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Mesectodermal Leiomyosarcoma of the Ciliary Body

Sun-Won Park, Hyung-Jin Kim, Hee-Seung Chin, Kee-Sun Tae, and Jee-Young Han

Summary: We report a case of low-grade mesectodermal leiomyosarcoma of the ciliary body, an extremely rare tumor of neural crest origin, occurring in a 12-year-old boy. MR imaging showed a well-margined, ovoid soft tissue mass at the right temporal ciliary body, accompanied by total retinal detachment. The mass was slightly hyperintense relative to contralateral vitreous on T1-weighted images and markedly hypointense on T2-weighted images and enhanced very well. Initial biopsy of the mass suggested a peripheral nerve sheath tumor. The mass grew rapidly after biopsy, and enucleation was performed. The final diagnosis based on histology and immunohistochemistry was a low-grade mesectodermal leiomyosarcoma. To our knowledge, this is the first reported case of a malignant form of mesectodermal leiomyoma of the ciliary body.

Leiomyoma of the ciliary body is extremely rare and is believed to originate from mesodermal tissue derived embryologically from the neural crest, so-called mesectoderm (1). This specialized type of leiomyoma, mesectodermal leiomyoma, microscopically more closely resembles a neurogenic than a myogenic tumor (1–3). Since the first report of this unusual hybrid neurogenic-myogenic tumor by Jacobiec et al in 1977 (1), a considerable number of cases have been reported worldwide (2–11). According to our thorough search of the literature, however, there has been no reported case of a malignant form of this tumor, and all published cases have been considered histologically benign. Furthermore, only a few cases concerning the MR imaging appearance of this rare tumor have been reported (6, 8, 12). We report a case of low-grade mesectodermal leiomyosarcoma of the ciliary body occurring in a 12-year-old boy, with emphasis on the MR imaging and histologic findings.

Case Report

A 12-year-old boy presented with an 8-month history of a progressive loss of vision in his right eye. On examination, visual acuity in the right eye was estimated as hand motion. Visual acuity in the left eye was 20/20. Intraocular pressure was

normal in both eyes. Ophthalmologic examination showed dilated episcleral vessels and a 1.0-cm-round soft tissue mass in the temporal aspect of the right ciliary body with an associated retinal detachment. The lens was mildly pushed forward and medially by the tumor.

Ultrasonography and MR imaging revealed a $1.0 \times 1.0 \times 1.3$ -cm well-margined, ovoid soft tissue mass at the temporal aspect of the right ciliary body, accompanied by a large amount of subretinal fluid. For the most part, the mass, as well as subretinal fluid, was heterogeneously hyperechoic on ultrasonographic examination. On MR imaging, the signal intensity of the mass was slightly hyperintense to contralateral vitreous on T1-weighted images and markedly hypointense on T2-weighted images. By contrast, subretinal fluid was markedly hyperintense on T1-weighted images and slightly hypointense on T2-weighted MR images (Fig 1A). The mass enhanced substantially after the intravenous injection of contrast material, whereas the subretinal fluid did not (Fig 1B).

Under the assumption that malignant melanoma or medulloepithelioma was present, an incisional biopsy was performed, disclosing a dark brown mass originating from the temporal ciliary body of the right eye. Several pieces of yellow-tan soft tissue from the mass were removed. On the basis of microscopic findings showing spindle cell proliferation with fibrillary cytoplasm and mild nuclear pleomorphism, the pathologist suggested neurofibroma as the most probable diagnosis. Other differential diagnoses were schwannoma and benign smooth muscle tumor. Immunohistochemical staining could not be performed, because of insufficient biopsy specimens.

Ten days after the first operation, the patient was readmitted to the hospital with complaints of swelling and palpable mass in the operated eye. Ophthalmologic examination revealed a 2×2 -cm protruding soft tissue mass in the anterolateral aspect of the right eye. The cornea was edematous and blood-stained, and the anterior chamber was grossly filled with blood as well. The intraocular pressure of the right eye measured 29 mm Hg.

Follow-up MR imaging showed the right eye to be macrophthalmic and peculiar in shape. There was a relatively well-margined, crescentic soft tissue mass along the protruding anterolateral ocular coat (Fig 1C). The signal intensity and enhancing characteristics of the mass were same as those of the original ciliary body mass shown on the MR images obtained before biopsy. The boundary of the uveoscleral coat in the vicinity of the mass was partly indistinct, which indicated tumoral infiltration. Also noted was a total retinal detachment, accompanied by subretinal fluid that was markedly hyperintense relative to vitreous on T1-weighted images and heterogeneously isointense and hyperintense on T2-weighted MR images, representing various stages of subacute hemorrhage (Fig 1C). The lens was subluxated laterally.

