

Multiple Choroidal Osteoma – A Rare Case Report

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Abstract

Choroidal osteoma is rare clinical entity of unknown etiology, characterized by formation of mature cancellous bone within the choroid. It typically affects young females, with no racial predilection. Vision loss occurs mainly due to photoreceptor degeneration secondary to decalcification and/or development of choroidal neovascularization especially if located at the sub foveal area. Our case is 9 y old male child identified incidentally with multiple yellowish white well demarcated lesions in the left eye suggestive of choroidal osteoma associated with nearby areas of RPE (retinal pigment epithelium) atrophy and depigmentation. SD-OCT (spectral domain optical coherence tomography) demonstrated high reflectivity from the choroid and atrophy of the overlying retinal layers. USG B-scan demonstrated multiple highly reflective calcified lesions within the choroid suggestive of choroidal osteoma.

Keywords: RPE; Choroid; Osteoma; Decalcification

Introduction

Choroidal osteoma is a benign ossifying disorder with formation of mature cancellous bone in the choroid. The exact etiology is still unknown. First case was presented at the meeting of Verhoeff Society in 1975 and was published by Gass et al. [1]. Incidence of the disease is extremely rare. No data available on the prevalence in the literature. Till now only few cases are reported. The largest case series include 74 eyes of 61 patients with choroidal osteoma followed up over a duration of 26 years [2]. There is no racial predilection, yet most reported patients were Caucasians [3]. It typically affects the young adults with female predilection. It appears as orange yellow to yellow white lesions with distinct margin with blood vessels overlying them. The lesion colour depends on the level of overlying retinal pigment epithelium (RPE) depigmentation [1]. In early stages they tend to have orange-red in color, whereas in later stages they have yellowish tint due to RPE depigmentation [4]. The most common cause of visual loss in these patients are due to choroidal neovascularization (CNV) and/or photoreceptor loss [5], choroidal and RPE atrophy associated with decalcification [2,6,7].

Case Report

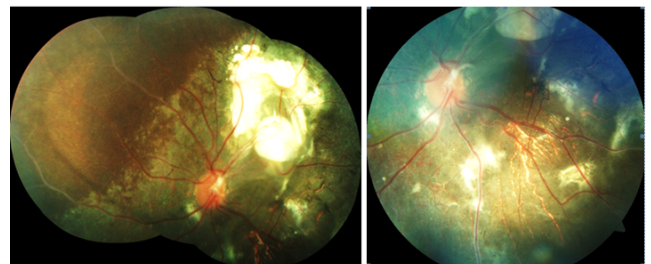
A 9 y old child diagnosed with pan-sinusitis was referred to us from the ENT department with complaints of swelling over left side of face involving left lower lid to rule out any ocular manifestation. Patient gave history of swelling over the left side of face, with duration of about 10 days. It was insidious in onset, gradually progressive in nature and involved the left lower lid, not associated with pain and fever. There was no associated systemic disease. On examination BCVA for left eye was hand movement and for right eye was 6/6. Patient was unaware of diminished vision in his left eye. On dilated fundus examination we observed two yellowish white lesion with well demarcated borders located superotemporally indicative of active lesion, associated with nearby areas of RPE depigmentation and pigment clumps extending

into macular area suggestive of degeneration of overlying retinal layers indicative of decalcified lesions in the left eye (Figures 1A and 1B).

Fundus fluorescein angiography (FFA) revealed areas of early granular hyperfluorescence corresponding to the areas of RPE depigmentation and late hyperfluorescence over the calcified lesion with some interspersed areas of hypofluorescence corresponding to the areas of pigment clumps in the left eye (Figure 2).

SD-OCT was performed and revealed high reflectivity from the choroid with marked thinning of overlying retinal layers including photoreceptor inner/outer segment junction (Figure 3).

USG B-scan of left eye demonstrated large irregular echogenic calcified lesion of 7.1 x 3.9 mm in the posterior choroid near to the optic disc region and extending up to optic disc and another smaller echogenic calcified foci in the posterolateral choroid both nasally and temporally. Nasal lesion was 1.3 mm and temporal lesion was 1.5 mm (Figure 4).



Figures 1 (A) and (B): Fundus photograph of the left eye showing multiple choroidal osteomas superotemporally and areas of RPE depigmentation.

