

Leiomyoma in the Posterior Choroid : A Case Report

Smooth muscle tumor of the uveal tract is rare, and mostly located in the cilio-choroidal area. We report a unique case of posterior choroidal leiomyoma in a 27-yr-old man. Ophthalmoscopic examination disclosed an 11 mm-sized mass on the fundus two-disc diameters apart from the optic disc. With a suspicion of amelanotic melanoma, the globe was enucleated. The mass occupied the whole thickness of choroidal stroma beneath the pigmented retinal epithelium and composed of spindle cells arranged in intersecting fascicles. Immunohistochemical studies demonstrated immunoreactivities of the tumor cells for smooth muscle actin, desmin, and vimentin. Ultrastructurally, numerous intracytoplasmic filaments with fusiform focal densities, scattered segmental external laminae, subplasmalemmal densities, and pinocytotic vesicles were noted. The leiomyoma in this case had several unusual features in that it was confined to the posterior choroid with no relation to the ciliary body, occupied the whole stroma of the choroid instead of suprachoroidal location, and occurred in a young male. It is important to include choroidal leiomyoma in the differential diagnosis of choroidal tumors.

Key Words : Eye; Uvea; Choroid; Neoplasms; Leiomyoma

Yoon Kyung Jeon, Hee Jung Cha,
Na Rae Kim, Chong Jai Kim, Je G. Chi

Department of Pathology, Seoul National University
College of Medicine, Seoul, Korea

Received : 2 January 2001
Accepted : 27 June 2001

Address for correspondence

Yoon Kyung Jeon, M.D.
Department of Pathology, Seoul National University
College of Medicine, 28 Yongon-dong, Chongno-gu,
Seoul 110-799, Korea
Tel : +82-2-760-2788, Fax : +82-2-743-5530
E-mail : junar@netian.com

INTRODUCTION

The intraocular smooth muscle tumor is rare and has been known to arise in the iris, the ciliary body, or the choroid. Choroidal leiomyomas usually involve the ciliochoroidal area (peripheral choroid) (1), and leiomyoma of the posterior choroid is extremely rare. Here we report a case of posterior choroidal leiomyoma in a young male.

The choroidal leiomyoma can mimic amelanotic melanoma, neurofibroma, neurilemmoma, and other tumors in several clinicopathologic aspects. Thus, electron microscopic examination and immunohistochemistry are essential for the definite diagnosis of leiomyoma. Shields and his colleagues described the major clinical and pathological features of leiomyoma of the ciliary body and the choroid, based on the personal observation of seven cases and literature review of additional seventeen cases (1). Some unique features to our case compared with these previous observations are described.

We represent this case to contribute to find out the characteristics of the intraocular leiomyoma and emphasize the possibility of leiomyoma in the differential diagnosis of choroidal tumors.

CASE REPORT

A 27-yr-old male was referred to the Department of Oph-

thalmology, Seoul National University Hospital for the further evaluation of a known intraocular tumor of the left eye. He had been with a good visual acuity until he experienced a sudden decrease of visual acuity and the defect of upper visual field of the left eye a month before. On examination, the visual acuity of the right eye was 20/20 and the left eye was only finger-countable at a distance of 30 cm. The intraocular pressure and the movement of both eyeballs were normal. Indirect ophthalmoscopic examination of the left eye disclosed a yellowish white, highly vascularized elevated mass at the temporal side of the fundus about two-disc diameters apart from the optic disc (Fig. 1). The serous retinal detachment was accompanied around the mass. MRI revealed an 1 cm-sized mass with lobulating contour in the retina, and the lesion had medium to low internal reflectivity with smooth attenuation and the acoustic quiet zone inside on ultrasonography (Fig. 2). The right eye showed no abnormalities. Under the suspicion of amelanotic melanoma of the choroid, the left globe was enucleated.

