

Bilateral Choroidal Osteoma - A rare entity

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Abstract

A 24 years old female presented to us with the chief complaint of dimness of vision in her right eye since few days and loss of vision in right eye since childhood. On examination she was having well defined yellow to orange slight elevated lesion at macula and peripapillary area and in the left eye marked depigmentation of macula with subretinal fibrosis. She had undergone Optical coherence tomography (OCT) and Ultrasonography (USG-Bscan) of both the eyes. On the basis of clinical findings and investigation, diagnosis of choroidal osteoma was made. The purpose of reporting this case here is that bilateral choroidal osteoma is a very rare entity and whenever a young female is presenting with this kind of lesion in retina, we should keep the possibility of choroidal osteoma in differential diagnosis.

Keywords: Choroidal osteoma, Choroidal tumors, Choroidal neovascular membrane, Antivascular endothelial growth factor.

Introduction

Choroidal osteomas are benign, juxta-papillary tumors that do not require any kind of treatment. They occur mostly in young females (90%) and are unilateral (75%)¹ Direct ophthalmoscopy, OCT, fundus fluorescein angiography, Indocyanine angiography, USG, X-ray, CT scan etc are used to make the diagnosis. Majority of patients with choroidal osteoma maintain good vision. The probability of loss of visual acuity (20/200 or worse) is more than 50% by 10 years in these patients, as reported by Shield's et al^{2,3} CNVM is one of the major cause of vision loss noted in these patients. Occurrence of subretinal fluid in the absence of CNVM is also found in few case reports. Anti VEGF intravitreal injections were used successfully in those cases.

Case Report

A 24 years old female presented to us with the chief complaint of dimness of vision in her right eye since few days. Her left eye was having low vision since childhood. Best corrected visual acuity was recorded 6/18, N8 in the RE and hand movements in the LE. Slit lamp examination and applanation tonometry was found to be normal in both the eye. Fundus examination showed a well defined yellowish orange, slightly elevated lesion at the macula and along the superior and inferior arcade as shown in (Fig. in1a). Fovea appears spared. There was no haemorrhage or CNVM noted near the edges of lesion. LE fundus showed diffuse retinal pigmentary atrophic changes at the macula and peripapillary area with signs of subretinal fibrosis at the macula and disc gliosis as seen in (Fig. 1b). OCT of RE showing elevated retino-choroidal layer along with presence of subretinal fluid (Fig. 2a). OCT of LE shows irregular elevation of choroid with thinning of all the retinal layers (Fig. 2b). On USG (B scan) we noted the high reflective multiple choroidal lesion persisting on low gain with

acoustic shadowing in the RE (Fig. 3). The same was noticed in the left eye also with irregular thickening of choroid with hyper reflectivity. On the basis of clinical findings, OCT and USG test diagnosis of choroidal osteoma is made. Fundus fluorescein angiography was deferred as patient was not willing for any interventional procedure.

In view of SRF overlying the osteoma in the right eye, she was advised intravitreal injection Ranibizumab (0.5 mg in 0.05 ml) and the same was given once. On follow-up SRF persisted so we preferred to watch. Patient was continued in follow up for two years with same size of lesion, persistent SRF on OCT and maintained same visual acuity.

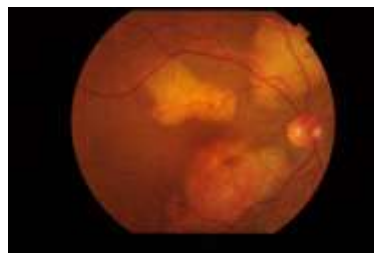


Fig. 1a: Fundus photo RE showing a distinguished yellow-to-orange lesion in the macula and along the arcade

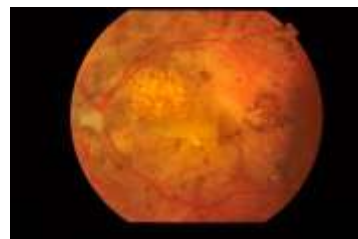


Fig. 1b: Fundus photo LE showing extensive retinal pigment epithelium atrophy at macula and peripapillary area along with subretinal fibrosis at macula and disc gliosis

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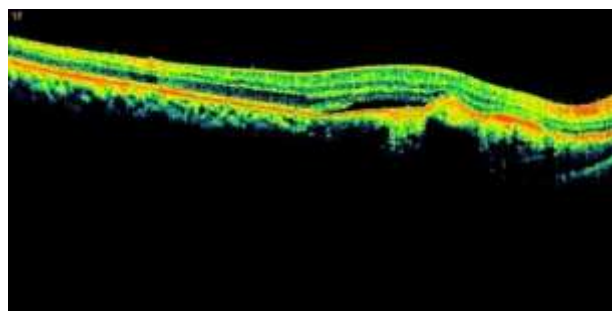


