Multimodal imaging in choroidal osteoma

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Abstract

Purpose: To describe characteristics of choroidal osteomas (CO), using ocular ultrasound, fluorescein angiography, ultra-widefield retinal imaging, ultra-widefield autofluorescence, optical coherence tomography, enhanced-depth-imaging OCT, and OCT angiography (OCT-A).

Methods: Retrospective, observational case series study. Clinical records from patients with diagnosis of CO who underwent complete imaging evaluation were analyzed.

Results: Sixteen eyes from 11 patients were included. Mean patient age was 33.4 years (range 20–61), 72.7% were female, 100% were Hispanic, and 54.5% had unilateral CO. Median visual acuity was 20/150 (range 20/20–2000). CO was completely calcified in 25%, partially decalcified in 50%, and decalcified in 25%. Other features included choroidal neovascularization (18.75%), focal choroidal excavation (12.5%), choroidal depression associated to decalcification (18.75%), thinning of outer retina and photoreceptor layers over decalcified tumor (75%). Decreased fluorescence on FAF was observed in decalcified regions while relatively preserved fluorescence was observed in calcified regions.

Conclusions: Nowadays, diagnostic tests provide important information about each stage of choroidal osteoma. Progressive decalcification of the tumor might have a common pathogenic role for development of FCE or choroidal depression. OCT-A/FA proved to be valuable tools for detection of CNV in patients with CO.

Keywords: Choroidal osteoma, Choroidal excavation, Choroidal neovascularization, EDI-OCT, OCT-A, Ultra-widefield retinal imaging

Introduction

Choroidal osteoma (CO) is a rare benign tumor of the choroid, which is composed of mature bone (trabecular and/or compact) and vascular channels [1–4]. Gass et al. made the first description of this kind of neoplasm in 1978, and since then multiple case reports and series have been published [5].

CO is typically unilateral (80% of cases) and it usually affects young healthy female patients [6]. It arises in late childhood or early adulthood and its most common symptoms are blurred vision, metamorphopsia and presence of a scotoma [7]. The clinical appearance of the tumor may vary from white-cream or yellow-gray to orange, well-defined, which according to some authors corresponds to the grade of calcification (orange pigmentation is present in areas with more ossification) [6].

Over time, ocular ultrasound (US), fluorescein angiography (FA) and optical coherence tomography (OCT) have been widely used for diagnosis and follow-up of CO. Enhanced depth imaging OCT (EDI-OCT) is a recent addition of OCT, that has been able to reveal the presence of bone lamella, tubular lamella with optically empty center, vascular channels and trabecular bone in patients with CO [8, 9].

In patients with CO, OCT angiography (OCT-A) is a new non-invasive imaging technique that employs motion contrast from blood flow to generate high-resolution angiographic images, in patients with CO has been able to show a dense irregular vascular network in the outer retinal layer (ORL) and choroid capillary layers [10]. On the other hand OCT-A [11].

The aim of this study is to describe the morphology of CO using a multimodal image system.
| Case | Age | Gender | Laterality | Localization | Decalcification | BCVA (Snellen) | Ophthalmic conditions | Clinically suspected CNV | Autofluorescence | FA | OCT-A findings (superficial choroid) |
|------|-----|--------|------------|--------------|----------------|----------------|----------------------|---------------------|-----------------|----|-----------------------------------|
|      |     |        |            |              |                | RE | LE | RE | LE | RE | LE | RE | LE | RE | LE | RE | LE | RE | LE |
| 1    | 21  | F      | L          | DNA          | XF             | WNL | C  | 20/150 | 20/200 | Keratocanus | N   | N   | WNL | H-hd | ND | ND | WNL | AV |
| 2    | 20  | M      | B          | SF           | SF             | PC  | PC | 20/800 | 20/200 | None          | Y   | Y   | h-He | h-Hd | WD | WD | DB  | DB |
| 3    | 21  | F      | B          | SF           | SF             | C   | PC | 20/20  | 20/20  | None          | Y   | Y   | H-Hd | H-he | CNV| WD | CNV | VN |
| 4    | 27  | F      | R          | XF           | DNA            | C   | WNL | 20/20  | 20/50  | Keratocanus | N   | N   | WNL | ND  | ND | ND | ND  | ND |
| 5    | 30  | F      | R          | SF           | DNA            | PC  | WNL | 20/50  | 20/25  | None          | N   | N   | h-Hd | WNL | WD | WNL | VN  | WNL |
| 6    | 43  | M      | B          | SF           | SF             | DC  | DC | 20/150 | 20/200 | None          | N   | N   | H-Hd | H-hd | ND | ND | ND  | ND |
| 7    | 61  | F      | L          | DNA          | XF             | WNL | PC | 20/20  | 20/25  | None          | N   | N   | WNL | H-Hd | WD | WNL | VN  | VN |
| 8    | 42  | F      | B          | SF           | XF             | PC  | PC | 20/200 | 20/20  | None          | Y   | Y   | h-Hd+h-He | h-He | CNV| CNV | CNV | CNV |
| 9    | 38  | M      | L          | DNA          | XF             | WNL | C  | 20/20  | 20/30  | CSC           | N   | Y   | WNL | h-He | WNL| CSC | WNL | VN |
| 10   | 30  | F      | B          | SF           | SF             | DC  | DC | 20/2000| 20/2000| None          | N   | N   | h-He | h-He | WD | WD | ND  | ND |
| 11   | 35  | F      | L          | DNA          | SF             | 20/2000| 20/150| None          | WNL | h-Hd | WNL | ND  | ND | ND | ND  | ND |