Gross examination of the enucleated eyeball revealed an oval to round nonpigmented mass measuring 11 × 11 mm in maximal dimension. The mass involved the lower temporal quadrant with retinal detachment around. The cut surface was gray to whitish and the mass had rubbery consistency. Hematoxylin and eosin stained sections revealed a well-circumscribed dome-shaped mass at the equator of the globe. The base of the mass was about 7 mm apart from the ciliary

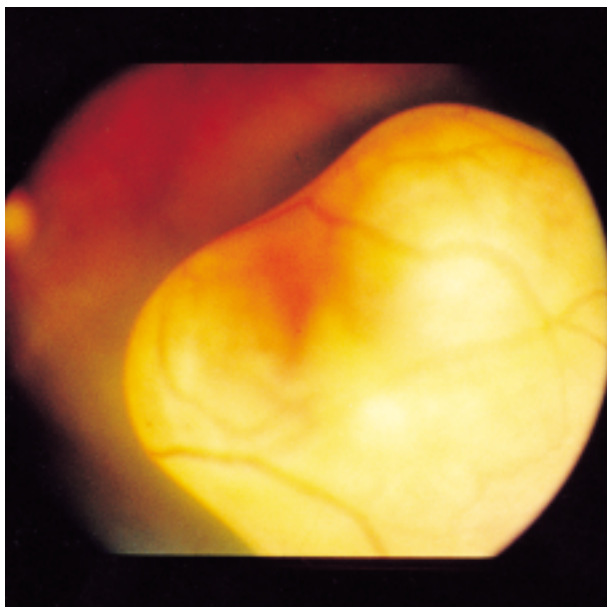


Fig. 1. Fundoscopy reveals a yellowish white vascularized mass.

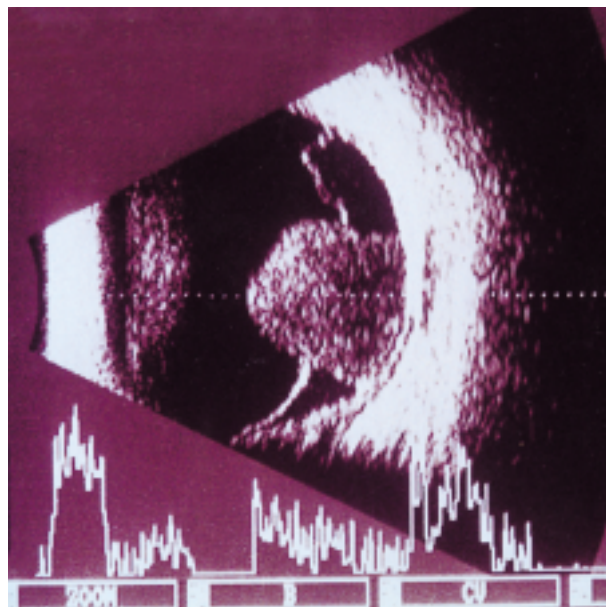


Fig. 2. On A scan ultrasonography, the lesion shows medium to low internal reflectivity with smooth attenuation. On B scan ultrasonography, the dome-shaped lesion shows the inside acoustic quiet zone without orbital shadowing.

