

Mesectodermal Leiomyoma of Ciliary body in a young female

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Abstract

We report a rare case of a young lady of 22 years who presented with minor decrease of vision in her left eye since two weeks and on examination showed a large brown mass lesion just behind the lens. Based on the clinical features, trans illumination, ultrasonography and magnetic resonance imaging of the orbit, a potential diagnosis of ciliochoroidal melanoma was made. After proper counselling and explanation to the patient and parents keeping in consideration the size of the lesion, the eye was enucleated. Histopathological examination confirmed it to be a mesectodermal leiomyoma of the ciliary body. Such benign lesions can rarely attain large sizes with clinical features simulating tumours like melanoma, thus making management, challenging for the physician.

Keywords: Leiomyoma, Melanoma, Ciliary, Mesectodermal.

Introduction

Intraocular leiomyomas are rare benign smooth muscle tumors that arise usually from the ciliary body or rarely choroid with a tendency of presenting in young females.⁽¹⁾ Case reports on leiomyomas have been published since 1950s⁽²⁾ and till date around only 29 cases have been published out of which seven cases have been beautifully explained by Shields et al in their article.⁽¹⁾ In a retrospective analysis by Shields et al of 12000 cases of mass lesions of uvea, out of the 1739 pseudomelanoma cases only 10 were reported to be leiomyomas which accounts for < 1% of the all pseudomelanomas.⁽³⁾ Considering the rarity of the lesion, reporting these cases becomes even more important, to enable one to be familiar with their varied presentation. Usually reported as an amelanotic mass lesion behind the lens they are the first differential for amelanotic melanomas. Other differentials being medulloepithelioma, schwannoma, and nonpigmented adenoma of the ciliary epithelium with smooth muscle differentiation.⁽⁴⁾

We report a case of a young girl with large mass lesion behind the lens which in contrast to the other reported amelanotic lesions was pigmented and closely simulated a pigmented melanoma.

Case Report

A young girl 22 years of age was referred to our institute with a diagnosis of large melanoma for further management. Her chief complaint was decrease of vision in her left eye since two weeks associated with loss of nasal field of vision. On examination she had 6/6, N6 vision in her right eye and 6/9 N6 in her left eye. Intraocular pressure (IOP) was 14 mm of Hg in right eye and 13 mm in the left.

Anterior and posterior segment examination of the right eye was normal. Slit lamp examination of the left eye revealed dilated episcleral vessels in the supero-

temporal and infero-temporal quadrants and a large brown coloured mass just behind the lens covering more than half the pupil (Fig. 1).

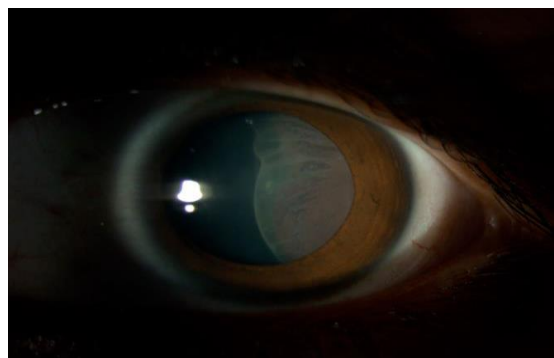
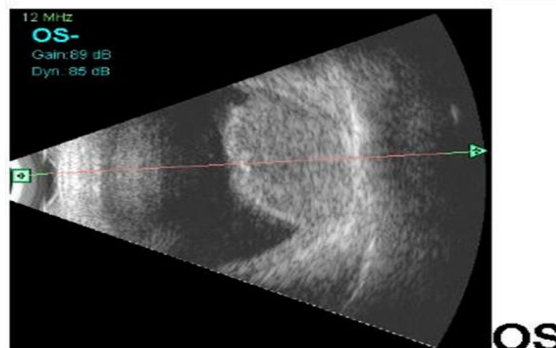


Fig. 1: Slit Lamp examination: Pigmented mass seen behind the lens

Fundus examination revealed inferior serous retinal detachment with clumps of tan coloured subretinal cells.

Ultrasonography (USG B-scan) of the left eye showed a mass lesion measuring 8.21×13.23×16.63 mm in the extreme temporal quadrant with high superficial and moderate internal reflectivity (Fig. 2).



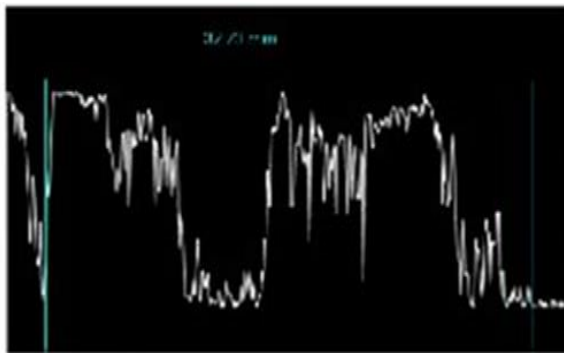


Fig. 2: USG B Scan: High Surface with moderate internal echogenicity

Trans-illumination test was negative.

Magnetic resonance imaging (MRI) scan revealed a 8.1×13.2 mm nodular lesion that was Hyper-intense on T1 and Hypo-intense on T2 weighted images (Fig. 3).

