Abstract: An adolescent girl presented with defective vision in her left eye. On examination of Left eye Anterior segment was normal and Posterior segment examination showed circumscribed orange hue elevated lesion with pigmentary changes more than 5 DD size involving and extending around macula, with high clinical suspicion of choroidal melanoma. Subjecting the patient to various diagnostic investigation techniques it was diagnosed as choroidal hemangioma. The case is being presented to emphasize the need of investigation techniques in such a suspicious choroidal lesion so that it would be helpful in proper management and prevent the eye from erroneous enucleation.

Keyword: Choroidal Hemangioma, Choroidal Melanoma, B-scan, FFA, ICG, MRI

INTRODUCTION
Choroidal Hemangioma is a rare benign vascular hamartoma of uveal tract. It is often mistaken for uveal melanoma and a misdiagnosis frequently leads to enucleation. However, due to the availability of specific investigation techniques and better clinical recognition of the condition, proper treatment now allows for preservation of vision and the eye. The present article is about the differentiation of benign from malignant lesion by using investigation techniques which aid in prompt management.

CASE SUMMARY
A 13 year old girl presented to our hospital with complaints of defective vision in her left eye for 3 months (Figure 1). Apart from defective vision in the left eye which was insidious in onset, no other clinically significant history could be elicited. On examination, visual acuity in the right eye by Snellen’s chart was 6/6, anterior segment was normal and pupil was reacting to light. The left eye had a visual acuity of 6/36 improving with PH to 6/18. Anterior segment examination was normal and pupil was reacting to light. Both eyes had normal Intra Ocular Pressure of 12 mm of Hg & Colour vision was normal.

The macular area showed a circumscribed elevated lesion with an orange hue and mild pigmentary changes. It measured more than 5 DD size involving and extending around macula. Retinal vessels running over the lesion were normal (Figure 2).

Figure 1 Clinical profile photograph of the patient

Figure 2 (a) Fundus picture of Right eye, (b) Fundus picture of Left eye showing a circumscribed orange hue elevated lesion with mild pigmentary changes

With these clinical findings we arrived at a differential diagnosis of
- Circumscribed Hemangioma
- Amelanotic Choroidal Melanoma
- Solitary Choroidal metastases
- Choroidal Granuloma
- Posterior Scleritis
- Choroidal Osteoma
- Amelanotic Choroidal Melanoma
On subjecting the patient to various diagnostic investigations, we obtained a better picture of the actual pathologic process. B-scan of the Left eye showed an elevated lesion in the temporal side over the macula 2-3 DD from optic disc measuring about 2.2mm in height, 9.3mm in circumference and having a diameter of 7.8mm, with a high intense echo seen over the top of the lesion and low to moderate internal reflectivity. There was no evidence of choroidal excavation nor acoustic shadowing (Figure 3).

Figure 3 (a) B-scan of LE shows elevated lesion seen in the temporal side over the macula 2-3 DD from optic disc (b) A-scan vector shows high intense echo seen over the top of the lesion and low to moderate internal reflectivity

FFA of Left eye was done which showed normal filling pattern of disc and vessels. Macular lesion showed hyperfluorescence in early choroidal phase with increasing in intensity in late phase and multiple blocked fluorescence corresponding to RPE stippling over the lesion (Figure 4). OCT showed a lesion in the choroid elevating the retina (Figure 5). CT scan showed focal thickening of retina on temporal side of left eye with minimal contrast enhancement (Figure 6). In MRI, lesion showed hyperintensity in T1 weighted & was isointense in T2 weighted images (Figure 7 & 8). Systemic examination was normal. Blood Investigations like TC, DC, ESR, Peripheral smear were normal. Chest X ray was normal, Mantoux was Negative & Ultrasound Abdomen was normal.

Figure 4 FFA of Both eyes . Left eye Macular lesion showsc hyperfluorescence in early choroidal phase with increasing in intensity in late phase and multiple blocked fluorescence corresponding to RPE stippling over the lesion.

Figure 5 OCT showing lesion in the choroid elevating the retina

Figure 6 CT scan shows focal thickening in the retina on temporal side of left eye

The above investigatory findings narrowed down our differential diagnosis to Choroidal hemangioma. Since Indocyanine green angiography (ICG) is an important investigation in making the diagnosis, patient was referred to another tertiary eye care hospital and it was done. ICG showed rapid filling and hyperfluorescence in early phase followed by early clearing and hypofluorescence. This confirmed our diagnosis of Choroidal hemangioma. Since Choroidal hemangioma is a benign tumour, the patient’s vision was not grossly affected. Hence the patient was kept under observation. 3rd month follow up of the patient showed the lesion is stationary without further progression. This case report is being published to emphasize the importance of diagnostic investigation techniques that differentiate benign Choroidal hemangioma from malignant Choroidal melanoma which helped in proper management instead of erroneous enucleation of eye for melanoma.

DISCUSSION

Choroidal hemangioma is a rare benign vascular hamartomatous tumour of uveal tract. These tumours exist in two forms (1) Circumscribed and (2) Diffuse, with diffuse tumors more commonly associated with encephalofacial angiomatosis (STURGE -WEBER SYNDROME). Previously, circumscribed lesions were considered as a part of Sturge-Weber Syndrome but now it is considered as separate entity1,2,3.Circumscribed Choroidal hemangioma occurs between 3rd and 6th decade. There is a speculation that these tumors are congenital in nature with the diagnosis made later in life. During embryogenesis, persistent AV shunts normally occurs and it is these shunts that may be responsible for the development of Circumscribed Choroidal hemangioma1,3,4. Clinical features of Choroidal hemangiomas depend on the secondary changes of hemangioma which includes Sub-retinal fluid, Neovascular glaucoma. Exudative retinal detachment, Irregular pigmentation on the surface of the tumor, Retinal Pigment Epithelial changes, Focal Ossification, Fibrous plate formation and Cystoid Macular Edema4.

Clinical characteristics of Circumscribed Choroidal hemangioma is unilateral presentation, occasional visual loss, tumour located in the posterior pole temporal to disc in the macular area, Orange red lesion, round to oval in shape, FFA showing early hyperfluorescence with late staining and B-Scan showing medium to high internal reflectivity4. Indocyanine green angiogram is an important tool in diagnosing Circumscribed Choroidal hemangioma. In early phases there is rapid filling and hyperfluorescence followed by faster clearing of the dye and relative hypofluorescence of the tumor with peripheral hyperfluorescence known as wash out pattern1,6.
In MRI, lesions show Hyper intensity in T1 weighted & appear isointense or Hyperintense in T2 weighted images. Circumscribed Choroidal hemangiomas have no malignant potential but may show progressive enlargement. Since Choroidal hemangioma is a benign tumour patient may simply be observed if vision is not affected and there is no extensive serous Retinal detachment. Treatment is generally reserved for cases in which vision is grossly affected or threatened. Factors such as size of the tumour, location of tumour, extent of subretinal fluid, presence of cystoid Macular edema and related ocular features help guide management. Treatment of Choroidal hemangioma is directed towards decreasing subretinal fluid, maintaining vision and preserving the eye. Enucleation was done as it closely resembles melanoma so that malignancy is not missed.

Current treatment options for Choroidal hemangioma includes:

(a) Laser photocoagulation - By using Argon Laser which induces regression of tumour and absorption of Sub-Retinal Fluid.

(b) Transpupillary thermotherapy (TTT) - By using infrared radiation which produce tumour necrotizing effect without coagulation.

(c) Radiation – Plaque Brachy therapy, External beam irradiation and Proton beam irradiation.

(d) Photodynamic therapy.

REFERENCES


