Case Report

Bilateral Choroidal Osteoma with Secondary Choroidal Neovascularization in a Teenager: A Rare Case

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ABSTRACT

Choroidal osteoma is a rare, benign ossifying tumor that may lead to vision loss due to complications such as choroidal neovascularization (CNV). While typically unilateral in young females, bilateral involvement is less common. This report presents a 16-year-old girl with gradual, painless bilateral vision loss. Her best-corrected visual acuity (BCVA) was 6/60 in the right eye and 6/36 in the left. Fundus examination revealed bilateral, orange, scalloped choroidal lesions in the macula with surface vascular networks. Fluorescein angiography confirmed CNV in both eyes. The patient received three intravitreal injections of ranibizumab (0.3 mg/0.05 ml) in each eye at four-week intervals. Post-treatment, her BCVA improved to 6/12 in the right eye and 6/9 in the left, with resolution of subretinal fluid. This improvement was sustained over a 12-month follow-up. The case highlights the importance of imaging in diagnosing choroidal osteoma and demonstrates the efficacy of anti-VEGF therapy in managing associated CNV.

Keywords: Choroidal Osteoma, Choroidal Neovascularization, Ranibizumab, Retinal detachment.

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INTRODUCTION

Choroidal osteoma is a benign tumor that is uncommon and is distinguished by the development of mature cancellous bone within the choroid.[1] The exact etiology remains unclear, and no specific risk factors have been identified. Gass et al, first described it in 1978.[1] It is most commonly a unilateral condition that primarily affects young females in their second or third decade of life and is frequently observed in the vicinity of the optic disc.[2] However, in rare cases (approximately 25%) bilateral involvement has been reported.

On fundoscopic examination, these tumors appear as distinct orange yellow to yellow-white lesions with visible blood vessels on their surface.[1] Their color changes over time, with early-stage lesions exhibiting an orange-red hue, while later stages develop a yellowish tint due to retinal pigment epithelium (RPE) atrophy. Though primarily diagnosed through clinical assessment, imaging techniques such as A-scan and B-scan ultrasonography assist in confirmation, revealing high reflectivity and acoustic shadowing.[3] While CT scans and X-rays can further verify the bony nature of the tumor, these imaging modalities are rarely used in modern practice.[3]

The primary causes of vision impairment in choroidal osteoma include choroidal neovascularization (CNV), photoreceptor damage, and RPE atrophy, which are often linked to tumor decalcification.[1,4] CNV frequently arises in tumors with an irregular surface and subretinal hemorrhage, potentially leading to complications such as serous retinal detachment.[5,6] Decalcification, first reported by Trimble in 1988, is observed in nearly 50% of cases and significantly worsens visual function when it involves the foveal region due to photoreceptor loss.[7,8]

Management of CNV secondary to choroidal

osteoma remains an evolving field, particularly in pediatric cases, where the long-term effectiveness of anti-VEGF therapy is still being explored.[2,5] Limited research has been conducted on the sustained benefits of antiangiogenic agents in this condition. This case report details a teenage girl diagnosed with bilateral choroidal osteoma complicated by CNV and highlights the role of intravitreal anti-VEGF injections as a potential treatment strategy.

Case Presentation

The patient provided informed written consent for the publication of her medical information, and Ethical Review Board (ERB) approval was obtained by institution. This case is reported in accordance with the Declaration of Helsinki and its subsequent revisions. A 16-year-old girl presented with gradual, painless vision loss in both eyes over the past few months. She had no prior medical or ocular history of significance. On examination, her best-corrected visual acuity (BCVA) was recorded as 6/60 in the right eye and 6/36 in the left eye. The anterior segment was normal, with no evidence of anterior or intermediate uveitis. Fundus examination revealed bilateral, slightly elevated, irregularly bordered orange choroidal lesions, predominantly in the macular region. These lesions had scalloped margins and multiple surface vascular networks. Additionally, yellow-gray sub-foveal changes were observed in both eyes (Figure 1).

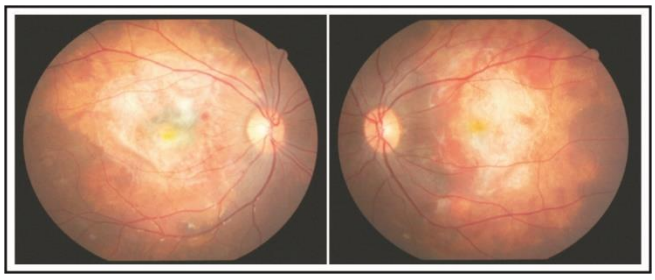

Figure 1: Fundus picture of a patient with choroidal osteoma.

