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RESEARCH ARTICLE

A rare giant leiomyoma of ciliary body & choroid-diagnostic dilemma & case report

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ABSTRACT

Leiomyoma is a rare benign tumor of smooth muscle origin, which in orbit can arise in the uvea (iris, ciliary body and heterotrophic smooth muscle of choroid). It often demonstrates a slow growth pattern & it is difficult to detect by routine ophthalmic examination at the early stage and the majority of cases of ciliary body tumor present a melanoma-like appearance clinically. Here, we are presenting a rare case of giant leiomyoma arising from ciliary body & choroid in a 21-year-old male presented with painful blind eye.

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INTRODUCTION

Leiomyoma of ciliary body is a benign smooth muscle tumour. Most patients are in the second or fourth decade of life at the time of diagnosis. Female to male ratio is 3:1. Most leiomyoma arise from ciliary body, but can also occur in iris & choroid. Clinically it presents with progressive blurring of vision with fleshy mass rarely exceeding 10 mm, readily transmitting light on transillumination.[1] Due to extreme rarity of occurrence, incidence estimates in general population are not available & very few case report/series are reported in English literature till date.

CASE HISTORY

A 21-year-old male came to out-door patient department with complaint of gradually progressive diminution of vision in right eye since 6 months associated with sudden pain in right eye since 2 days.

[Fig-1] His visual acuity was no perception of light in right eye and 6/6 in left eye .Slit lamp biomicroscopy revealed right eye proptosis, chemosis, circumciliary congestion, micro bullous corneal oedema, flat anterior chamber, semi-dilated fixed non-reacting pupil, anteriorly dislocated cataractous lens & increased intraocular pressure. These features are suggestive of secondary glaucoma due to mass effect of tumour. Fundus examination was not possible due to significant corneal oedema and cataractous lens. Magnetic resonance imaging (MRI) brain with orbit shows right sided intraocular mass lesion size 2.2x1.7x2.1cmat nasal side involving choroid, appearing hypointense on T2W images and iso to mild hyper intense on T1W images, suggestive of neoplastic etiology and a preliminary diagnosis of large ciliochoroidal melanoma was rendered and enucleation was planned due to painful blind eye [Fig-2]. After informed consent to the patient, right eye enucleation was done. On histopathological examination grossly a grey tan colour mass attached to ciliary body and choroid of size 2.5x1.8x2.2cm, without any extra-scleral extension [Fig-3]. Microscopic examination shows proliferation of spindle cells having round to ovoid bland nuclei & abundant eosinophilic cytoplasm in a fibrous stroma.

Fig. 1. Painful Right blind eye with proptosis, mechanical ptosis, chemosis, corneal edema

