

Circumscribed choroidal hemangioma with serous retinal detachment

Hemangioma coroideano circunscrito com descolamento seroso de retina

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ABSTRACT

Circumscribed choroidal hemangioma is a rare, benign vascular tumor of the choroid that can cause severe macular complications and irreversible vision loss despite its non-malignant nature. We report the case of a 41-year-old patient with no significant medical history who presented with progressive visual loss in the right eye. Fundus examination revealed a circumscribed yellow-orange choroidal mass located along the inferotemporal vascular arcade, associated with serous retinal detachment involving the macula. Multimodal imaging findings strongly supported the diagnosis of CCH. As photodynamic therapy was not available, proton beam therapy was considered as the best alternative therapeutic option. This case highlights the importance of differentiating circumscribed choroidal hemangioma from amelanotic melanoma and other mimicking lesions and illustrates the role of proton beam radiotherapy as an effective alternative when photodynamic therapy is not accessible.

RESUMO

O hemangioma coroideano circunscrito é um tumor vascular benigno e raro da coroide, capaz de causar complicações maculares graves e perda visual irreversível, apesar de sua natureza não maligna. Relatamos o caso de um paciente de 41 anos, sem antecedentes médicos relevantes, que apresentou perda visual progressiva no olho direito. O exame de fundo de olho revelou uma massa coroideana circunscrita de cor amarelo-alaranjada localizada ao longo da arcada vascular inferotemporal, associada a descolamento seroso de retina envolvendo a mácula. Os achados da imagem multimodal corroboraram fortemente o diagnóstico de hemangioma coroideano circunscrito. Como a terapia fotodinâmica não estava disponível, a radioterapia com feixe de prótons foi considerada a melhor alternativa terapêutica. Este caso destaca a importância de diferenciar o hemangioma coroideano circunscrito do melanoma amelanótico e de outras lesões que o mimetizam, além de ilustrar o papel da radioterapia com prótons como uma alternativa eficaz, quando a terapia fotodinâmica não é acessível.

INTRODUCTION

Choroidal hemangioma is a rare, congenital vascular tumor of the choroid. Two main forms are described: the diffuse type, usually associated with Sturge-Weber syndrome, and the circumscribed type, which is sporadic and typically becomes symptomatic in adulthood (4th to 6th decade).^(1,2) Although benign, circumscribed choroidal hemangioma (CCH) may lead to progressive vision loss due to exudation, serous retinal detachment (SRD), and secondary degenerative changes of the retinal pigment epithelium (RPE) and photoreceptors.

Because of its clinical similarity with choroidal melanoma, metastasis, nevus, or osteoma, accurate diagnosis is crucial to avoid mismanagement.

Despite its benign nature, CCH presents significant diagnostic and therapeutic challenges, particularly in settings where access to first-line treatment modalities is limited.

Therefore, the objective of this case report was to highlight the importance of multimodal imaging in establishing an accurate diagnosis of CCH and to discuss the available therapeutic options. This report was approved by the Ethics Committee of the Tunis Military Hospital (CAAE number: 92/2025/CLPP).

CASE REPORT

A 41-year-old patient with no relevant medical or family history presented with progressive visual decline in

the right eye. Best-corrected visual acuity (BCVA) was measured at 1/10 in the right eye and 20/20 in the left eye. Examination of the anterior segment was unremarkable in both eyes. Fundus examination revealed a circumscribed yellow-orange choroidal mass located along the inferotemporal vascular arcade, associated with an SRD extending to the macula (Figure 1A). Optical coherence tomography (OCT) demonstrated a dome-shaped choroidal mass with subretinal fluid, RPE alterations, cystoid macular edema and outer retinal damage involving the photoreceptors. (Figure 1B) Fluorescein angiography revealed early diffuse hyperfluorescence that persisted in late phases. Indocyanine green (ICG) angiography was not available. B-scan ultrasonography confirmed a homogeneous, echogenic dome-shaped choroidal mass with associated SRD. Orbital magnetic resonance imaging (MRI) revealed a lesion that was hyperintense on T1, isointense on T2, and exhibited intense homogeneous enhancement after gadolinium injection (Figure 2).

Since photodynamic therapy was not available, proton beam therapy was considered the most appropriate therapeutic alternative for our patient. Following treatment, the patient demonstrated structural improvement with a marked reduction in macular edema; however, visual acuity remained limited to 2/10, likely due to persistent outer retinal damage (Figure 3).