Red-free fundus imaging highlighted the lesion's distinct margins, areas of increased reflectivity due to RPE atrophy and prominent surface vascularization in CNV-affected regions (Figure 2 Top images). Fluorescein angiography (FFA) revealed early patchy hyperfluorescence with late leakage and staining, indicative of choroidal neovascularization CNV in both eyes. (Figure 2, Bottom images).

Ultrasound A-scan demonstrated a high-intensity echo spike characteristic of calcified nature of lesion with no internal vascular pulsations or low-reflectivity zones differentiating it from choroidal melanoma or other soft tissue tumors. B-scan showed highly reflective choroidal mass in both eyes, with posterior acoustic shadowing giving a characteristic "pseudo-optic nerve" appearance. (Figure 3).

OCT confirmed the presence of CNV with subretinal fluid accumulation. Neo-vessels can be seen in OCT-angiography (Figure 4).

Three intravitreal injections of Ranibizumab (Patizra) at a dose of 0.3 mg/0.05 ml were administered to the patient in both eyes at four-week intervals. Following treatment, her best corrected visual acuity improved to 6/12 in the right eye and 6/9 in the left eye, with resolution of subretinal fluid (Figure 5). This visual improvement was maintained over a 12-month follow-up period.

DISCUSSION

Choroidal osteoma is an uncommon benign intraocular tumor characterized by cancellous bone deposition within the choroid and primarily affects young females.[1] While the tumor is typically unilateral, bilateral involvement has also been noted particularly infemales.[2,6,7,9,10]Multifocality of choroidal osteoma is also a relatively uncommon condition.[2]

The exact pathogenesis of choroidal osteoma remains uncertain, but hypotheses suggest potential associations with intraocular inflammation, pregnancy, trauma, hereditary factors, and osseous choristoma.1 Shields et al, reported that tumor growth is often accompanied by decalcification, leading to complications such as CNV, sub macular hemorrhage, and progressive retinal atrophy.[8]

CNV is one of the most serious complications, occurring in 31%–47% of cases within 10 years and in up to 56% over 20 years, significantly worsening visual prognosis.[2,8] Decalcification, which affects nearly half of all cases, is particularly concerning due to its strong association with RPE atrophy, choriocapillaris degeneration, and photoreceptor loss.[8]

Histopathological studies have demonstrated that decalcified tumor regions exhibit severe thinning or complete loss of the outer retina and photoreceptor layers, while calcified areas tend to maintain retinal integrity.[8]These results indicate the significance of early intervention in the preservation of visual