*F* female, *M* male, *L* left, *R* right, *B* bilateral, *RE* right eye, *LE* left eye, *DNA* does not apply, *SF* subfoveal, *XF* extrafoveal, *WNL* within normal limits, *C* calcified, *PC* partially decalcified, *DC* decalcified, *BCVA* best corrected visual acuity, *CSC* central serous chorioretinopathy, *CNV* choroidal neovascularization, *N* no, *Y* yes, *h-He* hypo-autofluorescent with hyper-autofluorescent edge, *h-Hd* hyper-autofluorescent with hypo-autofluorescent dots, *H-hd* hypo-autofluorescent with hyper-autofluorescent dots, *H-he* hyper-autofluorescent with hypo-autofluorescent edge, *FA* fluorescein angiography, *WD* window defect, *OCT-A* optical coherence tomography angiography, *DB* dark background where decalcification was present, *VN* vascular network within tumor, *AV* absence of vascular flow within tumor.
Methods

The clinical records of patients with a diagnosis of cho-
roidal osteoma who had undergone multimodal fundus
imaging on the retina service at Asociación para Evitar
la Ceguera en México were reviewed. The diagnosis was
based on the presence of a yellow-white to orange-red
mass deep to the RPE and bone density on ultrasonogra-
phy. Institutional review board approval was obtained for
this retrospective study.

Patient data were extracted from medical records
and included age at diagnosis, gender (male, female),
chief complaint, ocular comorbidities. Ophthalmic fea-
tures included best-corrected visual acuity (BCVA),
tumor laterality (unilateral or bilateral), location (foveal,
extrafoveal).

Multimodal imaging analysis included ultrasonogra-
phy (US), fluorescein angiography (FA), ultra-widefield
retinal imaging (UWF), ultra-widefield autofluorescence
(UWF-FAF), optical coherence tomography (OCT) and
OCT angiography (OCT-A).

Enhanced depth imaging optical coherence tomogra-
phy (Spectralis HRA+OCT; Heidelberg Engineering,
Germany), data included tumor surface configuration
(flat or depressed), effects of tumor on overlying retina
(RPE, photoreceptor and inner retina status). One inde-
pendent physician manually measured osteoma thick-
ness with a caliper function through the epicenter of the
tumor.

Ultra-wide field color fundus photograph and ultra-
widefield fundus autofluorescence (Optos Daytona;
Optos PLC, United Kingdom) data included tumor loca-
tion (foveal, extrafoveal), tumor color (yellow, orange,
white), and fundus autofluorescence pattern. Decalcifica-
tion Calcification (complete, partial) was defined as pale
areas within the osteoma, RPE thinning and visibility of
underlying choroidal vessels.

Fluorescein angiography (FA) (Spectralis HRA + OCT;
Heidelberg Engineering, Germany) data included pres-
ence or absence of CNV. OCT-A images were analyzed in
patients in whom FA was performed and correlated with

![Fig. 1 Multimodal imaging in choroidal osteoma.](image)

- a Fundus photograph shows a flat well-demarcated orange lesion in the macular area.
- b, c Fluorescein angiogram showing early hyperfluorescence and quiescent late staining in the juxtapapillary area (yellow arrow).
- d FAF showing predominantly hyper-AF in the juxtapapillary area.
- e, f OCT-EDI showing a focal choroidal excavation (asterisk).
- g, h OCT-A Boundaries of quiescent CNV in deep plexus (red circle).
- i B-scan ultrasonography consistent with CO (green asterisk)
the presence or absence of CNV (SS OCT Angio; Topcon Corporation, Japan).

