

Long-Term Follow Up of Intra-Retinal Fluid and Outer Retinal Tubulations in a Case of Choroidal Osteoma Following Anti-VEGF Treatment

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Abstract

Purpose: To report long term follow up of intraretinal fluid (IRF) and outer retinal tubulations (ORTs) in a case of choroidal osteoma (CO) over 54 month.

Method: Case report.

Results: A 35 Saudi female presented because of poor vision in her left eye. Her right eye was essentially normal with 20/20 vision (VA). Her left eye had 1/200 VA and showed peripapillary CO involving the macular area. OCT imaging showed intraretinal cystic spaces that were interpreted as a sign of an active choroidal neovascular membrane (CNV). 5 Avastin injections were administered into her left eye with no anatomical or visual improvement. On reviewing her imaging, no CNV was detected at any point. The IRF in this case seems related to RPE damage overlying the CO. ORTs were detected and increased over time.

Conclusion: Clinicians should be careful when assessing cystic spaces in cases of CO. ORTs seem to represent a healing mechanism. IRF and ORTs in CO cases do not seem to benefit from anti-VEGF injections.

Keywords: Choroidal osteoma; Intra-retinal fluid; Outer retinal tubulations; Intravitreal bevacizumab; Choroidal neovascular membrane; Fundus autofluorescence

Introduction

Choroidal Osteoma (CO) is a rare ossifying choriostoma of the choroid that particularly affects young women in the childbearing age. Gass first described CO in 1978 [1]. It is usually appears as a well-defined, slightly elevated, yellow-white fundus lesion at the peripapillary or macular area [2]. Visual loss in cases of CO can happen due to atrophy of the overlying retina, accumulation of SubRetinal Fluid (SRF) or Choroidal NeoVascularization (CNV) [3]. Cases of CNV secondary to CO are usually treated nowadays with intravitreal injections of anti-VEGF drugs with good response [4]. Anti-VEGF therapy also showed good anatomical and visual results in treating SRF in cases of CO not associated with CNV [5]. We present here long term follow up of a case of CO with IRF and Outer Retinal Tubulations (ORTs) that was thought to have a CNV hence treated with 5 Avastin injections with no anatomical or visual change. We are not aware of long term follow up in similar cases or the results of Avastin use to treat such cases in literature.

Case Report

A 35-year-old Saudi female experienced poor vision in her left eye that started 10 years earlier. She presented to another ophthalmic unit in 2013. At that point of time she was told that she has a CO in her left eye associated a CNV and a course of 3 Avastin injections was given to

her left eye. She did not notice any change in her vision and she was referred to KKESH (King Khaled Eye Specialist Hospital). A year later, she was examined in the retina clinic in KKESH. Her right eye was essentially normal with 20/20 vision. VA in the left (affected) eye was 1/200. The left eye showed a fundal lesion compatible with a CO (Figure 1A). Ultrasound scan was done and that confirmed the diagnosis by showing a high reflective lesion with orbital shadowing behind it (Figure 1B). FFA was done at that point with Topcon fundus camera (Figure 1C) and no active CNV was seen on it. Spectral domain OCT (OCT, Heidelberg Engineering, Heidelberg, Germany) was also done (Figure 1D) showing some cystic spaces that seemed intra-retinal and subretinal. No further injections were recommended at that point. When we reviewed the OCT scans at that point, we could see at least 2 oval ORTs above the level of RPE with hyper reflective wall (Figure 1D) in addition to some IRF mainly at the plane of outer nuclear layer (ONL). 18 months later, during follow up of the case, the fundal appearance was not much different (Figure 2A).

OCT scans showed oval cystic spaces just inner to the RPE plane and in the middle of the retina (Figure 2B). Based on that, a CNV was thought to be active and 2 Avastin injections were given 6 weeks apart with no anatomical or visual change. Then follow up is decided with no further injections. On reviewing the OCT at that point, we could identify more ORTs (Figure 2B) with IRF as well. 2 years later, the patient came under care of the authors. The right eye examination was normal. Left VA at that point was 1/200. The left eye fundal appearance was pretty similar to before. OCT showed some IRF and ORTs. Observation was decided. 6 months later (in October 2017) the imaging was repeated. Figure 3A shows the fundal appearance with some decalcification of the CO at the macular area. OCT

demonstrated more ORTs in addition to IRF (Figure 3B). OCT also demonstrated RPE and photoreceptors disintegration at the nasal part of the macula overlying the CO. Fundus Autofluorescence (AF) as done with Optos fundus camera (Optos 200TX, Dunfermline, UK)

with 488 nm wavelength and that showed reduced AF over the lesion indicating RPE loss (Figure 3C). FFA was repeated to make sure no active CNV is there which was the case (Figure 3D).