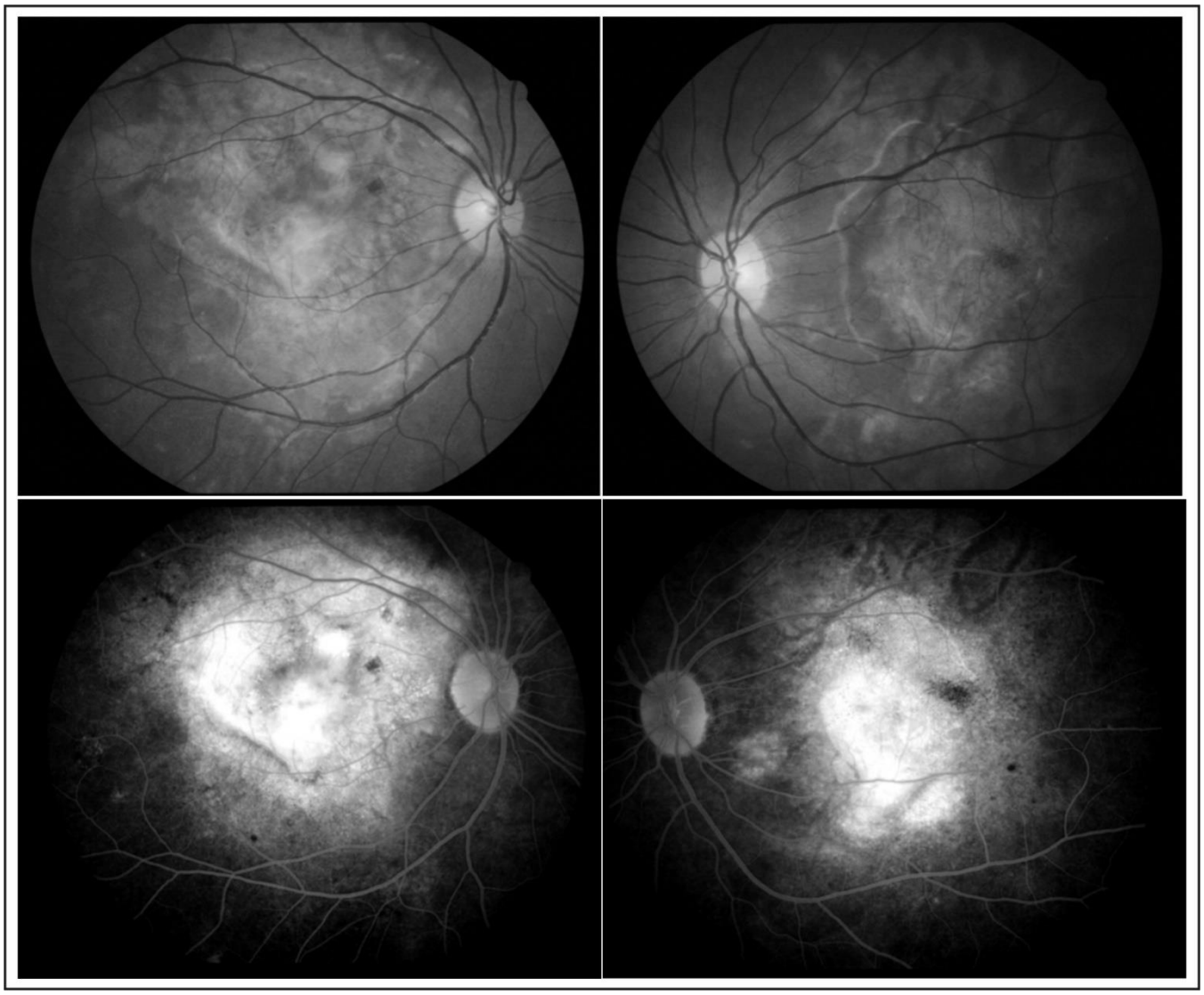

Figure 2: Top, Red-free images showing macular lesions with distinct margins in both eyes. Bottom, Late frames of FFA showing staining of the lesions and leakage.

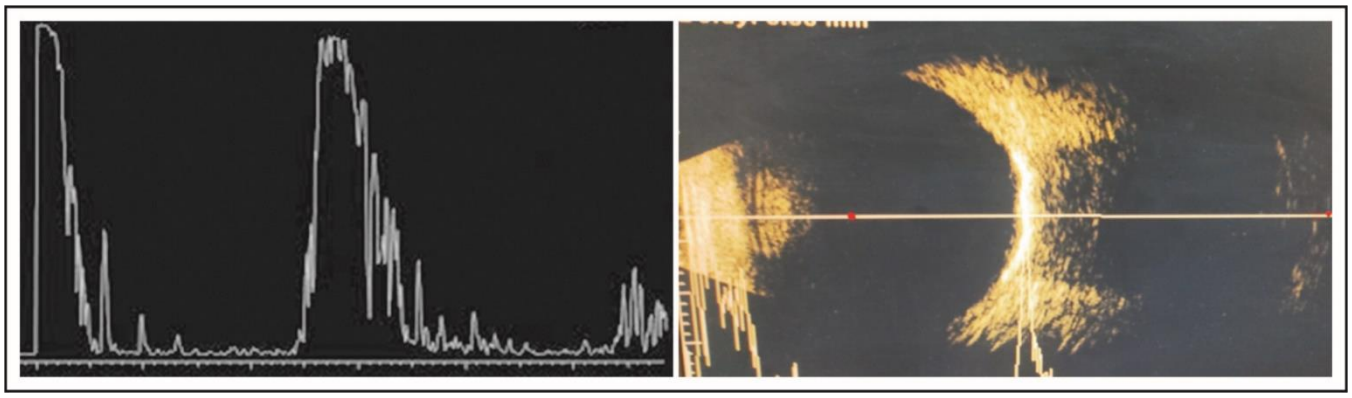

Figure 3: High spikes of the choroidal osteoma in A-scan and Orbital shadowing in B-scan.