**Results**

There were 16 eyes in 11 patients with choroidal osteoma included in this study. The demographic and clinical characteristics are summarized in Table 1. All patients were Hispanic and diagnosis corresponded to primary CO in 15 eyes, whereas one patient had CO secondary to choroidal hemangioma. The median age at presentation was 33.4 years (range 20–61 years). Most patients were female (72.7%).

Initial symptoms included blurred vision [9 patients, (82%)], metamorphopsia [1 patient, (9%)], asymptomatic [1 patient, (9%)]. Ocular conditions that accompanied the diagnosis of CO were keratoconus (2 patients) and central serous chorioretinopathy (1 patient). Visual acuity was 20/20–20/50 in 7 eyes (44%), 20/60–20/150 in 2 eyes (12%), 20/200 or worse in 7 eyes (44%). Poor visual acuity (20/200 or worse) was related to foveal photoreceptor loss overlying deossified osteoma (n = 6), subfoveal choroidal neovascular membrane (n = 1), keratoconus (n = 1).

Five patients (45.5%) had bilateral CO; while the other 54.5% had unilateral CO. The osteoma was completely calcified in 4 eyes (25%), partially decalcified in 8 eyes (50%) and decalcified in 4 eyes (25%). Tumor location was subfoveal in 12 eyes (75%); extrafoveal in 4 eyes (25%).

CO showed different FAF patterns, which we classified as normal autofluorescence (isoautofluorescent, 6.25%), predominantly hyper-AF (37.5%) and predominantly hypo-AF (56.25%); decreased fluorescence on FAF was observed in decalcified tumoral regions while relatively preserved fluorescence was observed in calcified regions. Patients with worse visual acuity (≤20/200) presented predominantly hypo-AF pattern (5 eyes, 31.25%).

OCT data demonstrated a mean central foveal thickness of 265.5 μm (range 101–599 μm), a mean subfoveal choroidal thickness of 498.17 μm (range 288–736), and a mean central tumor thickness of 574.86 μm (range 246–1084). Patients with decalcified portion of tumor displayed and overlying thinned inner retinal layers in 4 eyes (25%), thinned outer retina with thinned to absent photoreceptor layer in 12 eyes (75%), and overlying RPE hyperplasia in 3 eyes (25%).

Seven eyes with clinical suspicion of choroidal CNV were imaged with FA and OCT-A. Leakage of fluorescein dye was present in 2 patients (3 eyes, 18.75%); OCT-A made evident the location of abnormal vascular network in outer

![Multimodal imaging in choroidal osteoma.](image-url)
Fig. 3  Choroidal depression associated with tumor decalcification. Top: Optical coherence tomography progression of a patient with partially calcified CO. Middle and bottom: Choroidal vessels become prominent in decalcified areas. RPE-photoreceptor detachment induced by decalcification (asterisk). These images were taken after 13 and 17 months, respectively.
Table 2 Literature review of case reports analyzing choroidal osteoma characteristics

| Authors             | Year | n (eyes) | Results                                                                                                                                 |
|---------------------|------|---------|----------------------------------------------------------------------------------------------------------------------------------------|
| Shields et al. [12] | 2007 | 22      | OCT: calcified portion displayed an intact inner retina, outer retina and photoreceptors, but decalcified portion showed intact inner retina with thinned or absent outer retina and photoreceptors. BCVA was better in eyes with calcified osteomas |
| Margolis et al. [13]| 2011 | 13      | AF: hypofluorescence. Indocyanine green angiography showed relative hypofluorescence. SD-OCT: separation between outer retina and RPE within the excavation; and other cases in which the outer retina layers conform to the retina pigment epithelium within the excavation. Choroidal thickness of uninvolved choroid was thicker than normal |
| Freton et al. [9]   | 2011 | 11      | SD-OCT: different reflectivity pattern among hyporeflective, isoreflective and hyperreflective, besides retina exhibited degenerative changes |
| Navajas et al.      | 2012 | 3       | FD-OCT show in calcified tumors a distinctive latticework pattern resembling a spongy bone structure, decalcified areas show hyporeflective areas above Bruch membrane and absence of choroidal vessels. AF: Decalcified tumor had reduced over all fluorescence |
| Shields et al. [11] | 2015 | 15      | EDI-OCT: horizontal lamellar lines, hyperreflective horizontal lines, horizontal and vertical tubular lamella. Photoreceptors were intact in ossified tumors meanwhile those were atrophic or thinning in deossified osteomas |
| Pierro et al. [14]  | 2017 | 3       | FCE and CNV in CO. OCT-A is a useful skill to detect CNV |
| Cennamo et al. [17] | 2017 | 6       | OCT-A: fine vascular network within the tumor. EDI-OCT: horizontal lamellar lines, horizontal and vertical tubules and speckled regions. B-Scan echography: solid mass with acoustic shadowing |

**Discussion**

Choroidal osteomas may demonstrate decalcification, CNV, retinal pigment epithelium (RPE) alterations and vision loss [6]. Patients with calcified areas, even subfoveal ones, had better visual acuities; while patients with decalcified CO had lower visual acuities correlated with RPE disruption and outer layer thinning and photoreceptor loss, and corresponded to hypo-AF on AF [8, 12].