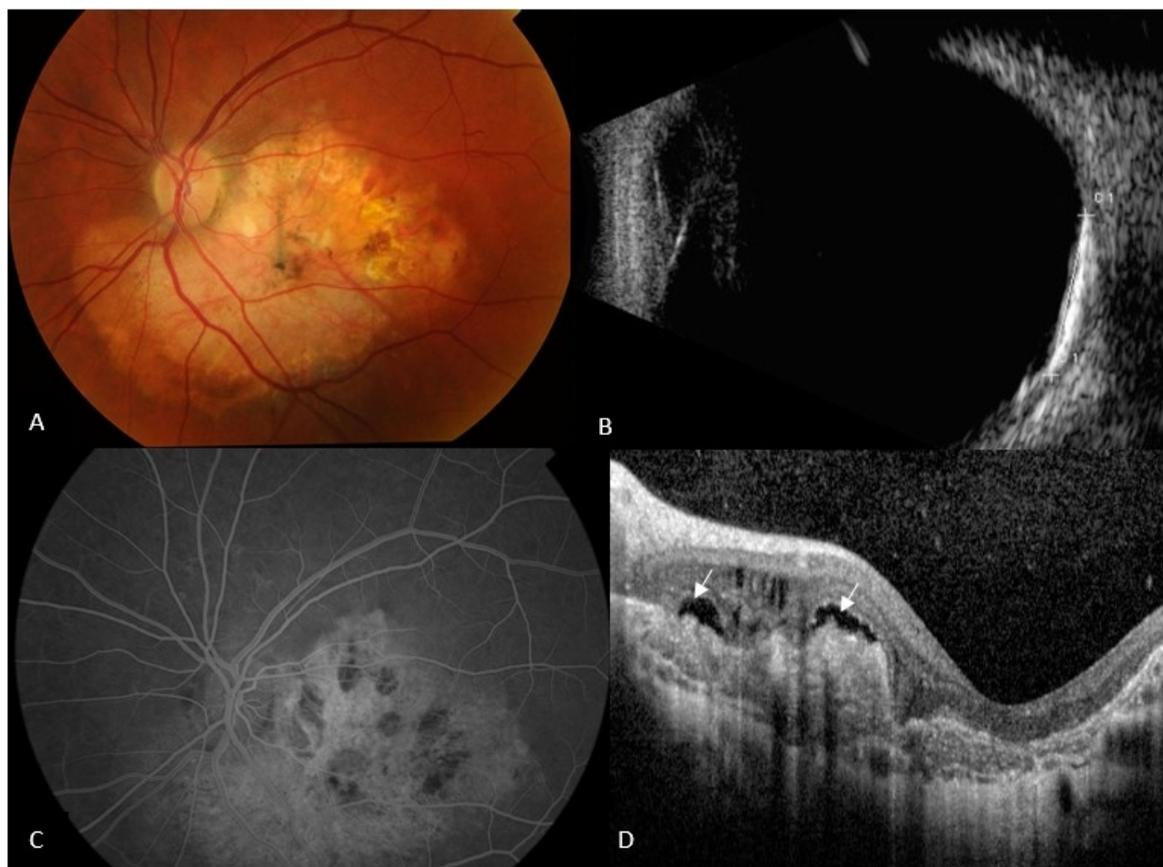


Figure 1: Imaging of a case of choroidal osteoma (CO) with intraretinal fluid (IRF) and outer retinal tubulations (ORTs) at presentation. (A: Color fundus photo of the affected left eye showing a well-defined yellow-white lesion involving the macular and infero-nasal peripapillary areas. B: ultrasound scans of the affected eye showing hyper reflective lesion with shadowing behind it. C: Fluorescein fundus angiography (FFA) of the affected eye (venous phase) showing hyperfluorescence over most of the lesion due to staining with small areas of hypofluorescence due to localized areas of RPE & choriocapillaries atrophy. No evidence of CNV seen on FFA. D: OCT scan (horizontal cut through the foveal center) showing hyperreflective lesion under the RPE at the nasal part of the macula with overlying RPE thinning/loss and loss of ellipsoid zone and external limiting membrane). There is also IRF, mainly in the outer nuclear layer and at least 2 oval cystic spaces above the RPE plane with well-defined hyper-reflective wall compatible with ORTs (marked with arrows).