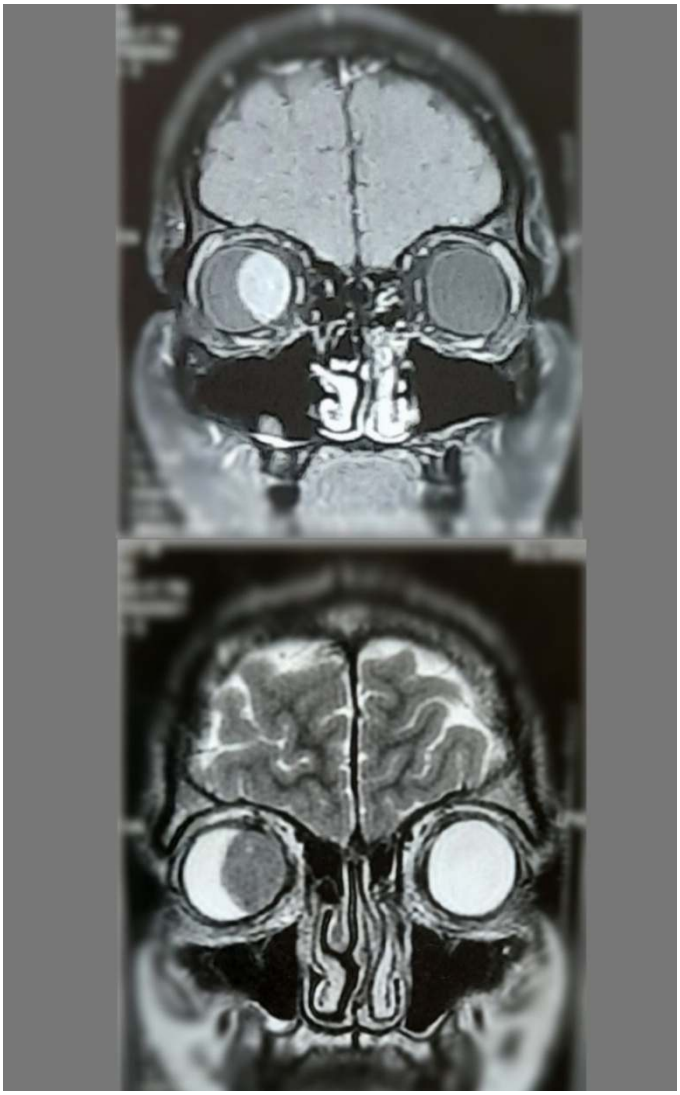

Fig. 2. MRI Brain with Orbit (P+C) - right sided intraocular mass lesion size 2.2x1.7x2.1 cm at nasal side involving choroid, appearing; Fig-2a. Iso to mild hyper intense on T1W image; Fig. 2b. hypointense on T2W image

No significant pleomorphism, atypical mitosis, pigmented cells or necrosis seen. [Fig-4] Suspected differential was schwannoma or leiomyoma, so IHC (Immunohistochemistry) was advised. On IHC, tumour was positive for markers-muscle specific actin, h-caldesmon and S-100 & negative for HMB-45 which confirmed the diagnosis of leiomyoma of ciliary body & choroid.

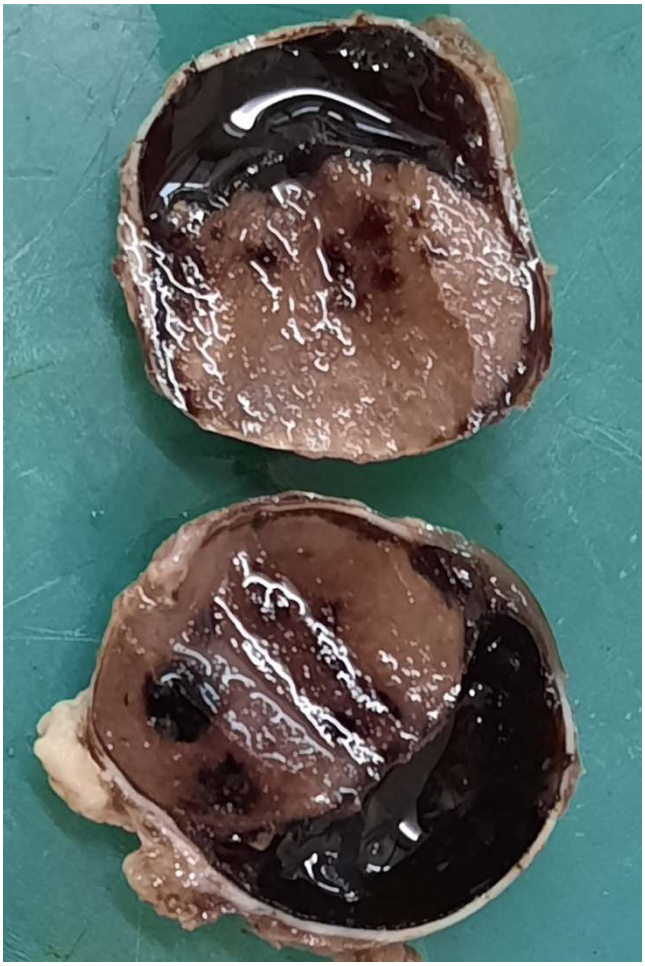

Fig. 3. Gross Examination- a grey tan colour mass attached to ciliary body and choroid of size 2.5x1.8x2.2cm, without any extra-scleral extension

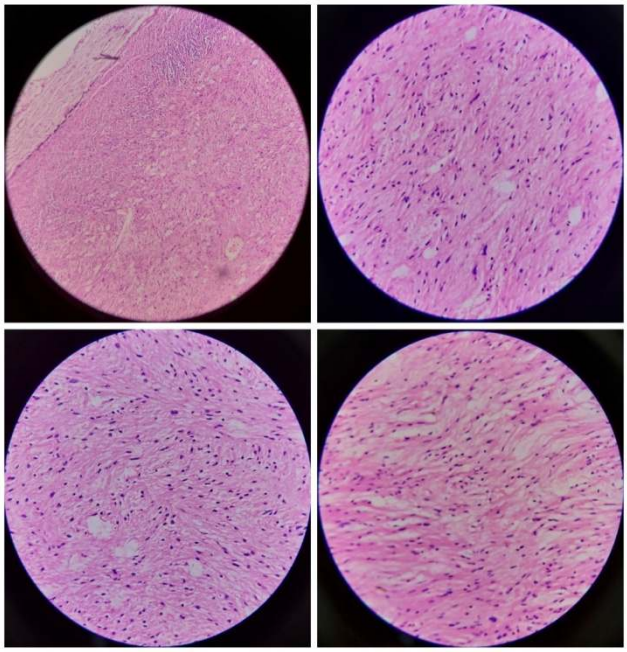

Fig-4. Microscopic Examination-Fig-4a-(Haematoxylin &Eosin stain,100x), Interlacingfascicles of spindle cells & dilated vessels. Fig-4b,4c,4d (Haematoxylin &Eosin stain,400x)- proliferation of spindle cells having round to ovoid bland nuclei & abundant eosinophilic cytoplasm in a fibrous stroma

