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A rare case report of incidental circumscribed choroidal hemangioma in a phthisical eye: An overview of clinical manifestations and histopathological diagnosis

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Abstract:

Circumscribed choroidal hemangioma can mimic other conditions affecting the posterior segment of the eye, such as choroidal melanoma, melanocytoma, Coats' disease, choroidal metastasis, central serous chorioretinopathy, or nonspecific retinal detachment. A 43-year-old woman reported experiencing significant vision loss in her right eye over the past three years. Upon slit-lamp examination, the right eye exhibited a total cataract, a shallow anterior chamber, and endophthalmos. Since the eye was no longer functional, enucleation followed by the placement of a silicone ball implant was recommended. Histopathological analysis of the removed eye revealed features consistent with phthisis bulbi and a localized overgrowth of numerous blood vessels in the choroid, indicative of a choroidal hemangioma. The presence of a choroidal capillary hemangioma in a phthisical eye is an extremely rare finding. It may lead to significant vision impairment due to secondary complications such as exudative retinal detachment, neovascular glaucoma, cataract formation, and optic nerve atrophy.

Keywords: Choroidal hemangioma, phthisis bulbi

Case history:

A 43-year-old woman presented to a tertiary care center in northeast India with a primary complaint of progressively worsening vision loss in her right eye over the past three years. She also reported experiencing intermittent, throbbing pain in the same eye for the past year. Her medical history

included blunt trauma to the right eye eight years ago, which had been resolved on its own.

On clinical examination, the right eye had no perception of light, while vision in the left eye was normal (6/6). Slit-lamp examination of the right eye revealed a mature (total) cataract, a shallow anterior chamber, and a sunken appearance

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(endophthalmos). The intraocular pressure in the right eye was significantly reduced to 6 mmHg. A dilated fundus examination provided no view of the retina in the right eye, whereas the left eye appeared normal.

B-scan ultrasonography of the right eye demonstrated reduced axial length, a dense (echogenic) lens, and highly reflective echoes in the vitreous cavity. There was a marked increase in retino-choroidal thickness along with features consistent with phthisis bulbi. [Figure 1a] The patient was referred to the vitreoretinal clinic for further evaluation. As the right eye was nonfunctional, enucleation followed by placement of a silicone ball implant was recommended and performed by an oculoplasty surgeon after obtaining informed consent from the patient and her guardians. The enucleated eyeball was sent to the ocular pathology lab for histopathological evaluation.

Grossly, the enucleated eyeball measured in all dimensions. The optic nerve was cut flush to the

surface. A transillumination defect was not seen. The eyeball was sectioned vertically. [Figure 1b]

Microscopic examination of the hematoxylin-eosinstained tissue revealed hypertrophy of the corneal epithelium with subepithelial stromal scarring and vascularization. The angle of the anterior chamber was obliterated with a shallow anterior chamber. The intraocular lens was disintegrated. There was disorganization of the uveal structures, noted with atrophy of the retinal layers. The vitreous cavity showed oedematous fibrocollagenous tissue with proliferation of engorged vessels and capillaries. A solitary zone of choroidal vascular engorgement and proliferation was also noted. Focal calcification with ossification was also noted within the choroidal tissue. The scleral coat was thickened. The overall vitreous cavity and choroid were moderately infiltrated by chronic inflammatory cells. These findings are consistent with phthisis bulbi, with a coincidental circumscribed choroidal hemangioma observed. [Figure 2a and 2b]

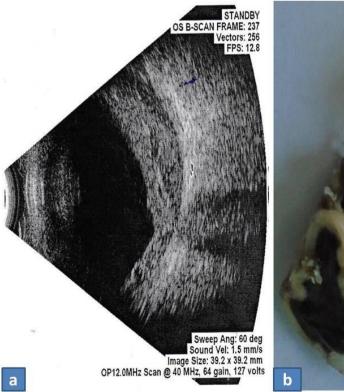




Figure 1: a) B-scan ultrasonography showing a marked increase in retino-choroidal thickness along with reduced axial length; b) Gross enucleated eyeball showing a small intraocular brownish mass at the posterior aspect of the eye

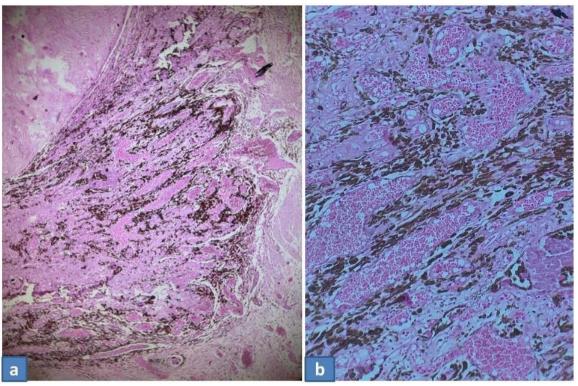


Figure 2 (a, b): A solitary zone of choroidal vascular engorgement and proliferation suggestive of solitary choroidal hemangioma (H&E-stained photomicrograph, 100X and 400X)

Discussion:

Choroidal hemangioma is a benign vascular tumor present from birth. It is classified into two types based on its extent. Circumscribed choroidal hemangiomas (CCHs) are single, clearly defined lesions typically located behind the equator of the eye. In contrast, diffuse choroidal hemangiomas appear as widespread, poorly defined thickening of the choroid, often resembling a "splashed ketchup" pattern and affecting multiple zones. Diffuse hemangiomas are commonly associated with other clinical features seen in Sturge–Weber syndrome [1].

These tumors are considered non-proliferative because the endothelial cells lining their blood vessels do not multiply. However, they can gradually increase in size due to venous congestion within the lesion, rather than from cellular growth or division [2].

CCH is a type of hamartoma that can be present from birth. However, most individuals begin to show symptoms and seek medical attention later in life, typically between their 40s and 60s [3]. Diagnosing CCH can be challenging in certain cases, as it can resemble various other conditions affecting the posterior segment of the eye. If there is any doubt, timely referral to a retinal specialist is essential for accurate diagnosis [4].

Various treatment options are currently available, such as argon laser photocoagulation, episcleral plaque radiotherapy, external beam radiation, proton beam therapy, transpupillary thermotherapy, photodynamic therapy, and intravitreal anti-VEGF injections. Enucleation is considered a last resort and is typically performed only in cases of painful, blind eyes when all other treatments have proven ineffective [5].

Conclusion:

Choroidal hemangioma can lead to significant vision impairment due to secondary complications such as exudative retinal detachment, neovascular glaucoma, cataract formation, and optic nerve atrophy. These issues affecting the posterior segment of the eye may eventually result in phthisis bulbi. It is important for pediatricians, neurologists, and ophthalmologists to be well-informed about this rare condition to ensure timely counseling and appropriate management for affected patients.

Disclosure:

The authors declare that they do not have any conflict of interest or financial involvement related to this study.

Contributions by each author:

- 1. **Dr. Bidhan Chandra Das** Interpretation of H&E slides, writing the manuscript, conceptualization, and designing of the case.
- Dr. Dipankar Das- Interpretation of H&E slides, documentations, and review of the manuscript.
- 3. **Dr Ganesh Chandra Kuri**-. Clinical diagnosis and providing surgical samples.
- 4. **Mr. Apurba Deka-** Preparation of slides, data collection, and typing of manuscript.

Institutional Ethics Committee approval was obtained for the publication.

Declaration of patient consent

The authors certify that they have obtained the appropriate patient consent form. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initial will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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