Discussion

In this case of CO, the VA was poor since presentation because of extension of the lesion to involve the foveal area with decalcification associated with overlying RPE and outer retinal disintegration. VA remained the same (1/200) since April 2014 to date. On reviewing the previous imaging, no CNV could be detected by the authors at any point. However, IRF and ORTs were noted at all points of time. The IRF could be related to the RPE damage at the nasal part of the macula overlying decalcified tumor. This is confirmed by the reduced AF signal over the tumor area. ORTs were recently described by Xuan et al. [6] in 26% of case series of 17 cases of CO. They found that big tumor size; decalcified tumor; IRF; and RPE alterations within the fovea were risk factors for the development of ORTs. All these risk factors are evident in our case. These ORTs are thought to be a healing/remolding

mechanism in the presence of chronic IRF. It is not necessarily associated with a CNV. In Xuan and colleague's series, CNV could be detected in only one out of 5 cases with ORTs. It is to be noted that in our case the number of ORTs seems to increase as time has gone. This, perhaps, supports the theory that ORTs is a healing/remolding mechanism in cases of chronic IRF. Song and colleague [5] reported the efficacy of Avastin in achieving anatomical and visual improvement in cases of SRF secondary to CO without CNV. But this does not apply in our case, as the fluid here was not SRF. This is probably the reason why 5 Avastin injections in our cases did not result in any anatomical or visual improvement. In conclusion, the diagnosis of a CNV in the context of CO should be based on clear angiographic evidence (FFA or OCTA). Care should be taken not to consider ORTs and IRF in cases of CO as being SRF. This could lead to unwarranted anti-VEGF injections that would not benefit the case and could potentially cause

side effects being an invasive procedure.

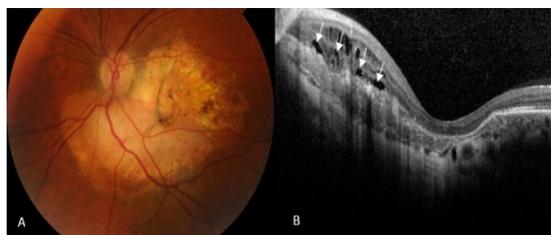


Figure 2: Imaging of a case of choroidal osteoma (CO) with intraretinal fluid (IRF) and outer retinal tubulations (ORTs) 18 month later. (A: Color fundus photo of the left eye showing the CO with no much change compared to the appearance at presentation in figure 1A. B: OCT scan (horizontal cut through the foveal center) showing hyperreflective lesion under the RPE at the nasal part of the macula with overlying loss of RPE, ellipsoid zone, and external limiting membrane). There is also IRF, mainly in the outer nuclear layer and at least 4 cystic spaces above the RPE plane with well-defined hyper-reflective wall compatible with ORTs (marked with arrows).

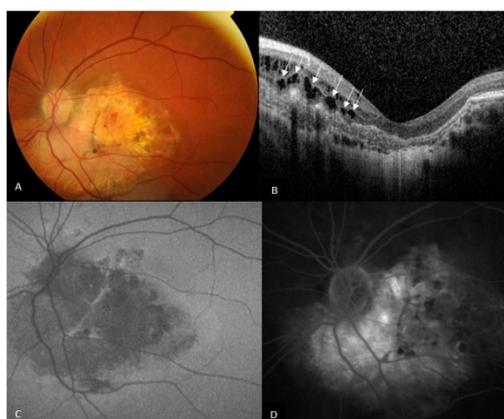


Figure 3: Imaging of a case of choroidal osteoma (CO) with intraretinal fluid (IRF) and outer retinal tubulations (ORTs) at last follow up (54 month after presentation to us). A: Color fundus photo of the left eye showing the CO with no significant change compared to the appearance at presentation in figure 1A. B: OCT scan (horizontal cut through the foveal center) showing hyperreflective lesion under the RPE at the nasal part of the macula with overlying loss of RPE, ellipsoid zone, and external limiting membrane. There is IRF, mainly in the outer nuclear layer and at least 6 cystic spaces above the RPE plane with well-defined hyperreflective wall compatible with ORTs (marked with arrows). C: Optos Fundus autofluorescence (AF) showing reduced signal over the CO (excluding a small streak in the middle corresponding to preserved area of pigmentation on color photos) indicating diffuse RPE loss. D: FFA angiography of the left eye in the late (recirculation) phase showing hyperfluorescence over most of the CO due to staining with small areas of hypofluorescence due to localized areas of atrophy of RPE & underlying choriocapillaries with no evidence of CNV.

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