DISCUSSION

Intraocular leiomyomas are rare benign tumors of uveal tract. Uveal Leiomyomas occur in relatively young age groups.[2] Tumor most frequently occurs in ciliary body followed by iris, posterior choroid, with ciliary body being predominant in females & iris, choroidal leiomyoma in males.[3] Mesoectodermal variant of leiomyoma shows hybrid histological features of neuroglial & smooth muscle differentiation.[4] In a review of 80 cases of uveal leiomyoma, Tomar AS *et al*, found the average largest dimension was 12.3±4.8(mean ± SD) (median: 13.0, range: 2.4-22) mm for ciliary body tumour and 12.0±1.0 (mean± SD) (median: 12.0, range: 11-13) mm for posterior choroid tumour.[3] While in our case, size of tumour was quiet large measured 25 x 22mm, considered as giant leiomyoma of ciliary body and choroid. Leiomyoma most commonly occurs in the anterior uvea and may exhibit slow progressive enlargement producing scleral thinning, ectasia, extra-scleral extension, cataract and secondary exudative retinal detachment with resultant vision loss.[3] There may be normal angle structures, focal angle closure and iridodialysis.[3] Fluorescein angiography reveals a vascularized tumour pattern and ultrasonography shows a well circumscribed mass with low to medium internal reflectivity.[1] CT scan depicts a mass with soft tissue density.[1]

While on MRI, with and without gadolinium enhancement, the tumor appears isointense to hyperintense in the T1-weighted images and exhibited marked signal enhancement with gadolinium. Leiomyomas exhibited a low signal in the T2-weighted images.[1] Leiomyoma, schwannoma & melanoma shows similar features on MRI.[5] Grossly leiomyomas are well circumscribed, greyish white, yellow smooth surfaced non pigmented mass. Leiomyoma is usually ovoid in shape whereas melanoma is frequently dome or mushroom shaped. Histopathological, leiomyomas are typically composed of interlacing bundles of spindle cell with blunt ended oval nuclei, moderate eosinophilic cytoplasm.[1] Leiomyoma shows immunoreactivity to smooth muscle actin (SMA), Muscle specific actin (MSA), h-caldesmon, calponin & desmin, while mesoectodermal variant shows positivity to CD56 & neuron specific enolase.[1] Goto H *et al* studied 32 cases of ciliary body tumour, found 4 cases of mesoectodermal leiomyoma & only 2 cases of ciliary body leiomyoma, both were female & treated by local resection & enucleation.[6] Differential diagnosis includemelanoma, schwannoma, neurofibroma, metastatic tumour and astrocytoma.[1,5] Uveal leiomyoma can closely simulate malignant amelanotic uveal melanoma from clinical and imaging perspectives. Both can appear as a dome-shaped, non-pigmented, smooth-surfaced, solid vascular mass originating in the uvea, which can extend through the wall of the eye and become extra scleral.[7] To differentiate by transillumination, in which leiomyoma usually transmits light and melanoma doesn't and ultrasonography shows leiomyoma as a dome-shaped mass in suprauveal location and melanoma as either dome or mushroom-shaped in uveal stroma.[2] The management of leiomyoma depends on tumour size and location. Local resection of entire tumour or observation is the treatment of choice. Local excision of iris leiomyomas in form of iridectomy or iridocylectomy is the treatment for well localized tumour.

In ciliary body and posterior choroidal leiomyomas trans scleral resection via partial lamellar sclerouveactomy or Endo resection via pars plana vitrectomy or enucleation. [8,9] Larger tumour is often managed by enucleation & prognosis is good. In summary, our case is a 21-year-old young male having painful blind eye suspected as choroidal melanoma, on enucleation diagnosed as giant ciliary choroidal leiomyoma & uneventful follow up till date.

CONCLUSION

Ciliochoroidal leiomyoma is a rare intra ocular benign tumour. There need to be clinicopathological correlation to diagnose ciliochoroidal leiomyoma as it is indistinguishable from ciliochoroidal melanoma. The only unequivocal way to confirm the disease is pathological evaluation with immunohistochemical studies. So Ophthalmologists will need to keep ciliochoroidal leiomyoma in their differential diagnosis of any uveal mass in young patients.

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