



## Case report

# Intravitreal ranibizumab for treatment of choroidal neovascularization secondary to a bilateral choroidal osteoma



Sidra Zafar <sup>a,\*</sup>, M.A. Burq <sup>b</sup>, Mustafa Ifhtikar <sup>a</sup>, Azam Ali <sup>b</sup>

<sup>a</sup> Aga Khan University Hospital, Karachi, Pakistan

<sup>b</sup> Consultant Ophthalmologist, Section of Ophthalmology, Department of Surgery, Aga Khan University Hospital, Karachi, Pakistan

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## ABSTRACT

**Purpose:** Choroidal osteomas are benign intraocular tumors that classically present in females. Despite their benign nature, significant visual acuity loss can occur due to retinal pigment epithelium degeneration. We report an unusual case of bilateral choroidal osteoma in a young boy.

**Observation:** A 16 year old boy presented to the Aga Khan University Hospital with a history of painless, bilateral deteriorating vision over past few months. Examination showed best corrected visual acuity as 20/200 in the right eye and 20/400 in the left eye. Funduscopy revealed a well-defined lesion in the juxtafoveal region of both eyes. A diagnosis of (bilateral) choroidal osteoma was subsequently made on the basis of optical coherence tomography, fundus fluorescein angiography, Indocyanine green and B-scan ultrasonography. The presence of choroidal neovascularization with sub retinal fluid was also established on OCT. The patient was treated with three intravitreal Ranizumab injections. At the follow up visit, vision had improved to 20/50 in the right eye and 20/200 in the left. Sub retinal fluid had also resolved.

**Conclusions:** and importance: Intravitreal Ranizumab may be an effective alternative in the management of choroidal neovascularization secondary to choroidal osteoma.

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## 1. Introduction

Choroidal osteomas are rare, benign intraocular tumors, first described in 1979 by Gass et al.<sup>1</sup> They are characterised by the presence of mature, cancellous bone within the choroid and are classically found as unilateral lesions in females in their second or third decade of life. On fundus examination, choroidal osteomas appear as slightly and irregularly elevated, orange-yellow plaques deep to the retina in the juxta papillary or macular region with well-defined geographic borders.<sup>1–3</sup> The pathogenesis and etiology of the tumor however, is still not well understood.

Despite being benign tumors, choroidal osteomas can often cause severe loss of visual acuity secondary to gradual atrophy of the overlying retina pigment epithelium (RPE) or by accumulation of either sub retinal fluid or sub retinal hemorrhage with or without choroidal neovascularization (CNV).<sup>4</sup> Treatment options have included laser photocoagulation, excision of CNV, photodynamic

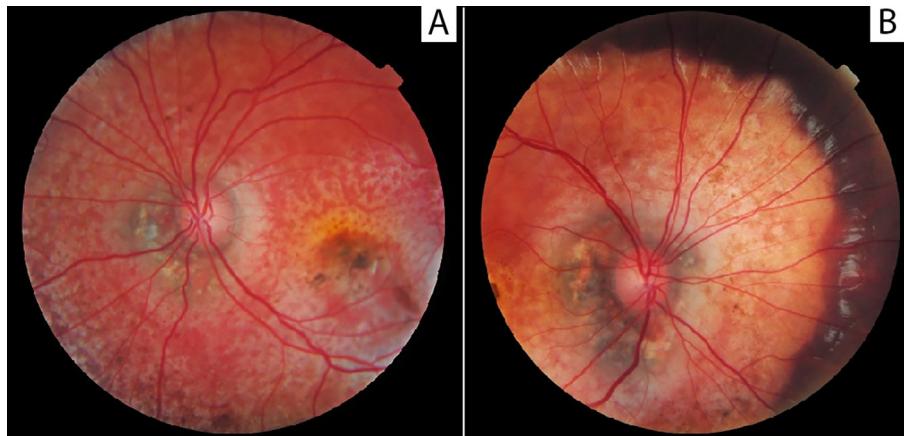
therapy (PDT) and trans pupillary thermal therapy (TTT) and more recently, the use of intravitreal anti-vascular endothelial growth factor (VEGF).

We evaluated the role of anti-VEGF therapy in the treatment of CNV in choroidal osteomas after their use in some case reports.