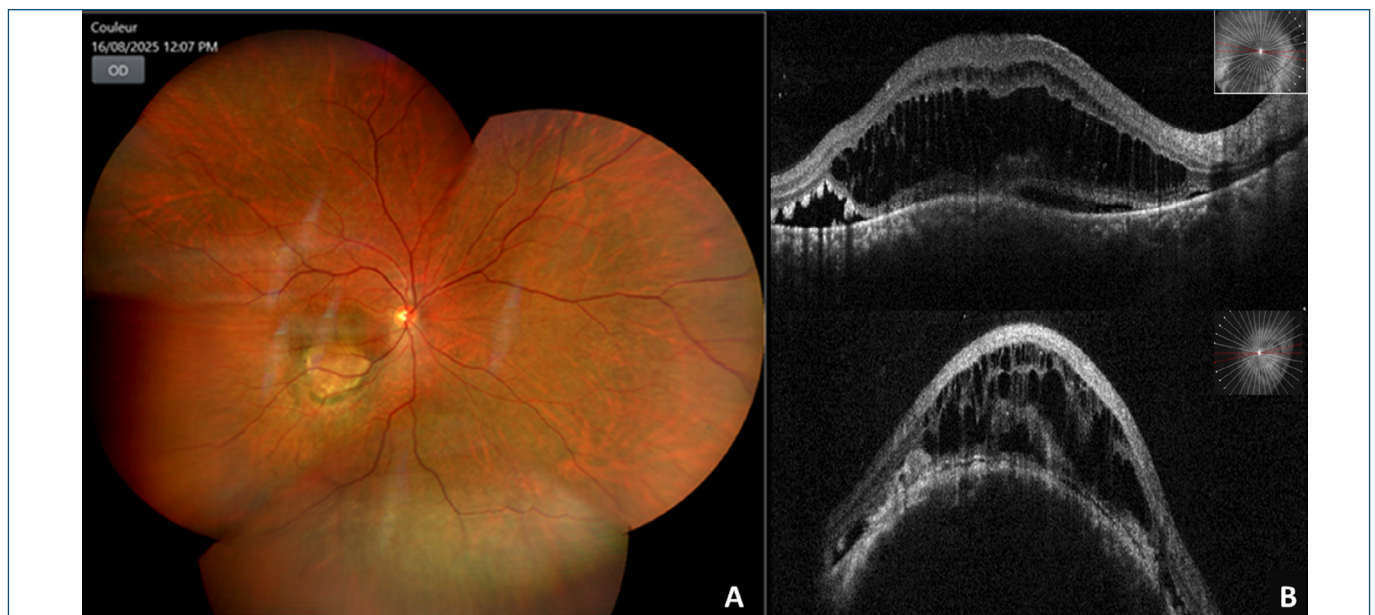


Figure 1. (A) Fundus photography of the right eye showing a circumscribed yellow-orange choroidal mass located along the inferotemporal vascular arcade. (B) Optical coherence tomography (OCT) demonstrating a dome-shaped choroidal mass with subretinal fluid, retinal pigment epithelium alterations, cystoid macular edema, and outer retinal damage.