Table 1. In our series two eyes (12.5%) presented CNV whoes medical records had FA and OCT-A. OCT-A analysis showed the following 4 patterns: absence of vascular flow within tumor (6.25%), dark background where decalcification was present (12.5%), vascular network within tumor (25%), and presence of a neovascular membrane (18.75%).

**Conclusions**

Choroidal osteoma is an ossifying tumor involving the choroid, its natural course may include tumor growth, calcification and decalcification; visual acuity depends on choroidal neovascularization and retinal changes associated to decalcification [12]. Duration of this condition is a mayor risk factor associated with vision loss. After 10 years, approximately 51% manifest evidence of growth and nearly 50% showed decalcification. Calcified and decalcified areas have demonstrated changes in outer retina. Optical coherence tomography changes have been shown that calcified areas have intact outer retina whereas decalcified portion have thinned to absent outer retina and photoreceptor layers [12].

In our series, the mean age at diagnosis was 33 years and females represented 72.7%, however 45.5% of our patients were bilateral, this patients showed osteomas located in the macular area with extension beyond the vascular arcades and showed RPE alterations due to osteoma decalcification.

Patients with CNV whose medical records had FA and OCT-A also where evaluated, both have good correlation in determining the site of neovascularization.
Choroidal excavations observed in this series correlate with previous descriptions made by Lampol et al. [15, 16]. Table 2. To our best knowledge Pierro et al. [14] described two patients with CO and FCE in the proximity of CNV. In this series two eyes had CNV in the proximity of FCE and one eye had FCE in the boundaries of the osteoma and normal choroidal tissue.

Although several authors have already described clinical and OCT characteristics of CO, to our best knowledge this is the first case series report of multimodal imaging findings of CO in Hispanic patients. Because this is a rare pathology, the number of cases reported in the literature is scarce, and this also constituted a limitation to our study. However, we believe these findings can deepen the information about the behavior of this uncommon tumor.

Abbreviations
CO: choroidal osteoma; CNV: choroidal neovascularization; OCT: optical coherence tomography; EDI-OCT: enhanced depth imaging optical coherence tomography; UWF: ultra-widefield retinal imaging; UWF-FAF: ultra-widefield fundus autofluorescence; FA: fluorescein angiography; FCE: focal choroidal excavation; APEC: Hospital “Dr. Luis Sánchez Bulnes” from the Asociación para Evitar la Ceguera en México I.A.P; BCVA: best-corrected visual acuity.

Authors’ contributions
FOM made substantial contributions to conception and design, acquisition of data, analysis and interpretation of data. MMG made substantial contributions to acquisition of the manuscript. NC organized and analyzed patient images. VSV contributed writing the manuscript. AE was a major contributor in writing the manuscript. NC organized and analyzed patient images. VSV contributed writing the manuscript. MMG made substantial contributions to acquisition of data. IA performed search and organization of data. VMC coordinated the investigation. JMJS analyzed and interpreted patient data. All authors read and approved the final manuscript.

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Acknowledgements
Carlos Andrés Valdés-Lara, MD for substantial contributions to acquisition of data. Miguel Espitia, MD for contributions to acquisition of reference articles.

Competing interests
The authors declare that they have no competing interests.

Availability of data and materials
The datasets used and analyzed during the current study are available from the corresponding author on reasonable request.

Consent for publication
All patients signed a consent publication form.

Data access and responsibility
The principal investigator, Francisco Olguín-Manríquez, had full access to all of the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

Ethics approval and consent to participate
Institutional review board approval was obtained for this study.

Funding
“Dr. Luis Sánchez Bulnes” Hospital from Asociación Para Evitar la Ceguera en México I.A.P provided funding for this study. The Hospital had no role in design and conduct of the study, collection, management, analysis, and interpretation of the data; and preparation, review, or approval of the manuscript.

Publisher’s Note
Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

Received: 10 February 2018 Accepted: 22 July 2018
Published online: 15 August 2018

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