Enucleation of the right eye followed by hydroxyapatite artificial eye insertion were performed. The specimen was cut along the anteroposterior axis, and the cut section showed marked outward protrusion of the anterolateral aspect of the ocular coat. The excised tumor was a $1.3 \times 1.2 \times 2.3$ -cm, relatively well-defined, crescentic, firm, grayish brown mass. It was firmly attached to the overlying sclera. There was a total

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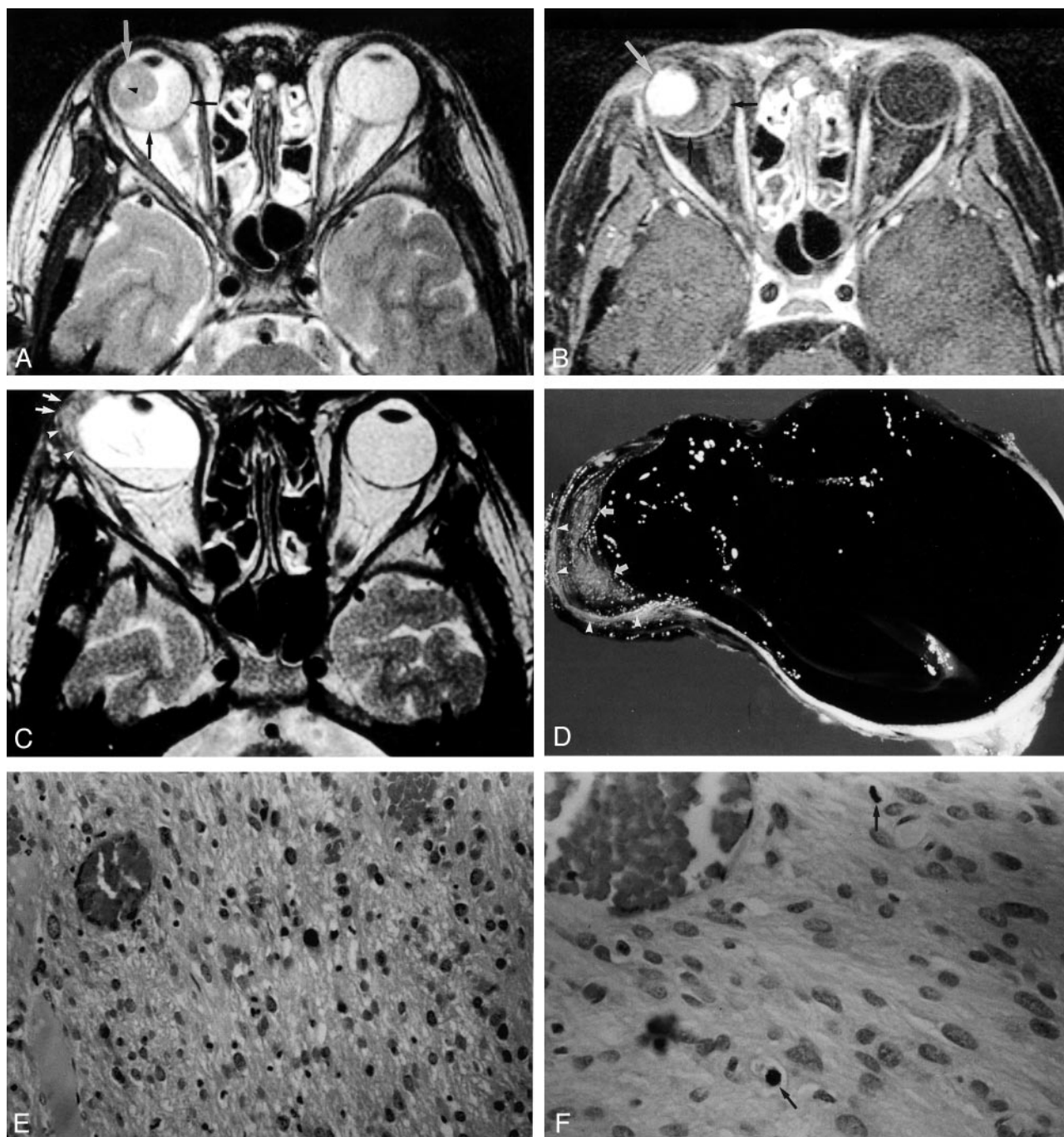


FIG 1. Low-grade mesectodermal leiomyosarcoma of the ciliary body in a 12-year-old boy.