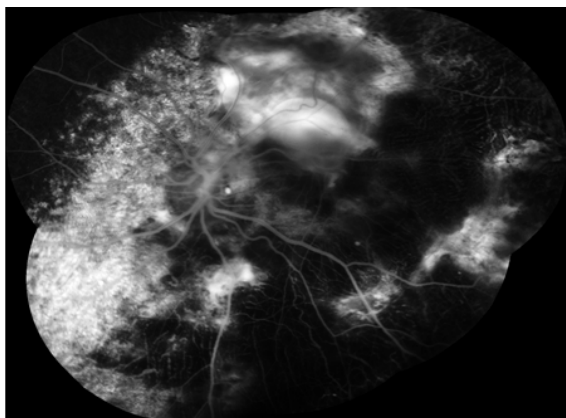


Figure 2: FFA of left eye showing areas of patchy hyperfluorescence corresponding to the areas of RPE depigmentation interspersed with areas of hypopigmentation corresponding to pigment clumps.

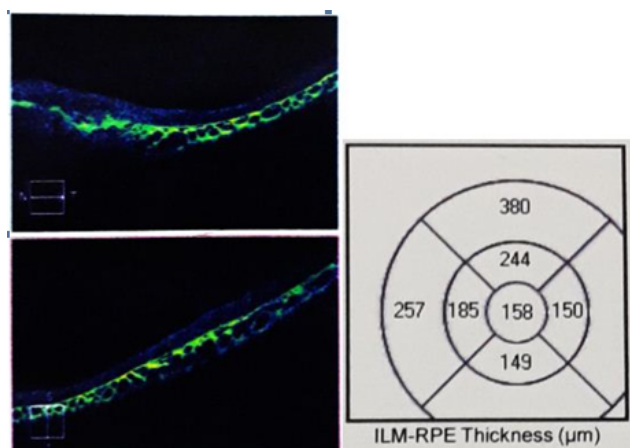


Figure 3: SD-OCT macula of left eye showing high reflectivity from the choroid and thinning of the retinal layers.

Examination of the right eye was normal. All routine blood investigations were normal including serum calcium and parathyroid hormone level. As there was already setup of decalcification with loss of RPE/photoreceptor and marked diminished vision, and also the lesion was extending into the foveal region, patient was asked for follow-up at monthly interval to monitor the subsequent progression or regression of the tumor, because of the non-availability to reliable treatment option.

Discussion

Choroidal osteoma is a rare benign ossifying disorder of the choroid. In our case the patient was 9 y old male child, whereas in largest case series on choroidal osteoma including 74 eyes of 61 patients followed up for a period of 26 years, Carol et al. demonstrated choroidal osteoma a disease of young females [2]. They found that choroidal osteoma showed evidence of growth in 51% of eyes and decalcification in nearly 50% of eyes by 10 years. In their series,

decalcification of choroidal osteoma was usually associated with poor vision [2]. Decalcification was hence considered as a significant risk factor for poor long-term visual acuity. Decalcification commonly occurs concurrently with overlying RPE alterations and atrophy of the choriocapillaries, both of which could lead to photoreceptor degeneration and poor visual acuity. Shields and colleagues found that the decalcified portion of osteoma displayed an overlying marked thinning to absent outer retina and photoreceptor layers (100%), compared with the calcified portion with preserved intact outer retina (95%) and intact photoreceptor layer (100%) [5]. The OCT (optical coherence tomography) findings of our case also revealed the diffuse atrophy of the outer retina on the decalcified lesion involving the macular area and explained why the vision was poor in our patient. The etiology of the choroidal osteoma is still unknown. It has been suggested that it is an osseous choristoma [8].



Figure 4: USG B-scan of left eye showing echogenic calcified lesion in the choroid.

This suggestion is supported by peripapillary location, a site favoured by other developmental tumors 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 tumor. A case of multiple osteoma developing in association with bilateral pseudotumors of the orbit raised the possibility that inflammation may have a part in the cause [9]. In our case there was associated pan-sinusitis with multiple choroidal osteomas. Treatment options for foveal choroidal osteoma are limited. PDT (photodynamic therapy) is a reasonable choice in the case of extrafoveal CNV (choroidal neovascularization) lesions. Observation is the indicated management where there are no symptoms, with fundus examination at regular intervals monitoring for signs of CNV. Shields et al. also reported a case of extrafoveal CNV successfully treated with PDT [10]. However, the author inserted a provision at the end of the case report that treatment of sub-foveal CNV with PDT may result in worse visual acuity due to decalcification and associated RPE loss. More recently anti-vascular endothelial growth factor (anti-VEGF) drugs have been used off license to treat CNV secondary to choroidal osteoma with good effect. In the future, more studies with long-term follow-up may help to define an appropriate time interval when intervention can be performed to prevent the tumor growth or decalcification.

Conclusion

Choroidal osteoma is a rare clinical entity of unknown etiology. The disease has the potential for poor long term visual acuity. Early detection of the disease with subsequent follow-up is mandatory to prevent vision loss due to early decalcification, photoreceptor loss and/or development of CNV.

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