

Choroidal Osteoma with Choroidal Neo Vascular Membrane



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ABSTRACT

Choroidal osteoma (CO) is a rare benign tumor usually unilateral and have a female preponderance. We report a case of a 34-year-old healthy male diagnosed with unilateral Choroidal Osteoma with an associated Choroidal Neo Vascular Membrane (CNVM). The diagnosis was confirmed based on clinical examination, typical findings on B-scan, Fundus fluorescein angiography, Indocyanine green angiography and Optical coherence tomography. The CNVM was treated with intravitreal Bevacizumab injections and showed a good response initially. There was a recurrence of CNVM over two years when patient was lost to follow-up. Treatment with intravitreal Ranibizumab bio similar injection helped to stabilize his vision. Regular follow-up is essential for timely treatment of recurrence of CNVM and optimum visual outcomes.

Key words: Choroidal Osteoma, Choroidal neo vascular membrane, Optical, Coherence tomography, Bevacizumab, Ranibizumab.

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INTRODUCTION

Choroidal Osteoma is a benign ossifying tumor of the choroid, usually unilateral. Young females are predominantly affected. Generally asymptomatic and slow growing tumor, defective vision can occur secondary to sub foveal decalcification, overlying RPE choriocapillaris atrophy and development of choroidal neovascular membrane (CNVM).^{1,2} The approximate 10-year risk for outcomes include tumor growth (50%), tumor decalcification (50%), vision loss (50%), and development of CNVM (30%).³

CASE PRESENTATION

A 34-year-old male presented with complaints of sudden painless decreased vision in the right eye for

last 15 days. His past ocular, medical and family histories were non-contributory. On examination his best-corrected visual acuity (BCVA) was 20/60 and N8 in the right eye and 20/20 and N6 in the left eye. Intra ocular pressure (IOP) and anterior segment examination were normal in both eyes. Fundus was normal in the left eye while right fundus showed a yellowish well circumscribed lesion of about 3-disc diameter in size with smooth surface and slight elevation with associated sub retinal hemorrhage and sub retinal fluid on the posterior pole. B-scan ultrasonography revealed a hyper-echoic lesion with acoustic shadowing, described in literature as a pseudo-optic nerve secondary to calcification (Figure 1). Fundus Fluorescein Angiography (FFA) and Indo Cyanine Green (ICG) showed an intense hyper fluorescence in the early phase with progressive leakage in the late phase, indicative of a classic choroidal neovascular membrane. There was an area of blocked fluorescence corresponding to the intra retinal hemorrhage and calcification of the tumor which was seen clinically. On Optical Coherence Tomography (OCT), normal inner retinal layer and abnormal outer retinal layer with sub foveal irregular hyperreflective lesion was indicative of the CNVM.

Hyperreflective lamellar pattern was suggestive of bony lamellae of CO. Sub retinal fluid (SRF) and intraretinal fluid (IRF) were also seen. A final diagnosis of Choroidal Osteoma with CNVM was made. Intravitreal injection of Bevacizumab (1.25mg in 0.05ml) was given. At one month follow-up, OCT showed resolving SRF and a second dose of intravitreal Bevacizumab injection was administered. After two months, there was an increase in metamorphopsia, though the BCVA was maintained at 20/30 and N8. OCT showed an increase in SRF. A third injection of intravitreal Bevacizumab was given, following which the patient was lost to follow-up.

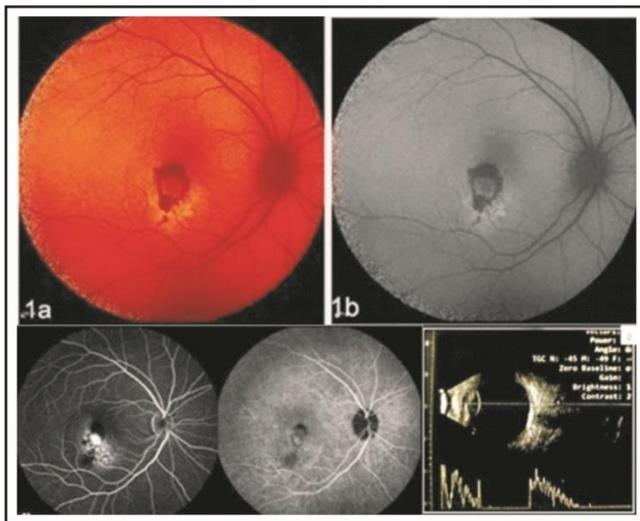


Figure-1: Top row shows 1a) Pseudo color image at presentation with elevated lesion of irregular margin and retinal hemorrhage at the fovea, 1b) FAF image shows central hypo Auto Fluorescence (AF) due to retinal hemorrhage surrounded by hyper AF margin. Second row: left image indicates FFA at presentation showing leakage due to type 2 CNVM. Middle: ICG image shows hypo fluorescence due to blockage by the calcified tumor. Right: B-scan showing Pseudo optic nerve due to calcification of Choroidal osteoma.