A and B, MR images obtained before biopsy demonstrate a well-margined, ovoid soft tissue mass (white arrows) at the temporal aspect of the right ciliary body, accompanied by a large amount of subretinal fluid (black arrows) caused by retinal detachment. Compared with contralateral vitreous, the signal intensity of the mass was slightly hyperintense on T1-weighted (not shown) and markedly hypointense on T2-weighted (A) images, whereas that of subretinal fluid was markedly hyperintense on T1-weighted and slightly hypointense on T2-weighted images. The mass enhanced very well, whereas the subretinal fluid did not (B). Note focal hyperintensity within the mass on the T2-weighted image (arrowhead in A), which suggests cystic or necrotic change.

C, Axial T2-weighted MR image obtained 10 days after biopsy shows the deformed, macrophthalmic right eye whose anterolateral coat is pushed outward in association with a total retinal detachment. There is a relatively well-margined, crescentic, markedly hypointense soft tissue mass (arrows) along this protruding portion of the ocular coat. The mass was slightly hyperintense to contralateral vitreous on T1-weighted images and enhanced well (not shown), as same as the original ciliary body mass shown on the MR images obtained before biopsy. The boundary of the uveoscleral coat in the vicinity of the mass is partly indistinct (arrowheads), which indicates tumoral infiltration. Subretinal fluid containing fluid-fluid level, which shows mixed isointensity and hyperintensity to contralateral vitreous on this T2-weighted image, demonstrated slight hyperintensity on T1-weighted images (not shown), representing various stages of subacute hemorrhage. Also noted was the lens subluxated laterally.

D, Macroscopic view of horizontal section of the enucleated right eye shows marked outward protrusion of the anterolateral ocular coat where a relatively well defined, crescentic, firm, grayish brown mass is located (arrows). It is firmly attached to the overlying sclera (arrowheads). There is a total retinal detachment and associated subretinal fluid containing brownish gray gelatinous material and blood clot.

retinal detachment, and the subretinal space was filled with brownish gray gelatinous material and blood clot (Fig 1D).

Microscopically, the tumor consisted of sheets of large polygonal cells that tended to become spindle shaped toward the base of the tumor. The tumor cells had large round to oval hyperchromatic nuclei, abundant eosinophilic cytoplasm with indistinct borders, and slender cellular processes blending into a fibrillary background (Fig 1E). Moderate cellular atypia and 4–6 mitotic figures per 10 high-power field strength were seen (Fig 1F). The sclera was partly invaded by the tumor.

Immunohistochemical studies obtained by using a standard immunoperoxidase technique disclosed strong immunoreactivity for muscle-specific actin, equivocal immunoreactivity for synaptophysin, S-100 protein, and neuron-specific enolase, and negative staining for HMB45. The final diagnosis based on light microscopy and immunohistochemistry findings was a low-grade mesectodermal leiomyosarcoma of the ciliary body. The postoperative course was uneventful, and the patient has shown no evidence of tumor recurrence during 3 years of follow-up.

Discussion

Smooth muscle of the iris and ciliary body is embryologically different from that of the other parts of the body. Whereas the latter is derived from the mesoderm, the former is believed to be of neural crest origin, the so-called mesectoderm. Although there is no uniform consensus, it seems likely that those cells derived from the neural crest give rise to most, if not all, uveal leiomyomas (6, 10). Cases describing observation of melanocytes scattered throughout the tumor (3) and tumor associated with a nevus (2) also favor the neural crest origin of this tumor. By convention, however, the term *mesectodermal* is generally reserved for tumors that have neurogenic features seen by light microscopy (6).

Mesectodermal leiomyoma of the ciliary body has a predilection for young women, and most patients are in the second, third, or fourth decade of life (6, 8–10). In contrast to melanoma, which is located in the uveal stroma, these tumors are usually located in the supraciliary or suprachoroidal space. During transillumination, it usually transmits light well, whereas most melanomas cast a shadow (9). Although these clinical features may be useful for differentiating this unusual tumor from the more common melanoma, the definitive diagnosis can be acquired only by the thorough histologic examination, often with the aid of electron microscopy (6, 11).

Because of its unique embryopathogenesis, mesectodermal leiomyoma of the ciliary body has a characteristic appearance on light microscopy suggestive of neuroid tumors, such as ganglionic, astrocytic, and even peripheral nerve tumors (1, 2). Whereas a classic leiomyoma comprises intertwining bundles of spindle-shaped cells with elongated nuclei, a mesectodermal leiomyoma consists of large polyhedral cells with ovoid nuclei and prominent nucleoids, abundant eo-