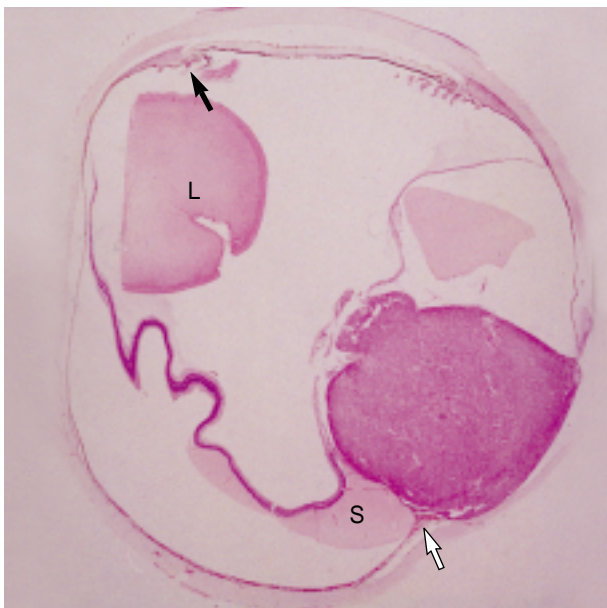


Fig. 3. Low power photograph of the left eyeball demonstrates the mass is apart from ciliary body (closed arrow) and occupies the whole stroma of the choroid (open arrow). L; lens, S; subretinal effusion (Hematoxylin & Eosin, $\times 3$).

body and from the optic disc. Pinkish subretinal serous fluid collection was observed around the proximal ora serrata region and the optic nerve. At the margin of the base, the retinal pigmented epithelial layer was elevated by the mass. The mass occupied the whole thickness of choroidal stroma, and was close to the sclera without erosion (Fig. 3). The retina overlying the mass was degenerated and atrophic. The tumor

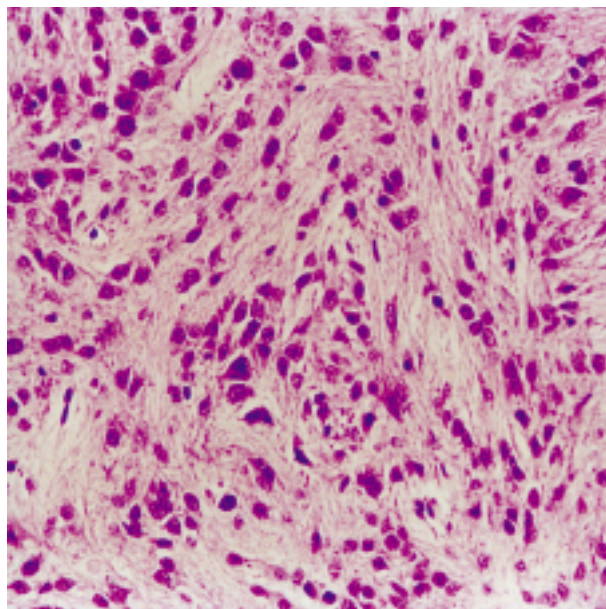


Fig. 4. The tumor was composed of spindle cells with oval nuclei and fibrillary cytoplasm (Hematoxylin & Eosin, $\times 200$).

was composed of spindle cells arranged in intersecting fascicles. There were vaguely palisading pattern of nuclei and mild collagen deposit amongst the cells (Fig. 4). The spindle cells had cigar-shaped, blunt-ended or oval to round nuclei, and abundant eosinophilic fibrillary cytoplasm. Cystic or slit-like vascular spaces lined by flat endothelium were dispersed

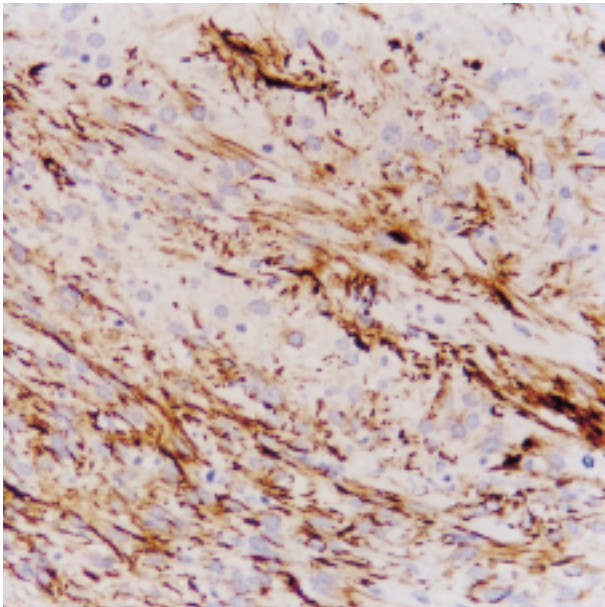


Fig. 5. Immunohistochemically, the tumor cells are reactive for desmin, a marker of smooth muscle cells ($\times 200$).