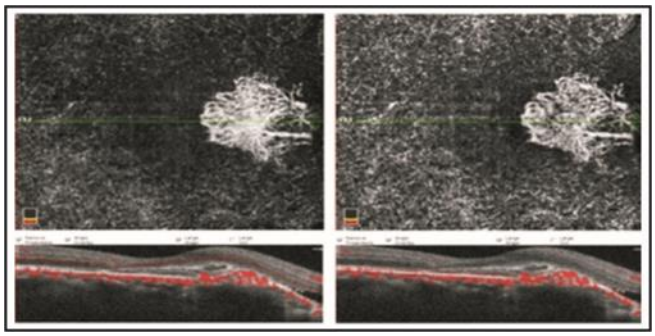

Figure 4: OCTA depicting abnormal vessels.

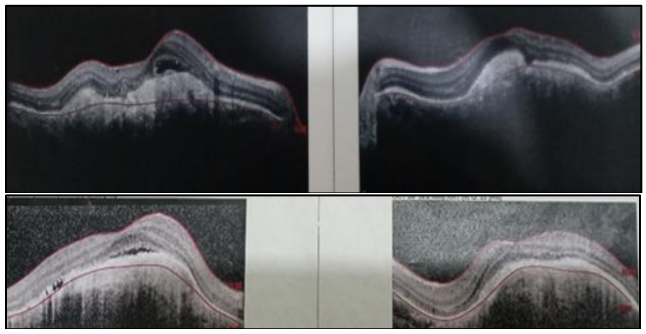

Figure 5: Top, OCT showing active CNV. Bottom, OCT of the patient after four intravitreal Ranibizumab injections.

function. Management of CNV secondary to choroidal osteoma remains a clinical challenge. Traditional treatments such as laser photocoagulation, surgical CNV excision, transpupillary thermotherapy (TTT), and photodynamic therapy (PDT) have shown limited efficacy, with some procedures causing further visual deterioration due to RPE damage and tumor decalcification.[2,4,5] Shields et al, recommended PDT for extrafoveal CNV, though its use for sub foveal CNV remains controversial due to potential exacerbation of visual decline.[8]

Recent advancements in anti-VEGF therapy have provided promising results. Gomez et al, reported that bevacizumab effectively reduced subretinal fluid and stabilized vision in patients with CNV secondary to choroidal osteoma.[2,8] Similarly, our patient responded well to three intravitreal injections of Ranibizumab, with significant visual improvement and CNV resolution. These findings further support the role of anti-VEGF agents as an effective treatment modality in such cases.

Although choroidal osteoma typically occurs in isolation, rare associations with other ocular conditions such as optic atrophy, posterior scleritis and polypoidal choroidal vasculopathy (PCV) have been reported.[7,9,10] The coexistence of posterior scleritis and choroidal osteoma is rare, with only three documented cases.[9] While the exact link between these two conditions is unclear, it is crucial to consider posterior scleritis as a potential comorbidity in patients with choroidal osteoma, as chronic inflammation may contribute to tumor progression. Choroidal osteoma-related vision loss is often attributed to CNV, but some cases suggest an association with PCV, particularly when sub macular hemorrhage is present. Unlike CNV, PCV tends to have a better visual prognosis and may respond more favorably to PDT.[5,10] Fine et al, documented a case of bilateral PCV in a patient with choroidal osteoma, highlighting the importance of indocyanine green angiography (ICGA) in distinguishing between CNV and PCV.[10]

CONCLUSION

This case report highlights the rare occurrence of bilateral choroidal osteoma with CNV in a teenage patient, successfully managed with anti-VEGF therapy. While the treatment response was favorable, the long-term prognosis remains uncertain, emphasizing the need for ongoing monitoring and further research on the sustained efficacy of anti-VEGF therapy. Choroidal osteoma can lead to significant vision loss due to decalcification, CNV, and RPE atrophy, making early diagnosis, vigilant follow-up, and timely intervention crucial to preserving vision. Intravitreal Ranibizumab demonstrated effectiveness in improving and stabilizing vision, consistent with existing literature. However, given the potential for disease progression and recurrence, long-term follow-up with multimodal imaging remains essential for optimizing visual outcomes.

Patient's Consent: Researchers followed the guide lines set forth in the Declaration of Helsinki.

Conflict of Interest: Authors declared no conflict of interest.

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