sinophilic cytoplasm, and a fine fibrillary background. Pleomorphic nuclei, the orientation of some of the cell processes toward the capillaries, and the rich vascular network also suggest a glial tumor (8). Because of the histologic features of the hybrid neurogenic-myogenic tumor mentioned above, a mesectodermal leiomyoma may closely resemble an amelanotic melanoma and a benign peripheral nerve sheath tumor on light microscopy (6), as in the present case. Other microscopic differential diagnoses include epitheloid leiomyoma, amelanotic nevus, paraganglioma, or hemangiopericytoma (7). Although not performed in this case, electron microscopy, by disclosing smooth muscle differentiation of the tumor, can provide the ultimate diagnostic clues, in cases where light microscopic examination is inconclusive. The electron microscopic findings include the occasional clusters of mitochondria near the perikaryon, abundant fine filaments with fusiform densities, basement membrane, and small caveolae of plasmalemma (1, 8). Recently, immunohistochemical study, which is less expensive, faster, and easier, has tended to supplant electron microscopic examination. In this study, mesectodermal leiomyomas usually show positive immunoreactivity for muscle markers and negative staining for melanoma-specific antigen and neural markers (7, 9, 10). Likewise, the tumor in the present case demonstrated strong immunoreactivity for muscle-specific actin and no immunoreactivity for HMB45. Although equivocal evidence of focal immunostaining was noted for various neural markers in the present case, their immunoreactivity was considered too nonspecific to rely on.

Mesectodermal leiomyoma of the ciliary body may be locally invasive, and cases showing forward extension to the root of the iris or into the anterior chamber were reported (5, 8); however, no mitotic figures were found in those cases. Although there have also been cases showing occasional mitotic figures, other clinical and histologic evidences for malignancy were lacking (3, 4). The present case is unique in that the tumor displayed a rather aggressive biologic and morphologic behavior. After the first operation, the tumor enlarged rapidly in a short period. Histologically, some portions of the sclera were infiltrated by the tumor, and there were moderate atypia and mitotic figures among the tumor cells, which seemed to be unusual for the ordinary mesectodermal leiomyoma. These biologic and histologic features favored the diagnosis of a low-grade mesectodermal leiomyosarcoma over a simple mesectodermal leiomyoma. To our knowledge, this probably is the first reported case of a malignant form of mesectodermal leiomyoma of the ciliary body. A careful search of the literature found only one case of mesectodermal leiomyosarcoma of the head and neck region, in which the orbit

E, Photomicrograph shows that the tumor is composed of sheets of large polygonal cells that have large round to oval hyperchromatic nuclei, abundant eosinophilic cytoplasm with indistinct borders, and slender cellular processes blending into a fibrillary background (hematoxylin-eosin stain, original magnification $\times 100$). F, High-magnification photomicrograph shows moderate cellular atypia and several mitotic figures (arrows) among the tumor cells (hematoxylin-eosin stain, original magnification $\times 400$).

was secondarily involved by the tumor originating from the maxillary sinus (13).

There have been only three reports describing the MR imaging features of leiomyoma of the ciliary body (6, 8, 12). Compared with that of the vitreous body, the signal intensity was hyperintense on both T1- and T2-weighted images in one case and hyperintense on T1-weighted images and hypointense on T2-weighted images in the other two cases. The signal intensity of the mass in the present case was hyperintense on T1-weighted images and hypointense on T2-weighted images, as in the cases reported by Shields et al (6) and De Potter et al (12). These MR imaging features are rather nonspecific, and similar patterns of signal intensity can also be observed in other more common intraocular tumors, such as retinoblastoma, medulloepithelioma, melanoma, and retinal or choroidal hemangioma (12). Mesectodermal leiomyoma of the ciliary body is often accompanied by various degrees of retinal detachment containing fluid in the subretinal space. The signal intensity of this subretinal fluid is variable and depends on the nature of fluid—that is, transudative, exudative, or hemorrhagic. The differentiation of the tumor from subretinal fluid is best achieved by contrast enhancement, as in the present case, where the former enhanced significantly but the latter did not. The enhancement pattern of leiomyoma of the ciliary body on MR imaging has been reported only once, by De Potter et al (12), who described marked enhancement of the tumor, more prominent than melanoma, as in the present case.

At present, it is difficult to conclude what would be the most appropriate management option for mesectodermal leiomyosarcoma of the ciliary body. As for intraocular leiomyomas, Shields et al (9) claim that they should be removed surgically by partial lamellar sclerouvectomy. They also suggest that surgery should be performed at a relatively early stage, before the occurrence of significant complications, such as progressive growth, subluxation of the lens, retinal detachment, and vision loss (9).

Conclusion

The malignant form of mesectodermal leiomyoma of the ciliary body is an extremely rare occurrence; however, in the appropriate clinical setting, if the tumor shows an aggressive biologic behavior, as in our case, it should be considered in the differential diagnosis of ciliary body tumors. MR imaging is useful for demonstrating not only the location, but also the morphologic characteristics of the tumor.

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