ABSTRACT
An 18-year-old male was clinically diagnosed as having choroidal osteoma in August 2005. Ocular examination revealed 4 disk diameter orange colored elevated lesion in the left eye. Ultrasonography and computed tomographic (CT) scan of orbit, fluorescein angiogram (FA) and optical coherence tomography (OCT) confirmed the diagnosis of choroidal osteoma. The patient was followed up at regular intervals and on the 4th year follow-up developed a choroidal neovascular membrane (CNVM) within the osteoma. It was treated with intravitreal bevacizumab. Patient was stable for next 2 years followed by a recurrence of the membrane for which further injections were given to regress it. During this period there was consistent progression of the size of osteoma as well as its decalcified component. We documented progression of the osteoma along with regression of the CNVM with color photo and OCT for 7 years. Spectral domain features of the osteoma were noted in relation to changes occurring in its decalcified area.

Long-term follow-up suggests that choroidal osteoma along with the decalcification part within it progresses with time. Intravitreal bevacizumab is effective in treating the CNVM secondary to choroidal osteoma.

Keywords: Choroidal osteoma, Choroidal neovascular membrane, Intravitreal bevacizumab, Calcification, Decalcification.


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INTRODUCTION
Choroidal osteoma is a benign intraocular tumor of unknown etiology and is composed of mature bone.\(^1\)\(^2\) It is commonly juxtapapillary or peripapillary but may extend to the macula.\(^3\) It is yellow-white to orange-red in color with clumping of pigments. Occasionally decalcification can occur and is characterized by thin, atrophic regions with associated retinal pigment epithelium (RPE) atrophy.\(^4\)\(^5\) Decalcification can occur spontaneously or as a result of laser photoocoagulation or photodynamic therapy.\(^6\) Choroidal neovascular membrane (CNVM) may also develop.\(^2\) Patients with a choroidal osteoma may be totally asymptomatic or present with mild-to-severe visual loss or metamorphopsia. Here, we report a case of choroidal osteoma that has been followed up for the past 7 years. Periodical evaluation was done with color fundus photograph (CF), fluorescein angiography (FA), stratus and spectral domain (SD) OCT.

CASE REPORT
An 18-year-old male presented to us for a routine checkup related to a refractive error in August 2005. His past ocular, medical and family history was noncontributory. He was myopic and using spectacles for the last 7 years.

On ocular examination best corrected visual acuity (BCVA) was 20/20 in both eyes. Anterior segment examination and intraocular pressures were within normal limits. On fundus examination right eye was within normal limits and left eye revealed an orange colored elevated lesion around 4 disk diameter with well-defined margin suggesting a diagnosis of choroidal osteoma (Fig. 1A). FA showed early patchy hyperfluorescence with late staining of the lesion (Fig. 1B). Ultrasound B scans and computed tomographic (CT) scan findings confirmed the diagnosis of choroidal osteoma. OCT (Stratus; Carl Zeiss Meditec, Dublin, CA) showed subretinal fluid over the osteoma (Fig. 1C). The overlying retinal layers were intact. Here, the osteoma signified calcification. For the next 2 years patient was asymptomatic but the osteoma increased in size to around 5 disk diameter. In March 2009, the patient complained of distortion of vision. BCVA of left eye was 20/30. A crescentic area of subretinal hemorrhage was noted near the macula within the lesion (Fig. 2A). It corresponded as blocked fluorescence in FA (Fig. 2B). The hyperfluorescence underneath the blocked fluorescence represents leaky CNVM (Fig. 2C). Stratus OCT was done which revealed hyporeflective area within retina suggesting edema with fusiform hyper-reflective area within the chorioretinal complex, suggesting an active CNVM. Moreover, the choroidal surface was irregular with bright transmission of light through the atrophic choroidal osteoma. The outer retinal layers including the photoreceptors were disrupted over this part of osteoma. This finding signified decalcification (Fig. 2D). Patient was treated with intravitreal bevacizumab 1.25 mg/0.05 cc. Next month follow-up showed resolution of the hemorrhage with an improved BCVA of 20/20.