Fig 2a: OCT RE showing SRF overlying the elevated choroid

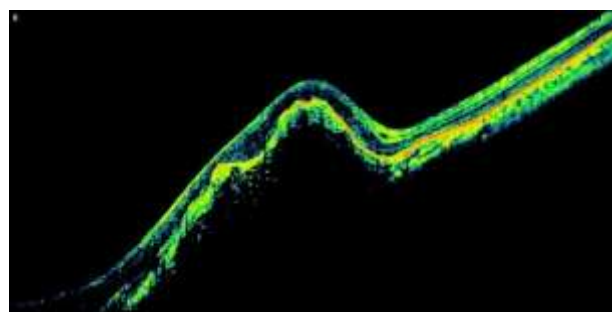


Fig 2b: OCT LE showing irregular elevation of choroid with thinning of retinal layers

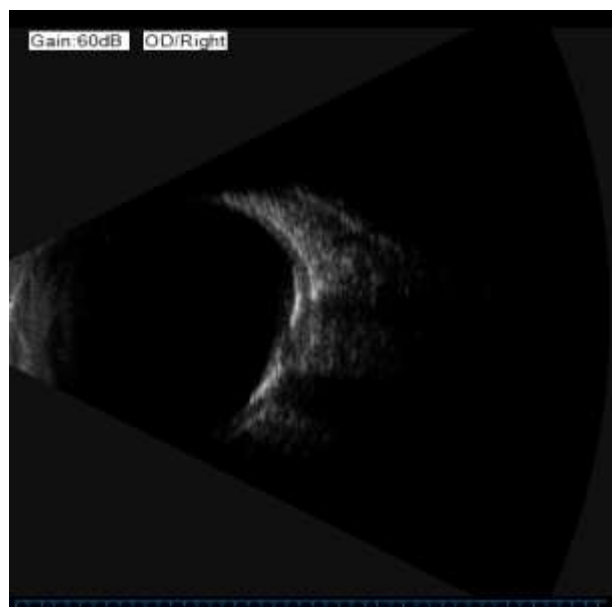


Fig. 3: USG B-scan RE showing hyperreflectivity of the retino-choroid layer with after shadowing

Choroidal osteoma is a rare condition. The largest case series in the literature includes 61 patients over a 26 year-span of ocular oncology practice in a major tertiary care centre.¹ Its precise frequency remains unknown. It occurs in all races but predominantly affects young adults in their early twenties with a large range from few months old to late sixties. It has a predilection for women and is unilateral in around 80 percent of cases.^{1,2,3} The exact etiology of choroidal osteoma is unknown. There has been found no

association between choroidal osteoma and any abnormality in blood chemistries such as calcium, phosphorus, alkaline phosphatase, complete blood count and urinalysis.³ Choroidal osteoma appear as a deep, orange-yellow lesion with distinct geographic or scalloped borders and branching vessels on the surface of the tumor. The lesion colour relates to the level of retinal pigment epithelium (RPE) depigmentation. In early stages choroidal osteomas tend to have an orange-red colour, whereas in later stages they have a yellowish tint due to RPE depigmentation.⁴ Histopathology illustrates dense bony trabeculae with marrow spaces traversed by pathognomonic dilated thin-walled blood vessels, termed spider or feeder vessels.⁵ Tumor decalcification and resolution, originally described by Trimble⁶ in 1988 occurs in around 50 percent of case and is associated with long term poor visual acuity as seen in our patient in LE.

Choroidal osteomas are included in the large differential diagnosis of ocular tumors and ocular calcification. These include amelanotic choroidal melanoma, amelanotic choroidal nevus, metastatic choroidal carcinoma, circumscribed choroidal hemangioma, disciform macular degeneration, posterior scleritis, idiopathic sclerochoroidal calcification, choroidal cartilage, leukemia and retinoblastoma.

Choroidal osteomas are also known to be associated with subretinal fluid, hemorrhages and serous retinal detachment. Serous retinal detachment occurs in choroidal osteoma frequently in the absence of CNV. It is speculated to be the result of multiple pinpoint RPE leakage sites over the osteoma as detected by fluorescein angiography. Alternatively, atrophy of the RPE and Bruch membrane is thought to decrease their capacity to remove subretinal fluid emanating from the disrupted outer blood-retinal barrier. In our case we noticed the same thing in the RE i.e. presence of SRF on OCT without any CNVM.

There is no standard protocol for treatment for choroidal osteomas, but therapies are directed for complications arising from choroidal neovascularization (CNV) and subretinal fluid. Focal Laser, Transpupillary therapy, photodynamic therapy and intravitreal anti VEGF injections are tried with variable success rate. We also used intravitreal anti VEGF injection once in the RE but the amount of SRF we noticed before giving injection was found to be the same even after giving injection in contrast to earlier reports⁶ We decided to watch this case and she maintained the same visual acuity in 2 year follow up.

Abbreviations

Optical coherence tomography (OCT), ultrasound (USG), choroidal neovascular membrane (CNVM), Retinal pigment epithelium (RPE), CT scan (computed tomography), right eye (RE), left eye (LE), subretinal fluid (SRF), vascular endothelial growth factor (VEGF)

Source of Funding

None.

Conflict of Interest

None.

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