Nonconforming Deep Focal Choroidal Excavation in a Patient With Choroidal Osteoma: A Diagnostic Dilemma

Rohan Chawla, MD, FRCS; Shorya Vardhan Azad, MS; Brijesh Takkar, MD; Anu Sharma, MCOptom; Bibhuti Kashyap, MD

ABSTRACT: Evolution of an osteoma may result in neurosensory detachment, deossification, and choroidal neovascularization (CNV). The authors report a rare case of choroidal osteoma with CNV associated with a deep non-conforming focal choroidal excavation.


INTRODUCTION
Choroidal osteoma (CO) is a benign ossifying tumor commonly occurring in young females.1 Due to its peripapillary/macular location, vision inevitably gets affected during the course of tumor evolution.2 The fate of CO may be variable. It can undergo decalcification leading to loss of overlying choriocapillaries with atrophy of the outer retina, or it may lead to chronic retinal pigment epithelium (RPE) decompensation leading to a persistent neurosensory detachment (NSD).3 The most common cause of vision loss is due to choroidal neovascularization (CNV).3 Also, as these findings may coexist together, it may blur the lines of our management.

Herein we discuss a case of CO with a CNV along with a deep focal choroidal excavation on optical coherence tomography (OCT).

CASE REPORT
A 25-year-old male presented with complaints of sudden-onset, painless diminution of vision in the right eye (OD) with metamorphopsia for 5 days. The patient gave no complaints of field loss, floaters, or flashes. Best-corrected visual acuity (VA) was 20/60 and 20/20 OD and in the left eye (OS), respectively. On examination, both eye anterior segments were within normal limits. Fundus examination OS was unremarkable. OD fundus examination revealed a focal yellowish-orange peripapillary plaque-like lesion with overlying areas of depigmentation and retinal atrophy. A choroidal neovascular membrane (CNVM) with subretinal hemorrhage was seen adjacent to the fovea (Figure 1A). Ultrasonography OD showed an elevated lesion with high acoustic reflectivity and shadowing (Figure 1B). Hence, a diagnosis of OD CO with CNVM was made. Investigations inclusive of fluorescein angiography (FA), OCT, and swept-source OCT angiography (SS-OCTA) were done. FA revealed a patchy hyperfluorescent choroidal filling with masked fluorescence due to blood near the fovea (Figure 2A). On autofluorescence (AF) imaging OD, a peripapillary area of hyperfluorescence with involvement of fovea was seen. This area corresponded to the decalcified portion of the osteoma whereas edges of osteoma showed hypofluorescence signifying calcification of the tumor in the peripheral area (Figure 2A). A lattice-like reflective
pattern at the level of choroid with area of overly-
ing retinal atrophy was seen on OCT OD (Figure 2B). Subretinal CNVM with minimal subretinal fluid was also present (Figure 2C). Peculiarly, an OCT line scan through fovea from superonasal to the inferotemporal macula showed a focal deeply excavated choroid superonasal to the fovea (Figure 3A). Although, the neurosensory retina did not dip into this depression, the RPE lined its entire course. However, unlike a neurosensory detachment, the overlying neurosensory retina was not lifted above this hyporeflective space; rather, the neurosensory retina was in its place, the RPE seemed to dip down into the choroid forming a deep trench, and the underlying choroid was thinned. This excavation was noted far from the CNVM and at the superior edge of the tumor and was not seen in any other scans through the other edges of the osteoma. There was no associated staphyloma of the sclera. OCT OD from the fovea showed an altered pattern of the choroidal vasculature in the area of the osteoma. It additionally unveiled a thicker lacey network of vessels at the level of choriocapillaries corresponding to the CNVM.

Due to an active neovascular membrane, the patient was advised to treat the right eye with the intravitreal anti-vascular endothelial growth factor, bevacizumab (Avastin; Genentech, South San Francisco) 1.25 mg/0.05 mL, subsequent to which there was improvement in vision (20/30) and a decrease in retinal thickness on OCT at 1 month follow-up. However, there was no difference in the depth of excavation, signaling it to be a preexisting structural abnormality, likely unrelated to the CNVM (Figure 3B). OCTA was helpful in assessing response to treatment, as there was a definitive decrease in the caliber of vessels suggestive of a CNVM. The vessels in the post-bevacizumab scan seemed to resemble the adjoining altered choroidal vasculature much more than those in the pre-injection OCTA scan. Patient was kept under observation and advised to follow-up. The patient was found to be stable at 6 months follow-up with VA of 20/30 and no recurrence of the CNVM. The choroidal excavation remained stable during this period.

