Mesectodermal Leiomyoma of the Ciliary Body

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Abstract: Mesectodermal leiomyoma of the ciliary body is a rare intraocular tumor. Clinically, the diagnosis of intraocular leiomyoma is challenging and the most important differential diagnosis is uveal melanoma. A 22-year-old female with high body mass index (34.5 kg/m^2) presented with deteriorated vision to hand motion in left eye. Ophthalmologic examination disclosed a pigment mass in the ciliary body of the left eye and magnetic resonance imaging revealed an intraocular lobulated tumor mass with hyperintense on T1-weighted images and hypointense on T2-weighted images. Enucleation was performed because the possibility of choroidal melanoma could not be absolutely excluded. Pathological examination revealed a mesectodermal leiomyoma. Although the association between mesectodermal leiomyoma and obesity requires further investigation, this case highlights the need for including mesectodermal leiomyoma in the differential diagnosis of patients with intraocular tumor especially for young female.

Keywords: Mesectodermal Leiomyoma, Ciliary Body, Obesity.

Mesectodermal leiomyoma is a rare benign tumor assumed to originate from the ciliary body smooth muscle, a neural crest derivative embryologically [1]. Clinically, the diagnosis of intraocular leiomyoma is challenging and the most important differential diagnosis is uveal melanoma. We report a case of this rare tumor in a young female with high body mass index (BMI).

CASE REPORT

A 22-year-old female presented with a 2-month history of gradual deterioration of visual acuity in her left eye. Her BMI was 34.5 kg/m^2. The best-corrected visual acuity was hand motion on 20cm in the left eye. Ophthalmologic examination disclosed a pigment mass in the ciliary body of the left eye (Figure **1**). On ultrasonography, the lesion showed medium to low internal reflectivity. Magnetic resonance imaging revealed an intraocular lobulated tumor mass with 19mm in maximum diameter with hyperintense on T1-weighted images and hypointense on T2-weighted images (Figure **2**). Enucleation was performed because these image findings could not absolutely exclude the possibility of choroidal melanoma. Gross examination disclosed a 24x24x23 mm mass with ciliary body and choroid involvement. Microscopic examination revealed that the tumor was composed of intersecting fascicles of spindle-shaped cells (Figure **3**). No mitotic figures were noted. Immunohistochemical analysis showed positivity for smooth muscle actin (SMA) and anti-muscle actin specific monoclonal antibody (HHF-35) and no reactivity with GFAP, S-100 and HMB-45. The diagnosis of mesectodermal leiomyoma was established, and no recurrence or metastasis was found after 18 months of follow-up.

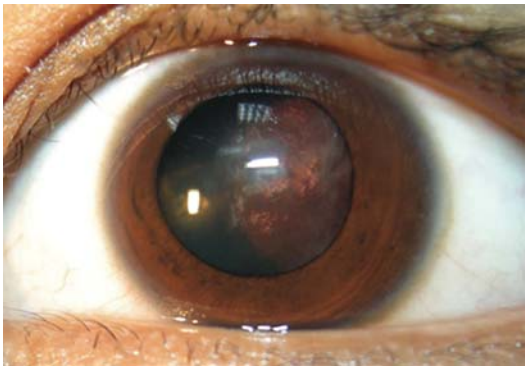

Figure 1: Slit-lamp photography showing a large dome-shaped ciliary body mass in the temporal quadrant of the left eye (after mydriasis).

DISCUSSION

This is the first report to demonstrate a case of mesectodermal leiomyoma of the ciliary body in an obese female. Although the pathogenesis of intraocular leiomyoma remains unclear, it is thought to arise either from ciliary body smooth muscle (mesectodermal leiomyoma) or from vascular smooth muscle (mesoderm leiomyoma). The clinical diagnosis of intraocular leiomyoma is often challenging, and the confirmed diagnosis often relies on histology with the aid of immunohistochemistry and/or electron microscopy [2].

In different to uveal melanoma with slight predilection for males, intraocular leiomyoma tends to

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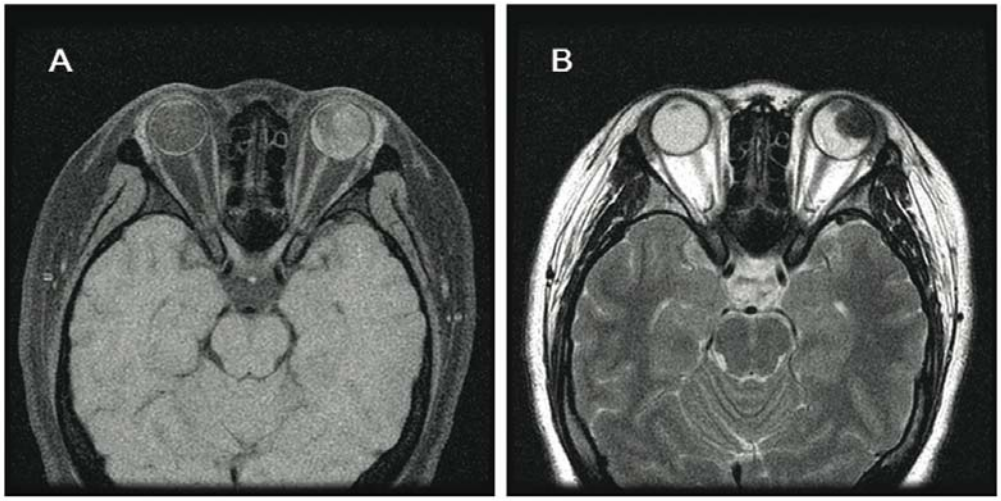