throughout the tumor. The vascular spaces tended to be more cystic at the periphery. The nuclei had mild pleomorphism and hyperchromasia with occasional nucleoli. There were mitoses up to three per ten high power ($\times 400$) fields, but atypical mitosis was not found. Immunohistochemical studies demonstrated immunoreactivity of tumor cells for smooth muscle actin, desmin, and vimentin (Fig. 5). The tumor cells were negative for S-100 protein and melanoma-specific antigen (HMB-45). Glial fibrillary acidic protein staining showed positivity only for glial fibers of retina. Epithelial membrane antigen, CD34, synaptophysin, chromogranin, and CD56 stainings were also negative. On ultrastructural examination, the tumor cells had irregular contour and their nuclei were oval with clumped chromatin and one or two small nucleoli. Cytoplasmic organelles were relatively well developed including mitochondria, rough and smooth-surfaced endoplasmic reticulum. Numerous intracytoplasmic filaments with fusiform focal densities were easily found. In addition, there were scattered segmental external laminae, subplasmalemmal densities along the cell membrane and micropinocytic vesicles. In intercellular space, small amount of thick and short collagen bundles were noted (Fig. 6).

DISCUSSION

Majority of the documented cases of intraocular leiomyomas have occurred in the ciliary body or peripheral choroid (ciliochoroidal) (1-4). Choroidal leiomyomas constitute only three ones among the total 26 reported cases of the leiomyomas in the ciliary body or the choroid (2, 3, 5). There was

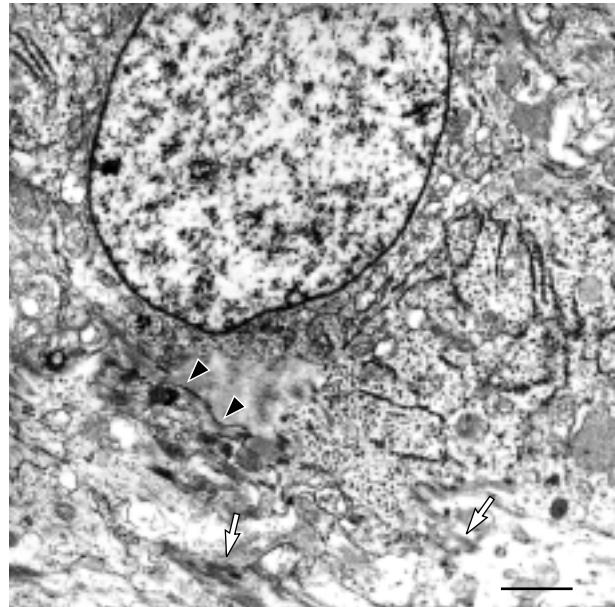


Fig. 6. Ultrastructurally, tumor cells show features of the smooth muscle cells that are characterized by intracytoplasmic filaments with focal densities (open arrows), subplasmalemmal densities (arrowheads), and suspicious pinocytotic vesicles ($\times 6,800$, bar; $1.5 \mu\text{m}$).

only one case confined to the posterior choroid without relation to the ciliary body (2). Among that 26 cases, 15 cases were confirmed by immunohistochemical or ultrastructural study. Clinically, it is very difficult to differentiate leiomyoma from melanoma, and actually all the reported cases were preoperatively considered as melanomas (6).