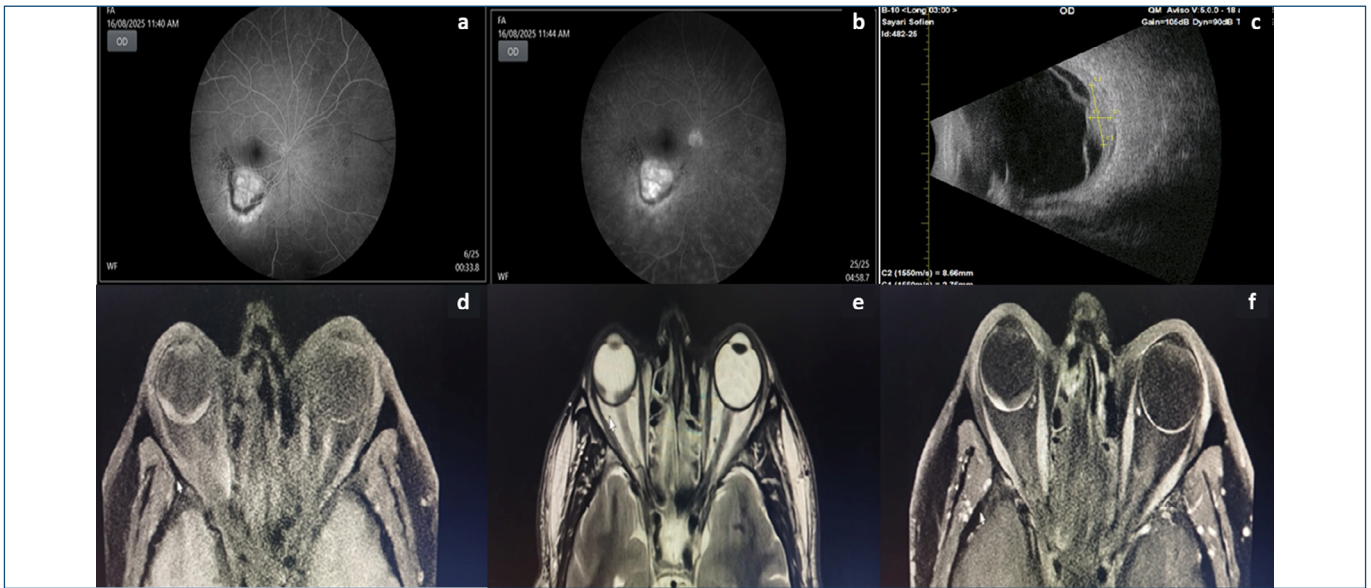


Figure 2. (A and B) Fluorescein angiography showing early diffuse hyperfluorescence that persisted in the late phases. (C) B-scan ultrasonography showing a homogeneous, echogenic dome-shaped choroidal mass with associated serous retinal detachment. (D to F) Magnetic resonance imaging revealed a lesion that was hyperintense on T1, isointense on T2, and exhibited intense homogeneous enhancement after gadolinium injection.

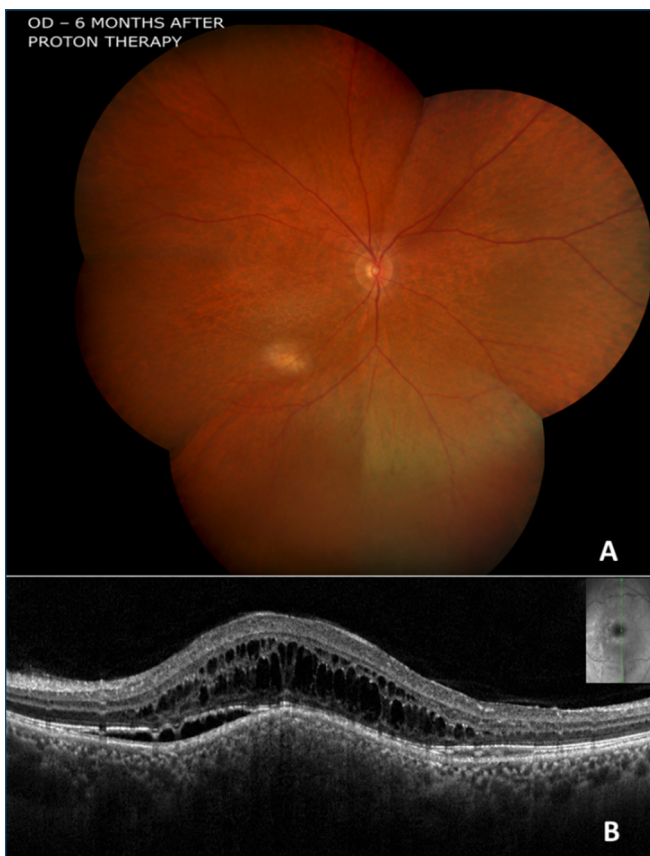


Figure 3. (A) Follow-up fundus photograph of the right eye obtained 6 months after proton beam therapy, showing partial regression of the circumscribed choroidal hemangioma. (B) Optical coherence tomography demonstrating significant reduction of cystoid macular edema and subretinal fluid, with residual outer retinal changes.