**DISCUSSION**

Nearly one-third of patients with CO are complicated by a CNVM. It occurs secondary to decalcification along with disruption of the RPE and Bruch’s membrane complex, allowing growth of new vessels. CNVM may present with hemorrhage, subretinal fluid and NSD, requiring some form of treatment. Although many modes of treatment have been tried such as laser, photodynamic therapy (PDT), and anti-vascular endothelial growth factor (VEGF) injections, the latter gives better results for similar CNVMs. Both laser and PDT have also been shown to cause retinal atrophy in the long term and are, therefore, not preferred.

The OCT of our case revealed a deep, trench-like hyporeflective space in the superior part of the osteoma, which had gone unnoticed on first examination. We feel it is unlikely to be due to the active CNVM due to its distant location from the possible source, and the fluid collection would
Figure 2. (A) Fluorescein angiography revealed a patchy hyperfluorescent choroidal filling and staining (blue arrow) with an area of masked fluorescence corresponding to the bleed near the fovea (white arrow). (B) Autofluorescence showing peripapillary area of hyperfluorescence with involvement of fovea (white arrow) (decalcified area), whereas edges of osteoma were hypofluorescent (blue arrow) (calcified area). (C) Optical coherence tomography noted a lattice-work reflective pattern at the level of choroid with an area of overlying retinal atrophy (white arrow). Subretinal choroidal neovascular membrane with minimal subretinal fluid was also present (blue arrow).

Figure 3. (A) Pre-injection optical coherence tomography (OCT) showing focal deeply excavated choroid adjoining the osteoma edge with neurosensory retina not dipping in this depression. There was no associated staphyloma of the sclera. (B) Post-injection OCT showed no difference in the depth of excavation, signaling it to be a preexisting structural abnormality.

Figure 4. (A) Optical coherence tomography angiography (OCTA) additionally unveiled a superficial lacy network of vessels at the level of outer retina (white circle). (B) OCTA is helpful in assessing response to treatment, showing a definitive decrease in the size of the choroidal neovascular membrane (white circle) as vessels post-injection seemed to resemble the adjoining choroidal vasculature much more than those prior to injection.
typically be expected inferiorly rather than superiorly. Also, there was no change in the height or rather the depth of this hyporeflective region following treatment with anti-VEGF, therefore ruling out the possibility of it occurring due to leakage from CNVM. A similar case reported recently, discussed NSD secondary to CNVM, which significantly decreased following anti-VEGF treatment.\textsuperscript{10} It could have been a NSD following decompensation of RPE, which is expected to occur at the same time during decalcification.\textsuperscript{11} Decalcification at the edges was highlighted by the AF images. However, its appearance is unlike a typical NSD, as the overlying neurosensory retina does not seem to be elevated; rather, the RPE seems to be dipping into a thinned-out choroid. Decalcification of CO itself may result in loss of choroidal tissue, but in such a scenario, it leads to the approximation of RPE to the sclera rather than a depression or an excavation, as the process is gradual.\textsuperscript{12} It could also be argued that the identified defect might rather be a failure of the RPE-retina complex to remain continuous with each other due to the anatomical change in their vicinity caused by the presence of the osteoma whose abrupt edge may have caused a mechanical disturbance similar to a posterior staphyloma resulting with RPE and retina incapable of coapting. But in such a case, the trench would be present all around the circumference of the osteoma and not localized to a single edge. Thus, corroborating all the above findings, the only logical alternative abnormality in our differential is a focal nonconforming choroidal excavation.

Focal choroidal excavation (FCE) was first described by Jampol et al. in 2006.\textsuperscript{13} The etiology still remains unclear with some believing it to be a defect in development of the eyeball.\textsuperscript{14} They have been described as two types, conforming and non-conforming.\textsuperscript{14} Our case was the latter type because the NSD did not conform to the depression along with the RPE, as remains the case in conforming type. Initially thought to be a benign developmental abnormality with little clinical implication, current literature has shown FCE to be associated with central serous retinopathy and CNVMs.\textsuperscript{15,16} Recently, a single report has affiliated it with CO, where the authors noted a conforming type of lesions, unlike ours.\textsuperscript{17} Nonconforming type of excavation can be misdiagnosed as an NSD due to an active CNVM or the atrophic RPE. However, the location, architecture and its nonresponsive nature to anti-VEGF is suggestive of this large, trench-like cavity being a FCE. Whether this is a coincidental finding or related in some way to the CO is difficult to comment. However, it did not seem to have any pathological effect on the CO or the CNVM or response to therapy per se.

Nonconforming type of FCE along with a CO may present as a diagnostic dilemma. Its location, morphology and unresponsiveness to anti-VEGF may help in differentiating it from a NSD.

REFERENCES