Choroidal Osteoma: A Rare Case Report

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Abstract
we report a case of choroidal osteoma in a 30-year-old female who came to us with complaint of gradual diminution of vision in right eye from one month. On fundus examination there was a peripapillary orange yellow coloured mass with well-defined margins and extending to fovea. B scan ultrasonography revealed sheet like calcification consistent with choroidal osteoma. Optical coherence tomography showed peripapillary mass, sub retinal fluid and choroidal neovascular membrane.

Keywords: choroidal osteoma, choroidal choristoma, choroidal neovascular membrane

Introduction:
choroidal osteoma is a benign tumour composed of mature bone which was first described by Gass et al in 1978(1)(2). The aetiology of this rare tumour remains unknown till date. It is typically found in posterior pole of healthy young women and is mostly unilateral (3). However, some male, young children and adults above 30 years of age have been found with choroidal osteoma. The diagnosis of choroidal osteoma is based on its characteristic clinical profile, ocular ultrasonography and fundus fluorescein angiography and OCT.

Case Presentation
A 30-year-old female presented to outpatient department of our hospital with complaints of diminution of vision in right eye, which was gradual in onset and rapidly progressive over a one-month period. There was no history of any systemic illness and the patient was not on any medication. There was no history of ocular trauma.

On examination Best corrected visual acuity was hand movements in the right eye and 6/6 in the left eye. There was right RAPD and left pupillary reaction were normal. Anterior segment examination was normal and IOP was within normal limits in both eyes.

Fundus examination revealed a yellow orange choroidal mass surrounding the optic disc temporally, superiorly and nasally. The mass extended to fovea and there was overlying RPE atrophy. The lesion had well defined margins and extended about 4-disc diameters on temporal side, 3-disc diameters superiorly, and 2-disc diameters on nasal side (Fig 1). The other eye fundus examination was unremarkable.

Investigations:
1. Fundus fluorescein angiography (FFA) showed early hyper fluorescence and late staining of CNVM (fig 1 b)
2. B-scan ultrasonography of right eye showed slightly elevated mass. It was highly reflective linear broad-based lesion centred just adjacent to right optic disc with acoustic shadowing giving pseudo-optic nerve appearance suggestive of choroidal osteoma. Fig 2a
3. Computed tomography showed calcific plaque on posterior wall of globe with bone like homogeneity. The plaque was seen around the right optic disc. Fig 2b
4. OCT of the right eye showed distorted retinal pigment epithelium, subretinal fluid and Choroidal neovascular membrane. (Fig3)

5. Vitreous tap of the patient was also taken and after thorough cytologic examination, only proteinaceous material was present and no cells were seen.

6. MRI revealed hypointense signal on T2 weighted image, no soft tissue mass was seen.

No abnormality was detected on chest, breast and abdominal examination as well as imaging.

All the routine baseline investigation were within normal limits including CBC, serum calcium, phosphorus and alkaline phosphatase concentration.

**Discussion**

Choroidal osteoma is a rare benign tumour characterized by the presence of cancellous bone within the peripapillary choroid (4). It mostly occurs unilaterally in young, healthy women, but 33% of cases occur in males and 21% of cases are bilateral. [5] It generally occurs as a sporadic trait. The incidence of the disease is extremely rare. No data on the prevalence are available in the literature. To date, only a few cases have been reported. There is no racial predilection; however, most reports were of white patients [3].

In contrast to other type of intraocular ossification choroidal osteoma occurs in otherwise healthy eyes. Gass In 1978 first described healthy young women with characteristic ophthalmoscopy appearance of a slightly elevated, juxtapapillary, yellowish orange, choroidal lesion with well-defined margins.[6][2]. The lesion colour depends on the level of overlying retinal pigment epithelium (RPE) depigmentation [6]. In the early stages, they tend to be orange-red in colour, whereas in later stages they have a yellowish tint due to RPE depigmentation [7]. The shape of the tumour is generally oval or round with characteristic well-defined scalloped or geographic margins. The margins can have blunt pseudopod-like projections. The vasculature within the choroidal osteoma is sometimes visible. The most common causes of visual loss in these patients are due to serous detachment, sub retinal fluid
Choroidal neovascularization (CNV), photoreceptor loss, choroidal and RPE atrophy associated with decalcification and macular scarring (7). Sub retinal fluid and CNVM in our patient is the possible reason for her vision loss. There are case reports of atypical presentation of choroidal osteoma with elevated juxtapapillary lesions producing diagnostic dilemma. (8)

The pathogenesis of choroidal osteoma is unknown. It has been suggested that it is an osseous choriostoma [8]. This suggestion is supported by peripapillary location, a site favoured by other developmental tumours and by occurrence of the osteoma in the absence of any other disease process. An alternative cause is secondary ossification following inflammation or trauma to the orbit or periorbital tumour [9] Case report of de-novo appearance of a choroidal osteoma in an eye, following years after laser photocoagulation for BRVO (Branch Retinal Vein Occlusion) has been reported. [10] Choroidal osteoma resembles a choriostoma in its tissue composition, its apparent onset in youth, and the absence of other preceding ocular disease processes. Features that are atypical for a choristoma are sexual preponderance in females and new development and growth of the tumour during adulthood. Other etiological factors like inflammation, trauma, hormonal changes, and heredity have been speculated. It has also been seen in association with histiocytosis, stargardt disease, and polyoid choroidal maculopathy.

Choroidal osteoma needs to be differentiated from amelanotic choroidal melanoma, amelanotic choroidal nevus, metastatic carcinoma to choroid, circumscribed choroidal haemangioma, idiopathic sclerochoroidal calcification, posterior scleritis and choroidal cartilage (2). Amelanotic choroidal melanoma, amelanotic choroidal nevi and metastatic carcinoma are usually more elevated and have less defined margins. Metastatic carcinomas are usually associated with serous retinal detachment. Idiopathic sclerochoroidal calcification is rather multifocal in pattern and is more likely to be bilateral compared to choroidal osteoma. (11)

Complications of choroidal osteoma include serous detachment choroidal neovascularization (CNV), subretinal haemorrhage, subretinal fluid (SF), decalcification status, and overlying retinal pigment epithelium (RPE) atrophy; of these, sub retinal fluid with serous detachment and CNV is known to be a major cause of visual impairment. (5)

The growth of the choroidal osteoma is about 0.37 mm per year. 50% of tumours show spontaneous resolution by the process of decalcification. This decalcification can also be triggered by various treatment modalities like Laser photocoagulation, photodynamic therapy by inducing osteoclastic activity in tumour. This resolution is characterised by atrophic yellow greyish lesion with overlying retinal pigment epithelium atrophy.

There are no known treatment methods for choroidal osteoma per se; however, there have been attempts to treat secondary CNV in patients with choroidal osteoma by surgical removal, laser photocoagulation therapy, transpupillary thermotherapy, and photodynamic therapy; however, these have shown limited effectiveness. (5) As anti-vascular endothelial growth factor agents have been accepted as the most effective method for treating CNV, they have also been used to treat patients with CNV accompanying choroidal osteoma, and several case reports have shown acceptable efficacy in improving retinal structure and visual acuity. (5)

After giving proper information to the patient and after taking her consent we injected one dose of anti vefg bevacizumab (Avastin) in our patient. She reported back with remarkable improvement in her vision from hand movements to 5/60 on 5th day. The improvement in her vision is possibly due to decrease in SRF as shown by optical coherence tomography and optic nerve decompression.

Optical coherence tomography after one week showed decrease in subretinal fluid and cnvm.

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