Choroidal Hemangioma: A Diagnostic Challenge

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ABSTRACT
Choroidal hemangioma is an uncommon, benign vascular tumor and typically presents between second to fourth decade of life. It is usually asymptomatic but may presents with reduced visual function, metamorphopsia, and photopsia. We report a case of 33-year-old woman with sudden diminution of vision and floaters. On clinical and radiological findings, a suspicion of choroidal melanoma/metastasis was kept for which enucleation of eye was performed. But in contrast with the above findings a diagnosis of choroidal hemangioma was made on histopathology. Diagnosis of choroidal hemangioma is challenging as it can mimic serious and malignant intraocular lesions. Our case is interesting in view of its rarity and the diagnostic dilemma which it presents to the ophthalmologists.

Keywords: Hemangioma, Eye Ball, Choroidal Melanoma, Metastasis

Introduction
Choroidal hemangioma is a rare, benign vascular tumour however exact incidence is not known. It typically presents between second to fourth decade of life. It can either be asymptomatic or presents with varied symptoms like reduced visual function, metamorphopsia, photopsia and even retinal detachment1. It can either presents as a circumscribed mass or as a diffuse variant. Circumscribed tumors occur without any associated local or systemic anomalies. Diffuse choroidal hemangiomas are usually evident at birth, as a part Sturge-Weber syndrome and are associated with ipsilateral cutaneous lesions, bilateral ocular signs and venous hemangioma of leptomeninges2,3. Diagnosis of choroidal hemangioma is challenging as it can mimic serious intraocular lesions like choroidal melanoma, metastasis and other inflammatory lesions4. We are reporting a case of choroidal hemangioma which was misinterpreted as choroidal melanoma/metastasis on clinical and radiological findings.

Case Report
A 33-year-old woman presented with history of sudden diminution of vision and floaters in left eye with no complaints in right eye. Her visual acuity was reduced in left eye i.e. finger counting close to face (FCCF) and 6/6 in right eye. There were no signs of redness, itching, watering or squint and diplopia in both eyes. However, intraocular pressure was slightly raised to 19mm Hg in left eye. Patient has no history of hypertension, diabetes or any other significant family history. On fundus examination of left eye, vitreous hemorrhage was seen and steroids were given for a week for proper visualization of the lesion. After one week the fundus examination showed orange-red area measuring 3mm near choroid inferior to optic disc. MRI showed lesion in left globe which was hyperintense on both T1 and T2, showing intense enhancement (Figure 1). Thus suspicion of choroidal melanoma or metastasis was kept. As the visual acuity was greatly reduced and a suspicion of malignant tumour was kept on radiological examination; enucleation of the left eye was performed and salvaging the right eye.

We received a specimen of left eyeball measuring 2.2 x 2 x 2 cm with attached part of optic nerve 0.3 cm in length. On examination, external surface was smooth and on cut it was filled with vitreous humor and hemorrhage. As no visible growth was identified so whole of the eyeball was processed. Ill defined thickening of 3mm in wall of posterior chamber was observed. On microscopic examination of this thickened area; fibrous bands composed of collagen were noted (Figure 2). Overlying this area there was a tumorous mass measuring 1.2 mm, composed of numerous thin walled vascular channels of different sizes lined by endothelial cells. The surrounding stroma showed few melanin containing macrophages dispersed in fibrocollagenous tissue. There was no stratification, nuclear atypia and mitotic activity in lining of vascular channels (Figure 2). Hence, final diagnosis of hemangioma was given.

Discussion
Choroidal hemangiomas are a diagnostic challenge as they clinically and radiologically mimic intraocular tumors. Differential diagnosis includes choroidal melanoma, metastatic tumors, choroidal osteoma, central serous chorioretinopathy and various inflammatory lesions4. In 38% of cases it is misinterpreted as malignant tumour so
appropriate diagnostic tests like ophthalmoscopy, fluorescein angiography, B-scan ultrasonography, indocyanine green angiography, Computerised tomography/Magnetic Resonance imaging (CT/MRI), guided fine needle aspiration cytology (FNAC) will facilitate the correct diagnosis. In our case it was mistaken as choroidal melanoma or metastasis on clinical and radiological findings however on histopathology a definitive diagnosis of hemangioma was made. Choroidal hemangioma on A and B scan shows high internal reflectivity whereas in melanoma the reflectivity is either low or medium however both appears as button or dome shaped. Ultrasonography provides a great deal of information to allow differential diagnoses of intraocular diseases. However, ultrasonography alone is not enough to make a precise diagnosis. Sometimes, it is necessary to depend on other imaging methodologies. Choroidal melanoma on MRI shows hyperintensity on T1 whereas it is hypointense on T2; however, in certain conditions like extensive necrosis or hemorrhage, retinal detachment melanoma can show varying intensities. On gadolinium diethylenetriamine penta-acetic acid (Gd-DTPA) it showed moderate to marked enhancement. Choroidal metastasis can also show varying signal intensities like hyperintensity on both T1 and T2 or hyperintense on T1 and hypointense on T2.

Choroidal hemangioma on the other side shows characteristic features on MRI due to high vascular flow. Mostly hemangioma shows hyperintensity on both T1 and T2. Choroidal osteomas show calcification on CT scan and ultrasonography.

It is evident from the above discussion that at times clinical and radiological findings can be confusing while differentiating these entities. Hence, histopathological examination is required for a definitive diagnosis. Choroidal hemangioma can be either circumscribed or diffuse. Circumscribed hemangiomas (CCH) are commoner and occur in absence of systemic disease whereas diffuse hemangiomas are uncommon and are usually associated with Sturge Weber syndrome and also, they require aggressive treatment to prevent visual loss from retinal detachment. The management of hemangiomas depends on the character of lesions. Various modalities of treatment include laser photocoagulation, radiotherapy, trans pupillary thermotherapy and photodynamic therapy and anti-vascular endothelial growth factor (VEGF). VEGF is a potent stimulator of endothelial proliferation and when secreted by tumor cells increase permeability of vascular endothelium. Anti-VEGF therapy leads to reduction in endothelial fenestrae and alter intercellular adhesion molecules which leads to decreased exudation. Anti-VEGF can help in decreasing macular edema and subretinal fluid complications resulting from choroidal hemangioma. Thus, the newer anti-VEGF therapy can be a promising adjuvant therapy for treating longstanding CCH with macular edema along with photodynamic therapy.

This case is interesting and challenging as hemangiomas are benign tumours and are generally mistaken as malignant ones so definitive diagnosis must be made for prognostication and appropriate management of patients.

References

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