Recurrence 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. Clinico-radiologic 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 cytoplasm. 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.

Keywords: 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 × 1 × 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 and electron microscopic studies may be useful for the correct diagnosis by showing smooth muscle differentiation in the tumor cells.
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   | NA   | NA        | Enucleation      | 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                        |
| Crozatto & 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| Resection        | malignant tumor            | NA                        |
| Burk et al. (15)    | 1989 | 63       | F   | NA   | NA   | NA        | Resection        | 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 staphylcoma      | 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   | Resection        | 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        | Enucleation after resection | 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.
Recurrent Mesectodermal Leiomyoma

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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