Histopathologically, the intraocular leiomyoma is composed of interlacing bundles of spindle cells with blunt-ended oval nuclei, moderate amounts of fibrillary cytoplasm, and intercellular myogial fibrils which are not distinguishable from neurofibrils of Schwann cell tumor on trichrome staining (7). Light microscopic examination alone often cannot discriminate leiomyoma from other spindle cell tumor, such as amelanotic melanoma, nevus, neurofibroma, neurilemmoma, hemangiopericytoma, and meningioma. So electron microscopic examination and immunohistochemistry are essential for the definite diagnosis of intraocular leiomyoma (9). The ultrastructural characteristics of smooth muscle tumor include cytoplasmic filaments with fusiform densities, subplasmalemmal densities, scattered segmental external laminae and pinocytotic vesicles in the cell membrane, all of which were the features of the present case. The intracytoplasmic filaments of astroglial tumors lack fusiform densities (8, 11). Leiomyomas show positive immunoreactivity for smooth muscle actin and antibodies against intermediate filaments such as desmin and vimentin. The immunohistochemical findings of our case were compatible with the those of smooth muscle tumor. In addition, we performed additional stainings for CD34 and epithelial cell membrane antigen to rule out

hemangiopericytoma and meningioma.

Shields and coworkers reported seven cases of intraocular leiomyoma. Of these cases, one involved the ciliary with extension to the iris, one involved the episcleral area, and five involved the ciliary body and choroid. Ciliochoroidal leiomyomas had a tendency to involve women in their reproductive ages and a supraciliary or suprachoroidal space separate and distinct from the uveal stroma (1). They mentioned that other rare uveal tumors such as neurilemmoma and hemangiopericytoma also have a tendency to involve the supraciliary space and sclera, but melanoma almost never occupies such a supravveal location.

Our case shows salient features in several aspects. At first, it occurred in a young male in contrast to the observations of Shields et al. Of the three cases of choroidal leiomyoma reported, only one case confined to the posterior choroid is available in the world literature as previously mentioned (2, 3, 5). However the tumor was diffuse and infiltrative (2). To the best of our knowledge, the leiomyoma in our case is the first one forming typical dome-shaped mass in the posterior choroid with no relation to the ciliary body. It occupied the whole thickness of choroidal stroma rather than suprachoroidal area, which is also an unusual feature. Like ours, Ceballos and coworkers also recently documented an unusual ciliochoroidal leiomyoma in the choroid rather than the suprachoroidal space (3).

Intraocular leiomyomas are believed to originate from the smooth muscle in the iris, the ciliary body muscle, blood vessel-associated smooth muscle or pericyte, or heterotopic smooth muscle cells in the choroid (2, 10). The term "mesectodermal leiomyoma" has been used for uveal leiomyoma emphasizing the pathogenesis when the tumor are composed of cells with both myogenic and neurogenic features by light and electron microscopy (11, 12). The cells of the neural crest that contribute to the formation of bone, cartilage, connective tissue, and smooth muscle in the region of the head and neck are called "mesectoderm". Ocular and periocular supportive tissue including the ciliary body muscle arises from the neural crest and only the external ocular muscles and vascular endothelia arise from the true mesoderm. Thus uveal leiomyomas often have been divided into two groups: those that are mesodermal and derived from vascular smooth muscle, and those that are mesectodermal and presumed to originate from the ciliary body smooth muscle which is a neural crest derivative (2, 9, 11). The possibility of mesectodermal origin may explain the frequent confusion of the disease with neuronal, glial, and nerve sheath tumors. However, some authors maintained that the separation of intraocular leiomyoma is unduly speculative and unimportant in terms of the treatment (4, 9), and the term of mesectodermal leiomyoma should be restricted to those that show distinct myogenic and neurogenic differentiation. We could not find any evidence of neurogenic differentiation in this case on immunohistochemical and ultrastructural examinations. The present case

is supposed to originate from the vascular smooth muscle of the ciliary vasculature entering posterior choroid.