He presented again, 2 years later with complaints of worsening of vision to 20/80 and N12 in the right eye. An increase in tumor size to five-disc diameter was noted on fundus examination. Repeat FAF showed increase in the central hypo fluorescence with prominent hyperfluorescent margin (Figure 2). OCT was suggestive of normal inner retinal layer with abnormal outer retina and SRF. An enlarged hyperreflective mound like lesion was suggestive of CNVM, which had now enlarged, with significant SRF and IRF were seen. There was also an increase in the lamellar reflective pattern which extended from

inferior to the fovea to inferior arcade indicative of enlargement of the CO (Figure 3). A repeat FFA and ICG showed a well demarcated intense hyperfluorescence with progressive leakage in the late phase suggestive of the CNVM. Optical coherence angiography (OCTA) at this visit showed network of abnormal blood vessels surrounded by relatively homogenous choriocapillaris in the En face image of outer retina choriocapillaris slab. The corresponding OCT B scan showed increased flow signals below the level of RPE involving the lesion. (Figure 2). Despite a repeat intravitreal Bevacizumab injection, on one month follow-up there was worsening of vision with BCVA of 20/80 and N12.

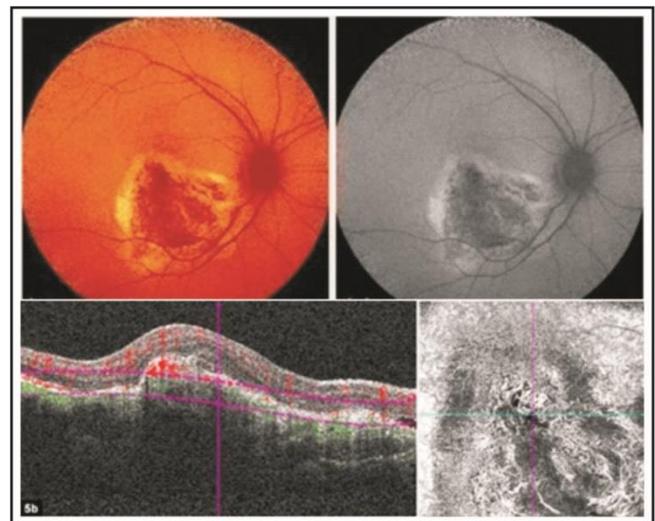


Figure 2: Top images show 2 years after the first presentation with increased size of the lesion, corresponding AF image shows central hypo AF surrounded by hyper AF margin. Bottom: En face image of outer retina choriocapillaris slab with network of abnormal blood vessels surrounded by relatively homogenous choriocapillaris.

In view of sub-optimal response, he was switched to intravitreal Ranibizumab bio similar (Razumab, Intas Pharmaceuticals, India), and received two injections at monthly interval. Post injection OCT showed reduced SRF (Figure 3) with his BCVA of 20/80 and N8. Metamorphopsia was reduced at 1 month review. The patient did not review back following this visit.

DISCUSSION

In 1978, Gass was the first to describe choroidal osteoma.¹ Few cases have been reported among males