DISCUSSION

Circumscribed choroidal hemangioma is a rare, benign vascular tumor that typically becomes symptomatic in adulthood. Circumscribed choroidal hemangiomas are typically found at the posterior pole, most often in the superotemporal quadrant near the macula, and are characterized by sharply demarcated borders composed of compressed melanocytes and choroidal lamellae.^(3,4)

Histologically, CCH is characterized by a proliferation of thin-walled, blood-filled vascular channels within the choroid. Two principal morphologic patterns are recognized: cavernous, composed predominantly of large dilated vascular spaces separated by delicate stromal septa, and capillary, consisting of smaller, capillary-sized vessels embedded in a loosely organized connective tissue matrix. Most lesions exhibit a mixed pattern, demonstrating variable proportions of both vascular types. The endothelial lining is typically flat and inconspicuous, without evidence of cytologic atypia, mitotic activity, or significant perivascular smooth muscle proliferation, supporting the non-neoplastic nature of the lesion.⁽⁵⁾

Although non-malignant, it may lead to significant visual loss through chronic subretinal fluid, SRD, and secondary degenerative changes such as RPE hyperplasia, subretinal fibrosis, retinoschisis, and photoreceptor degeneration.⁽⁶⁾

On fundus examination, the lesion typically appears as a sharply demarcated, red-orange mass, sometimes bordered by subtle basal pigmentation. It is most

commonly identified at the posterior pole, representing roughly two-thirds of reported cases.⁽⁷⁾ A predilection for the superotemporal juxtamacular region has been noted.⁽⁴⁾ CCHs are almost always solitary and affect only one eye; bilateral involvement remains uncommon, although a few exceptions have been described in the literature.^(8,9)

Differentiating CCH from other choroidal lesions is crucial, particularly from amelanotic choroidal melanoma, which usually presents as a pigmented dome- or mushroom-shaped mass with low-to-medium echogenicity and compression of the choriocapillaris on OCT.⁽¹⁰⁾ Other differential diagnoses include choroidal metastases, which are often multifocal and yellowish; choroidal nevus, generally flat with drusen on its surface; and choroidal osteoma, a calcified whitish lesion with high reflectivity and posterior acoustic shadowing on ultrasound.^(4,11)

Management depends on the presence of symptoms and lesion characteristics. Observation is recommended for asymptomatic cases, while treatment is indicated in patients with visual decline or persistent subretinal fluid.⁽¹⁾

Photodynamic therapy with verteporfin is considered the first-line treatment. Photodynamic therapy may induce regression or atrophy of the vascular channels within the hemangioma while sparing the neurosensory retina, leading to reduced vascular leakage and subsequent improvement or stabilization of vision.⁽¹²⁾

Transpupillary thermotherapy may be used as an alternative but is primarily indicated for extrafoveal, post-equatorial circumscribed choroidal hemangiomas characterized by mild subretinal fluid, with a basal diameter under 10 mm and a thickness below 4 mm.⁽¹³⁾

Transpupillary thermotherapy can result in complete or partial tumor regression in approximately half of the treated cases, although it carries an increased risk of fibrosis and choroidal atrophy.⁽¹⁴⁾

Radiotherapy, particularly proton beam therapy, represents a valuable and minimally invasive treatment option for larger tumors (> 3 mm) or extensive SRDs, offering good functional outcomes in a single session.⁽¹⁵⁾

Adjuvant therapies such as intravitreal anti-vascular endothelial growth factor (VEGF) or dexamethasone implants may reduce macular edema by reducing vascular wall permeability and enhancing the efficacy of photodynamic therapy.⁽⁷⁾ Enucleation is reserved for painful blind eyes with neovascular glaucoma.

Circumscribed choroidal hemangioma is a rare but clinically significant benign vascular tumor whose macular complications may lead to severe and irreversible visual impairment. Accurate diagnosis is essential, as

its clinical and imaging characteristics may mimic other amelanotic choroidal tumors with major therapeutic implications. Ultimately, early recognition and appropriate selection of the therapeutic modality are key to optimizing visual outcomes in patients with CCH.

AUTHORS' CONTRIBUTION

Oueslati Yassin and Mrabet Farah contributed to the conception, writing, and critical review of the manuscript. Elkassar Ghita and Houatmi Ahlem contributed to the writing and preparation of illustrations. Khallouli Asma and Maalej Afef contributed to the critical review of the manuscript. All authors approved the final version of the manuscript and are responsible for all aspects of the manuscript, including ensuring its accuracy and integrity.

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