actin reactivity by immunohistochemistry.

To our knowledge, 27 cases of mesectodermal leiomyoma in the ciliary body have been reported in the literature (Table 1). Twenty-three patients were female and four were male. Thirteen cases were right, eleven were left, and three were unknown. The tumor size ranged from 0.4 to 1.6 cm. Enucleation was performed in 9 cases and resection was performed in 15 cases. The type of operation was not documented in 3 cases. Among seven cases with follow-up data, there was no recurrence or metastasis. The maximum follow-up period was 6 yr. In the present case, the tumor was 2 cm in its greatest dimension, which is the largest one in the reported cases and is the only case that recurred after simple resection, which was not iridocyclochoroidectomy but just local resection by curettage.

Although the tumor in the present case recurred because the initial tumor removal was incomplete, it was thought to be benign on light microscopic appearance because of its well circumscription, moderate cellularity, and lack of mitosis. Based on our literature review, the mesectodermal leiomyoma can exhibit slowly progressive enlargement and can produce a large mass with complications that may require enucleation. However, in most reported cases, the local resection of the tumor such as modified partial lamellar sclerouvectomy was performed. Enucleation seems inappropriate for this tumor, and radiotherapy generally has little effect on benign tumor.

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