Figure 2: Magnetic resonance imaging. **A**: T-1 weight image showing the mass to be hyperintense to vitreous. **B**: T2-weighted image showing the mass to be hypointense to vitreous.

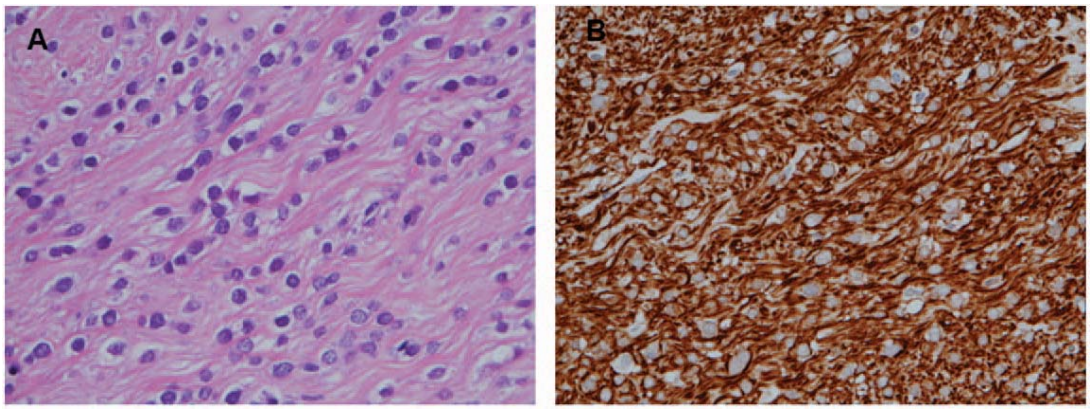

Figure 3: A: Light microscopy shows that mesectodermal leiomyoma is composed of ovoid or spindle-shaped cells with eosinophilic fibrillar cytoplasm, without evidence of pleomorphism or mitotic activity (H&E, x400). **B**: The neoplastic cells show intense positive immunoreactivity for smooth muscle actin confirming diagnosis.

occur in young women [2-5]. None was found to have systemic association except one case had both intraocular leiomyoma and uterine leiomyoma [6]. It has been proposed that intraocular leiomyoma have some relationship to uterine leiomyoma, which also tends to develop during the reproductive age [3]. Epidemiological studies have documented a positive association of uterine leiomyoma with body mass index and weight gain [7]. Obesity is noted to be associated with increased secretion of adipokines that significantly influence growth and proliferation of uterine stromal tumor [8]. It is noteworthy that our current case also had a high BMI (34.5 kg/m^2). Recent evidence further showed intraocular mesectodermal leiomyoma expressing sex steroid hormones**,** in a manner similar to conventional uterine leiomyomas [9]. However, the association between mesectodermal leiomyoma and obesity requires further investigation.

Clinically, it is very difficult to differentiate leiomyoma from melanoma, and most intraocular leiomyomas were initially diagnosed as melanoma. Image findings of MRI and ultrasonography for intraocular leiomyoma are usually similar to those of uveal melanoma.

In contrast to uveal melanoma, which most occurred in the white population, intraocular leiomyomas have often been reported in Asian patients [4-6, 10-14]. Although mesectodermal leiomyoma are pathologically benign and most have favorable treatment outcome, intraocular leiomyomas may show progressive growth to a large size and cause anterior chamber extension, cataract, retinal detachment, or visual loss [3, 15]. Large-sized tumors or those mimic malignant melanoma leave no choice other than enucleation. Partial lamellar sclero-uvectomy, endoresection, and transscleral resection have been performed for

peripheral localized intraocular leiomyomas with sparing of the eye [16]. Recurrent mesectodermal leiomyoma develops in one case 7 years after excision biopsy [14]. In addition, one case of mesectodermal leiomyosarcoma of ciliary body was reported to grow rapidly after initial biopsy, and enucleation was performed afterwards [17]. Long-term follow-up of these patients is mandatory especially for those underwent partially resected or biopsy only.

In conclusion, our case describes a mesectodermal leiomyoma of the ciliary body in a young obese female. Although rare, intraocular leiomyomas should be considered in differential diagnosis of intraocular tumors, especially in reproductive women patients.

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