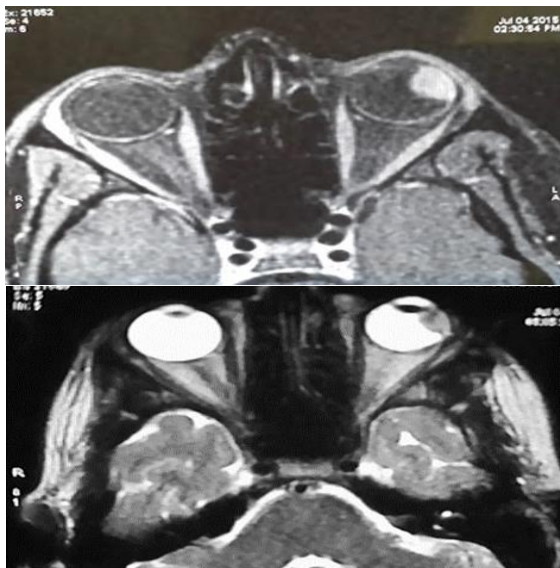


Fig. 3: MRI orbit: Lesion Hyperintense on T1 and Hypointense on T2 (with irregular intensity in T2)

Ultrasound examination of the abdomen, chest X-Ray and liver function tests were within normal range.

Considering the presence of a large pigmented mass that was not transilluminant and the findings of ancillary investigations, high possibility of the lesion being a ciliochoroidal melanoma was discussed in detail with the patient and her parents. After an informed consent, the eye was enucleated.

Histopathological examination (HPE) of the enucleated globe revealed a well circumscribed tumour arising from the ciliary body composed of sheets of spindle cells. There was no evidence of mitosis or necrosis. Tumour cells stained positive with SMA. Sections were negative for any malignancy (Fig. 4). It was reported as mesectodermal leiomyoma of the ciliary body.

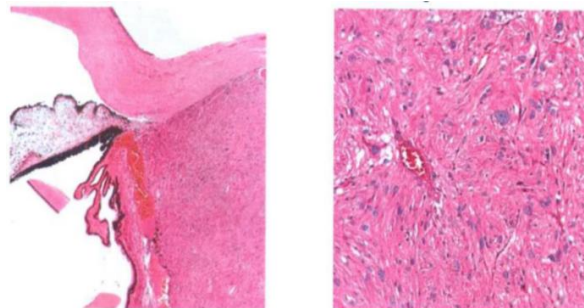


Fig. 4: Haematoxylin and eosin staining: Spindle cells seen

Discussion

Leiomyomas are rare benign intraocular smooth muscle tumours seen in young females involving the iris, ciliary body or choroid. 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.^(5,6,7) The pathogenesis is not very clear.

The biggest hurdle faced by ophthalmologists is to differentiate it from the malignant ciliary body amelanotic melanoma because of the similar clinical and imaging appearance. Both can appear as a dome-shaped, non-pigmented, smooth-surfaced, solid vascularized mass originating in the uvea. Both can extend through the wall of the eye and become extra scleral.⁽⁸⁾

Very few tests have been described in literature to differentiate these two pathologies - leiomyoma being suprauveal in location on ultrasonography in contrast to melanoma involving the uveal stroma and on trans illumination test leiomyomas transmitting light in contrast to melanomas blocking light. MRI features have been occasionally described and found to exhibit similar features in both. Diagnosis is usually confirmed on HPE, IHC (immunohistochemistry) and TEM (transmission electron microscopy).

In the above case the mass lesion clinically was pigmented, large, USG revealed high surface and moderate internal reflectivity, MRI T1 hyper and T2 hypo and the transillumination test blocking light all features strongly fitting well with the described features of melanoma. Trans illumination in this case might be negative because of the lesion being pigmented, confounding the diagnosis.

Uveal leiomyoma can grow slowly and extend into the anterior chamber^(9,10) leading to complications such as subluxation of the lens, glaucoma, and retinal detachment. These can extend through sclera as well into the orbit.⁽¹¹⁾ Despite the benign cytology, therapeutic intervention is usually necessary which is usually partial lamellar sclerouvectomy for lesions upto 3-4 clock hours. In larger lesions enucleation has been

described to prevent the additional complications and also because of the close possibility of the lesion being a large melanoma.

Out of the total 29 reported cases of leiomyoma in literature 10 underwent enucleation because of their large size and the others resection with one case having recurrence 7 years later in which resection was undertaken initially in spite of the large size but had to be later enucleated.⁽¹²⁾ In our case, even if we had reached a pre-operative diagnosis of leiomyoma, it would have been difficult to resect, considering the large size of the lesion.

In people of Indian and Asian origin, uveal melanomas occur at a younger age, making it further difficult to differentiate a pigmented leiomyoma from a melanoma.⁽¹³⁾ Learning from the above case a differential of leiomyoma can be kept based on the moderate internal reflectivity of the lesion on USG B Scan, irregular intensity changes on MRI and the young age of the patient. The MRI when seen carefully in this case was showing irregular intensity both in T1 and T2 specially in T2 weighted images.

This is a case of pigmented Leiomyoma simulating Melanoma proved histo-pathologically and is different from the previously described amelanotic ones. Being a rare pathology, all of us might not come across all the varied presentations of this, thus reporting such cases becomes crucial. Depending on the size of the lesion lamellar sclerouvectomy or enucleation is done. Confirmation is usually with HPE and IHC only but a differential should be kept in mind specially in lesions lesser than 3 clock hours in which cases eye can be saved functionally and structurally. In addition to the management of the tumour aspect, managing the psychological aspect especially for a young female is vital.

Conflict of Interest: None declared

Subjects informed consent has been obtained and the study protocol has been approved by the institute's committee on human research.

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