Although intraocular leiomyoma is cytologically benign, it may show progressive growth to a large size and cause subluxation of the lens, cataract, retinal detachment, and visual loss. So surgical resection is recommended for this disease. The tendency for peripheral location of the tumor facilitates a local resection. Local excision is preferable to enucleation even when a tumor of the ciliary body is suspected to be malignant, due to the diagnostic difficulties and the higher percentage of benign lesions than malignant lesions in the ciliary body (13-15). The estimation of the malignancy of smooth muscle tumor has been a point of dispute in other organs. The rarity of leiomyoma in the ciliary body and the choroid further makes it difficult to determinate the biological behavior. Although there is no single case of "leiomyosarcoma" in the ciliary body and the choroid, the pleomorphism and mitoses in our case made us to recommend a close follow-up of the patient.

In summary, we described a case of choroidal leiomyoma with unusual features and suggest that choroidal leiomyoma should be included in the differential diagnosis of choroidal tumors.

REFERENCES

1. Shields JA, Shields CL, Eagle RC Jr, De Potter P. *Observations on seven cases of intraocular leiomyoma: the 1993 Byron Demorest Lecture. Arch Ophthalmol* 1994; 112: 521-8.
2. Jakobiec FA, Witschel H, Zimmerman LE. *Choroidal leiomyoma of vascular origin. Am J Ophthalmol* 1976; 82: 205-12.
3. Ceballos EM, Aaberg TM Jr, Halpern RL, Grossniklaus HE. *Choroidal leiomyoma: report of a case. Retina* 1999; 19: 349-51.
4. Biswas J, Kumar SK, Gopal L, Bhende MP. *Leiomyoma of the ciliary body extending to the anterior chamber: clinicopathologic and ultrasound biomicroscopic correlation. Surv Ophthalmol* 2000; 44: 336-42.
5. Naumann GO. *Leiomyoma of the choroid. Presented at the seventh biennial meeting of the Association of Ophthalmic Alumni, Armed Forces Institute of Pathology; Washington, DC; June 3-4, 1997.*
6. Chang M, Zimmerman LE, Mclean I. *The persisting pseudomelanoma problem. Arch Ophthalmol* 1984; 102: 726-7.
7. Green WR. *The uveal tract. In: Spencer WH, Font RL, Green WR, Howes EL, Jakobiec FA, Zimmerman LE, editors. Ophthalmic Pathology. An Atlas and Textbook. Philadelphia: WB Saunders, 1990; 3: 1685-9.*
8. Meyer SL, Fine BS, Font RL. *Leiomyoma of the ciliary body. Electron microscopic verification. Am J Ophthalmol* 1968; 39: 2102-13.
9. Foss AJ, Pecorella I, Alexander RA, Hungerford JL, Garner A. *Are most intraocular "leiomyomas" really melanocytic lesions? Ophthalmology* 1994; 101: 919-24.
10. Shields CL, Shields JA, Varenhorst MP. *Transscleral leiomyoma. Ophthalmology* 1991; 98: 84-7.

11. Jakobiec FA, Font RL, Tso MOM, Zimmerman LE. *Mesectodermal leiomyoma of the ciliary body: a tumor of presumed neural crest origin. Cancer* 1977; 39: 2102-13.
12. Yu DY, Cohen SB, Peyman G, Tso MOM. *Mesectodermal leiomyoma of the ciliary body: new evidence for neural crest origin. J Pediatr Ophthalmol Strabismus* 1990; 27: 317-21.
13. Takagi T, Ueno Y, Matsuya N. *Mesectodermal leiomyoma of the ciliary body: an ultrastructural study. Arch Ophthalmol* 1985; 103: 1711-4.
14. Ishigooka H, Yamabe H, Kobashi Y, Nagata M. *Clinical and pathological status of mesectodermal leiomyoma of the ciliary body: a case report and review of the literature. Graefes Arch Clin Exp Ophthalmol* 1989; 227: 101-5.
15. White V, Stevenson K, Garner A, Hungerford J. *Mesectodermal leiomyoma of the ciliary body: case report. Br J Ophthalmol* 1989; 73: 12-8.
16. Peyman GA, Martinez CE, Hew A, Peralta E, Kraut RJ. *Endoresection of a ciliary body leiomyoma. Can J Ophthalmol* 1998; 33: 32-4.