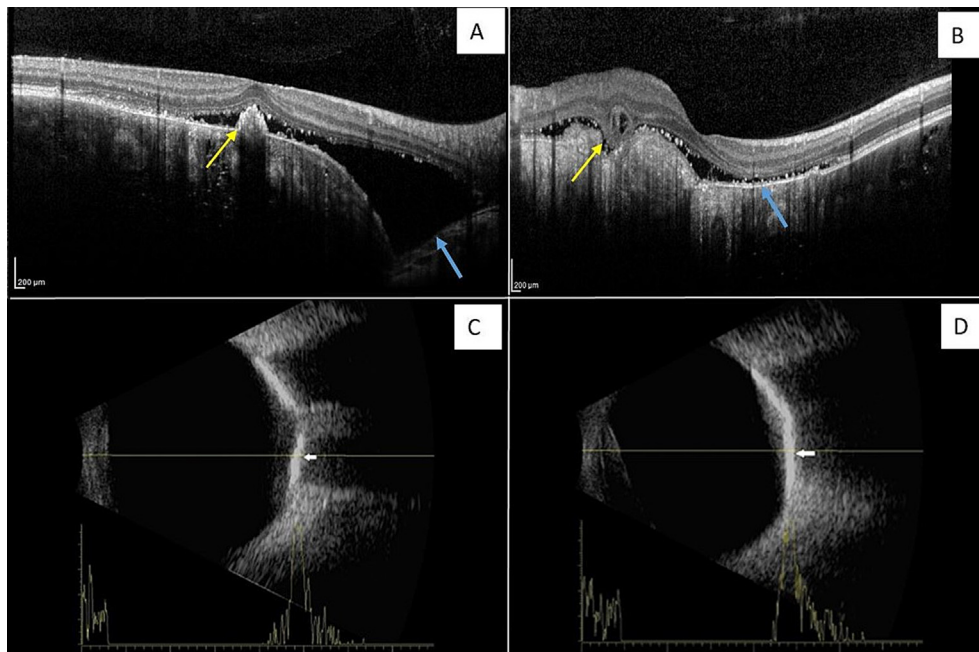
## 2. Case report

A healthy 16 years old boy presented to the ophthalmology consulting clinics, Aga Khan University Hospital with bilateral deteriorating vision over the past few months. Best corrected visual acuity (BCVA) was 20/200 in the right eye and 20/400 in the left eye. Past ocular history was significant for receiving 7 bevacizumab injections (Avastin; Genentech, Inc., South San Francisco, CA) for a choroidal hemangioma. Rest of the history was unremarkable. On clinical examination, an orange-yellow plaque was noted in the juxtafoveal region of both eyes with associated calcification (Fig. 1A and B). The osteoma gave the characteristic appearance of a pseudo-optic nerve on B-scan ultrasonography, appearing as a slightly elevated, highly reflective choroidal mass (Fig. 2C and D). Fundus fluorescein angiography (FFA) showed early, patchy hyper

\* Corresponding author. B-123, Block-H, North Nazimabad, Karachi, Pakistan.  
E-mail address: [sidrazafariqbal@gmail.com](mailto:sidrazafariqbal@gmail.com) (S. Zafar).



**Fig. 1.** (A) A yellowish-choroidal lesion is noted in the left eye with well-defined borders. (B) A similar lesion can also be appreciated in the right eye. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)



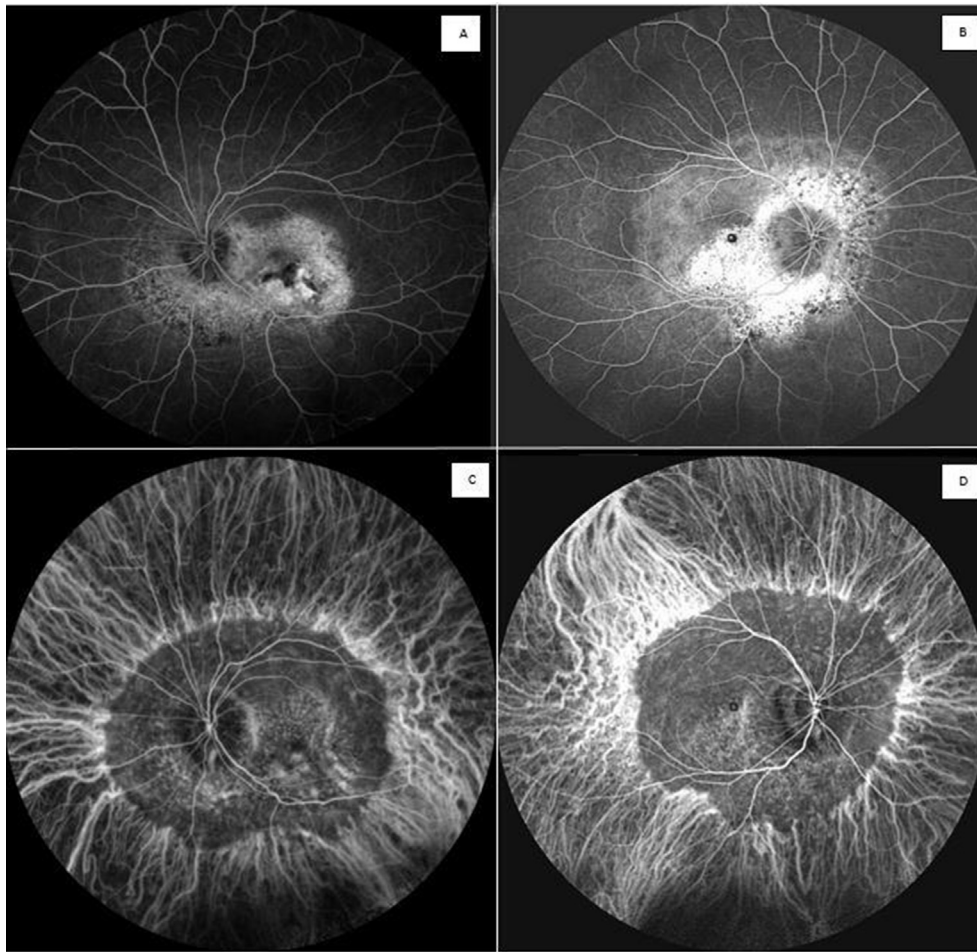
**Fig. 2.** (A) and (B) Optical coherence tomography scan of the right eye and left eye, respectively. The choroidal neovascular membrane can be visualized on scans of both the eyes highlighted using the *yellow* arrow. The *blue* arrow demonstrates presence of sub retinal fluid. (C) and (D) B-scan ultrasonography appears as slightly elevated, highly reflective choroidal mass with acoustic shadowing bilaterally. The *white* arrow highlights the characteristic pseudo-optic nerve finding associated with choroidal osteomas. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