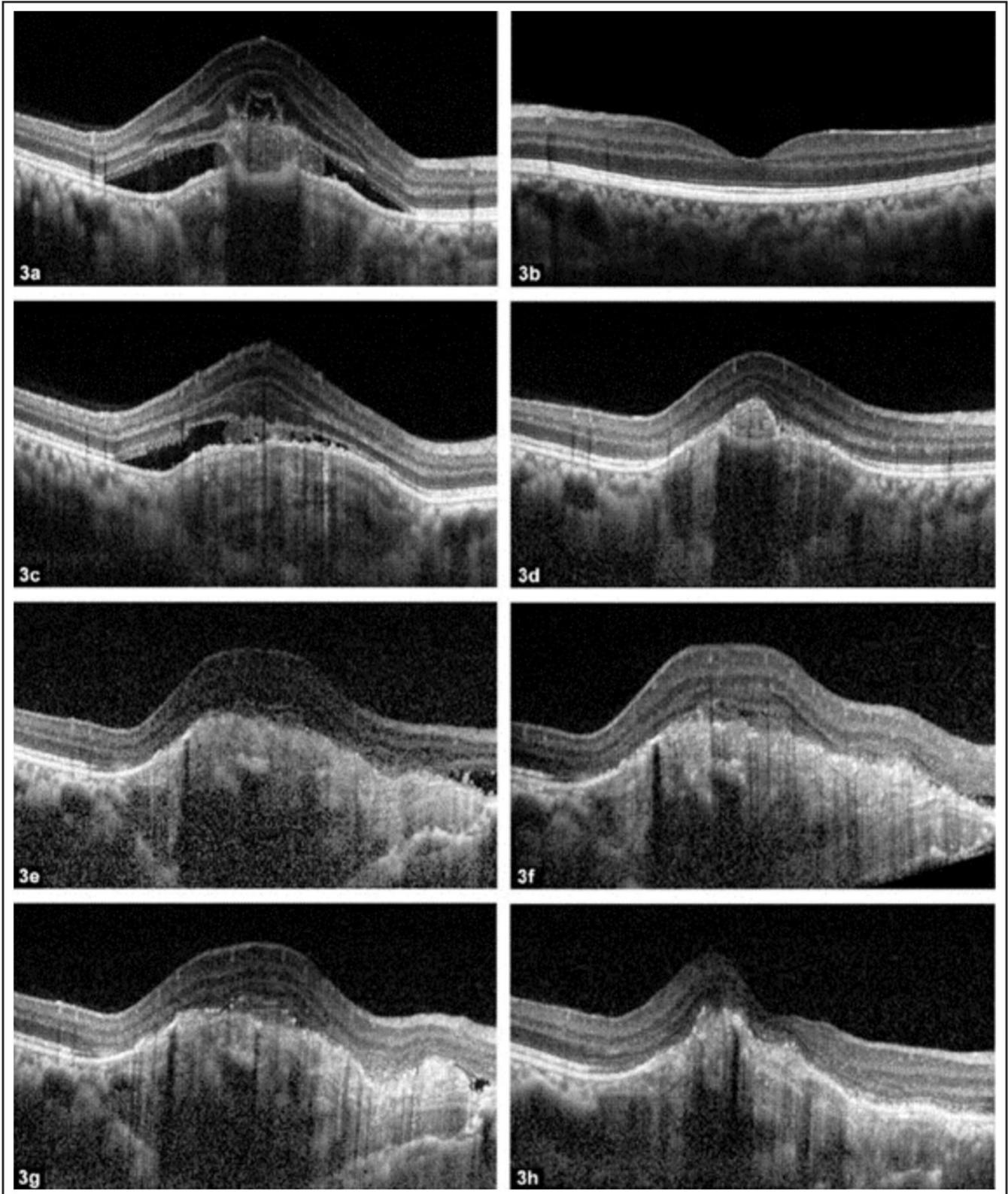


Figure 3: **A:** Shows OCT at presentation with normal inner retinal layers, SRF and a hyper-reflective mound like lesion below the level of RPE with lamellar reflective pattern. **B:** Shows normal retina in left eye. **C and D:** Show response to Bevacizumab injection. **E:** shows increase in the size of the tumor after 2 years. **F and G:** Shows responses to Ranibizumab therapy. **H:** shows response after 2 doses of Ranibizumab with regressed CNVM and well maintained inner retinal layers in the fovea.

and children.⁴ Our patient is a young man with a unilateral presentation. Generally, patients with calcified CO maintains good vision when compared to decalcified CO as decalcification induces overlying RPE atrophy and photoreceptor loss. Decalcification in CO can be assessed by OCT in which focal choroidal excavation with neuro sensory detachment, choroidal caverns and speckled hyperreflective dots suggestive of trabecular bone tissue within the tumor can be seen.^{5,6} There is no standard treatment to prevent the growth of choroidal osteoma or to stop decalcification. Therapies are directed to the management of Choroidal OsteomawithCNVM.^{7,8}

The possible pathogenesis of CNVM in CO is that atrophied RPE overlaying the tumor permits growth of new blood vessels or the osteoma itself facilitates the growth of new blood vessels.¹ Multi modal imaging plays an important role in diagnosing and prognosticating the outcome post-intravitreal injection. A high-quality OCT-A has proven to be a useful, non-invasive and cost-effective imaging method for detecting and monitoring treatment response of CNVM in Choroidal osteoma.⁹

Treatment modalities available for CNVM are laser photocoagulation, photodynamic therapy and anti-vascular endothelial growth factor (VEGF).¹⁰ Intravitreal anti VEGF is an effective therapeutic option for choroidal osteoma-associated CNVM, particularly when the CNVM is juxta foveolar or sub foveolar in location.¹⁰ In our case the BCVA improved from 20/60 and N12 (at presentation) to 20/30 and N8 with treatment. Worsening of the lesion with poor response to treatment following a lapse in review was noted after two years.

Switching the anti-VEGF to Ranibizumab showed significant response, with a decrease in the SRF and visual improvement on follow up. The increase in CNVM was associated with increased size of CO in this case, which was predominantly inferior to the fovea. The persistence of normal inner retinal layers and no signs of decalcification prove to be reasonable for sustained BCVA of 20/80 and N8.

CONCLUSION

The study underscores the importance of regular follow-up in patients with CNVM associated with CO to ensure timely detection and treatment of recurrence, thereby optimizing visual outcomes.

Conflict of Interest: Authors declared no conflict of interest.

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Manavi D Sindal; Head of Department: *Concepts, Design, Literature search, Data acquisition, Data*

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