Patient had an uneventful follow-up until March 2011 when he again complained of diminution of vision. His
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BCVA in OS was 20/30. Clinical examination revealed elevation in the macular area but no sign of subretinal hemorrhage. At this stage we had access to spectral domain OCT (Spectralis: Heidelberg Engineering, Germany) and the patient was scanned using the same. There was hyperreflectivity of choroidal osteoma with thinning of outer retina temporal to fovea. There were some RPE alterations noted in this area (Fig. 3). Foveal thickness had increased along with presence of subretinal fluid. Patient was treated again with Intravitreal bevacizumab. The subretinal fluid reduced on next month follow-up. We advised further two injections of bevacizumab but the patient was reluctant to take them and wanted to follow-up and take injections in case of any symptomatic worsening. In September 2011 we repeated the OCT examination which showed that the outer layers of retina overlying osteoma were clearly defined which represented calcification (Fig. 4).

Figs 1A to C: Fundus photograph, FFA of choroidal osteoma, OCT showing subretinal fluid

Figs 2A to D: Active CNVM shown in fundus photograph and FFA OCT showing irregular surface of the choroid with bright transmission of light through the atrophic decalcified choroidal osteoma.
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Fig. 3: OCT showing subretinal fluid with deposits between the RPE and sensory retina above the osteoma

Fig. 4: OCT showing calcified hyper-reflective osteoma with overlying intact outer retinal layers

Fig. 5: OCT showing thickened decalcified hyper-reflective osteoma with overlying disruption of outer retinal layer

He came a month after with symptoms of metamorphopsia and on doing an OCT examination we found cystoid spaces in the macular area and the outer layers over the hyper-reflective part of the osteoma were not clearly defined. This part of the osteoma represented decalcification (Fig. 5). Intravitreal injection of bevacizumab was repeated at this stage. The foveal thickness decreased following injection.

In January 2012 OCT there was an increased foveal thickness of 403 microns with cystoid spaces within it. There was also an optically empty space noted temporal to the fovea (Fig. 6). Intravitreal bevacizumab was repeated at this stage. In February, foveal thickness reduced to 381 microns and there were reduction of cystoid spaces (Fig. 7). BCVA improved to 20/30. Eventually the patient has been further followed up on a monthly basis and the condition remains stable till the most recent examination done in September 2012. At time of submitting this report he has been treatment free and stable for 7 months.

DISCUSSION

The etiology of choroidal osteoma remains unclear. Hence, the management of this tumor is essentially limited to baseline documentation and periodic follow-up ocular examination. Up to 33% of eyes with choroidal osteoma may develop CNVM.1 Subretinal neovascularization in such cases has been treated in the past with photodynamic therapy or intravitreal antivascular endothelial growth factor (anti-VEGF) injection.7,8 In our case there was an active CNVM on the 4th year follow-up in 2009. Patient was periodically treated with intravitreal injections of bevacizumab.

We noticed a gradual increase in size of the osteoma throughout our 7 years follow-up. This was also seen through photographic documentation in a mean 10 years of G William Aylward et al study at the Bascom Palmer Eye Institute.9 Treatment of CNVM in their group was done with argon laser photocoagulation. In our case study Stratus OCT revealed optical shadowing of the choroid with overlying retina mostly intact representing calcification. The irregular hyper-reflective surface with bright distorted photoreceptor represented decalcification of the osteoma. This was also found in Shields et al study.10 Their study also noted a poor visual acuity of a calcified subfoveal osteoma compared to a good visual acuity in a calcified subfoveal osteoma. In our case visual acuity was good as the fovea was spared of decalcification as per documentation on the OCT.9 Aurélien Freton et al noted isoreflectivity of osteoma in SD OCT in 7 cases, hyper-reflectivity in two and hyporeflectivity in two of their 11 cases.11 In our case we noted hyper-reflectivity of the osteoma with scattered deposits all over the space between RPE and sensory retina.

Through the FA, Stratus and SD OCT, we documented the course of the disease treated with bevacizumab. We also have a serial documentation of the calcification as well as the decalcification part of the osteoma in a 7-year time. The SD OCT was able to show the peculiarity of the decalcification with its effect on the outer retinal layers. To the best of our knowledge there is no existing report of such a long-term follow-up along with treatment with intravitreal bevacizumab of choroidal osteoma.
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Fig. 6: OCT showing multiple cystoid spaces above the osteoma

Fig. 7: OCT showing reduction of the size of cystoid spaces after the injection

REFERENCES


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