fluorescence of the choroidal lesion with late diffuse staining in areas of calcification (Fig. 3A and B). On optical coherence tomography (OCT), an irregular hyper reflective area was observed in the choroid with sub retinal fluid overlying the osteoma (Fig. 2A and B). Indocyanine green showed areas of hypofluorescence in the early phase as well as hyperfluorescent in late frames (Fig. 3C and D). A diagnosis of choroidal osteomas was made for both eyes with associated choroidal neovascularization (CNV) and sub retinal fluid. A decision was made to treat the patient with intravitreal ranibizumab ([0.5 mg] Lucentis; Genentech, Inc., South San Francisco, CA). The potential risks and benefits of the intravitreal injection of ranibizumab were explained to the patient and written consent was obtained. A total of three injections were administered, each separated by a month's duration. At the 2-week follow up, visual

acuity had improved to 20/50 in the right eye and 20/200 in the left. The sub retinal fluid had also resolved.

### 3. Discussion

Choroidal osteomas are rare, ossifying tumors that classically occur as unilateral lesions affecting middle-aged women.<sup>2</sup> In two of the largest long-term studies involving choroidal osteomas, analysis of 36 and 74 patients over a period of 10 and 26 years respectively, revealed bilateral lesions in only 25% of the cases with 67%–89% affected being female.<sup>2,3</sup> Our case is therefore unique as it presented as a bilateral lesion in a young male, which is an uncommon finding.<sup>5</sup>



**Fig. 3.** (A) and (B) Fundus Fluorescein Angiography with early patchy hyperfluorescence and late diffuse staining in areas of calcification. 3(C) and 3(D) demonstrate hypo fluorescence of mass on Indocyanine green with diffuse hyper fluorescence in late frames.

Loss of visual acuity in choroidal osteomas is most commonly attributed to decalcification occurring from disruption of the tumor. Subsequently, the insult may result in damage to the RPE, CNV, and appearance of sub retinal fluid and/or hemorrhages.

Management of CNV associated with choroidal osteomas is a challenging issue. Various treatments have included laser photocoagulation, surgical removal of CNV and TTT but have achieved limited success. Laser photocoagulation of CNV associated with choroidal osteoma has largely been unsuccessful. In a long-term study conducted by Aylward et al. successful treatment was achieved in only 25% of the eyes. It is hypothesized that, owing to depigmentation of the RPE, there is decreased absorption of laser energy and hence, reduced treatment efficacy.<sup>3</sup> Surgical removal of CNV and TTT have also been tried but with poor visual outcomes.<sup>6–8</sup> More recent treatments have included photodynamic therapy (PDT) and intravitreal bevacizumab (Avastin) or ranibizumab (Lucentis).

Anti-VEGF has successfully been used in the treatment of CNV associated with different ocular diseases including age-related macular degeneration (ARMD) and diabetic retinopathy. Various studies have also reported efficacy of anti-VEGF in inducing remission of CNV secondary to choroidal osteomas.<sup>4,7,9–11</sup> Use of PDT has also shown to have multiple benefits including improved visual acuity and metamorphopsia as well as a reduction in the size of the CNV. However, relative to anti-VEGF, PDT is associated with poor visual outcome and the possible need for multiple re-

treatments.<sup>10</sup> Focal decalcification of the tumor may also occur following treatment which may again be visually detrimental.

After assessing all of the possible treatment options, we successfully treated our patient with intravitreal ranibizumab. Ranibizumab reduces permeability of choroidal vessels through its influence on VEGF levels resulting in a significant decrease in sub retinal fluid, as well as improved visual acuity overall.

#### 4. Conclusion

Although choroidal osteomas remain a rare ocular pathology, significant progress has been made in treating choroidal osteoma associated CNV, a significant cause of visual impairment in these patients. Anti-VEGF, the newest therapeutic modality has successfully shown to resolve CNV and/or associated sub retinal fluid and hemorrhages. Furthermore, it is associated with few complications and a good visual outcome.

#### Conflict of interest

The authors declare no conflict of interest.

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