

Retinal astrocytic hamartoma versus choroidal osteoma: an interesting case

Hamartoma astrocítico da retina versus osteoma de coróide: um caso interessante

Hamartoma astrocítico de la retina versus osteoma de la coróide: un caso interesante

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ABSTRACT

The clinical case presents an interesting discussion about differential diagnosis between choroidal osteoma and retinal astrocytic hamartoma. A healthy 19-year-old man presented with a dark spot on temporal field of the left eye with one month of evolution. Examination of the ocular fundus of the left eye revealed a well-circumscribed and elevated orange-yellow plaque around the optic disc and hemorrhages in the macular region. The patient was submitted to fluorescein angiography, ocular ultrasound, optical coherence tomography and optical coherence tomography angiography. The yellowish mass, the optically empty spaces and the calcifications seen in the optical coherence tomography, besides the macular hemorrhage are inherent characteristics of the two tumors, making it very difficult to reach the etiological diagnosis. An Enhanced depth imaging optical coherence tomography and a cytological evaluation through fine needle biopsy would be useful tools to clarify the diagnosis.

Keywords: Tomography Optical Coherence; Retina; Hamartoma; Osteoma; Choroid.

RESUMO

Este estudo de caso clínico apresenta uma discussão interessante sobre o diagnóstico diferencial entre osteoma de coróide e hamartoma astrocítico da retina. Um indivíduo saudável do sexo masculino, de 19 anos de idade, apresentou uma mancha escura no campo temporal do olho esquerdo com 1 mês de evolução. O exame de fundo ocular do olho esquerdo revelou uma placa laranja-amarelada bem circunscrita e elevada ao redor do disco óptico e hemorragias na região macular. O paciente foi submetido a angiografia com fluoresceína, ultrassonografia ocular, tomografia de coerência óptica (OCT) e angiotomografia de coerência óptica (OCTA). Uma massa amarelada, espaços opticamente vazios e calcificações foram detectados na OCT, além das hemorragias maculares que são características inerentes aos dois tumores, dificultando enormemente o diagnóstico etiológico. Uma tomografia de coerência óptica com profundidade de imagem aprimorada e uma avaliação citológica por biópsia por agulha fina podem ser ferramentas úteis para esclarecer o diagnóstico.

Palavras-chave: Tomografia de Coerência Óptica; Retina; Hamartoma; Osteoma; Coróide.

RESUMEN

Un individuo saludable del sexo masculino, de 19 años de edad, presentó una mancha oscura en el campo temporal del ojo izquierdo con 1 mes de evolución. El examen de fondo ocular del ojo izquierdo reveló una placa naranja-amarillada bastante incrustada y elevada alrededor del disco óptico y hemorragias en la región macular. Se sometió el paciente a angiografía con fluoresceína, ultrasonografía ocular, tomografía de coherencia óptica (OCT) y angiotomografía de coherencia óptica (OCTA). Se detectaron en la OCT una masa amarilla, espacios ópticamente vacíos y calcificaciones, además de las hemorragias maculares que son características propias a los dos tumores, lo que hace muy difícil el diagnóstico etiológico. Una tomografía de coherencia óptica con profundidad de imagen primoreada y una evaluación citológica por biopsia con aguja delgada pueden ser herramientas útiles al aclaramiento del diagnóstico.

Palabras Clave: Tomografía de Coherencia Óptica; Retina; Hamartoma; Osteoma; Coroides.

Funding: No specific financial support was available for this study.

CEP Approval: Not applicable

Disclosure of potential conflicts of interest: None of the authors have any potential conflict of interest to disclose.

Received on: May 30, 2018

Accepted on: Jul 25, 2018

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How to cite: Silva R, Moreira Neto CA, Alencar L. Retinal astrocytic hamartoma versus choroidal osteoma: an interesting case. eOftalmo. 2018; 4(3): 137-142 <http://dx.doi.org/10.17545/eoftalmo/2018.0029>

INTRODUCTION

Choroidal osteoma (CO) is a benign and rare tumor that is composed of mature bone cells and is more prevalent in females¹. CO is often located in the juxtapapillary or macular region and is unilateral in the majority of cases². The most common complication of CO is choroidal neovascularization (CNV), an important cause of visual loss, occurring in up to 31% of cases³.

Retinal astrocytic hamartoma (RAH) is a benign glial tumor arising from the retinal nerve fiber layer. This lesion represents the most common ophthalmoscopic finding in patients with tuberous sclerosis complex; however, this mass can also manifest as a sporadic condition that appears as a yellow–gray mass, often with minimally dilated retinal vessels, fine retinal traction, and glistening intrinsic calcification⁴.

CASE REPORT

A 19-year-old Caucasian male presented with a dark spot on the temporal field of the left eye with 1 month of evolution. He had no other relevant past ocular, medical, or family history. His best corrected visual acuity was 20/20 in the right eye and 20/20 in the left eye. Examination of his anterior segment was unremarkable, in addition to a normal ocular fundus examination of the right eye. Fundoscopy of the left eye revealed a well-circumscribed and elevated peripapillary orange-yellow plaque and hemorrhage in the macular region (Figure 1A). Fundus autofluorescence (FAF) revealed a hyperautofluorescent lesion throughout the tumor extension and a fluorescent hypopigment in the hemorrhage area (Figure 1B).

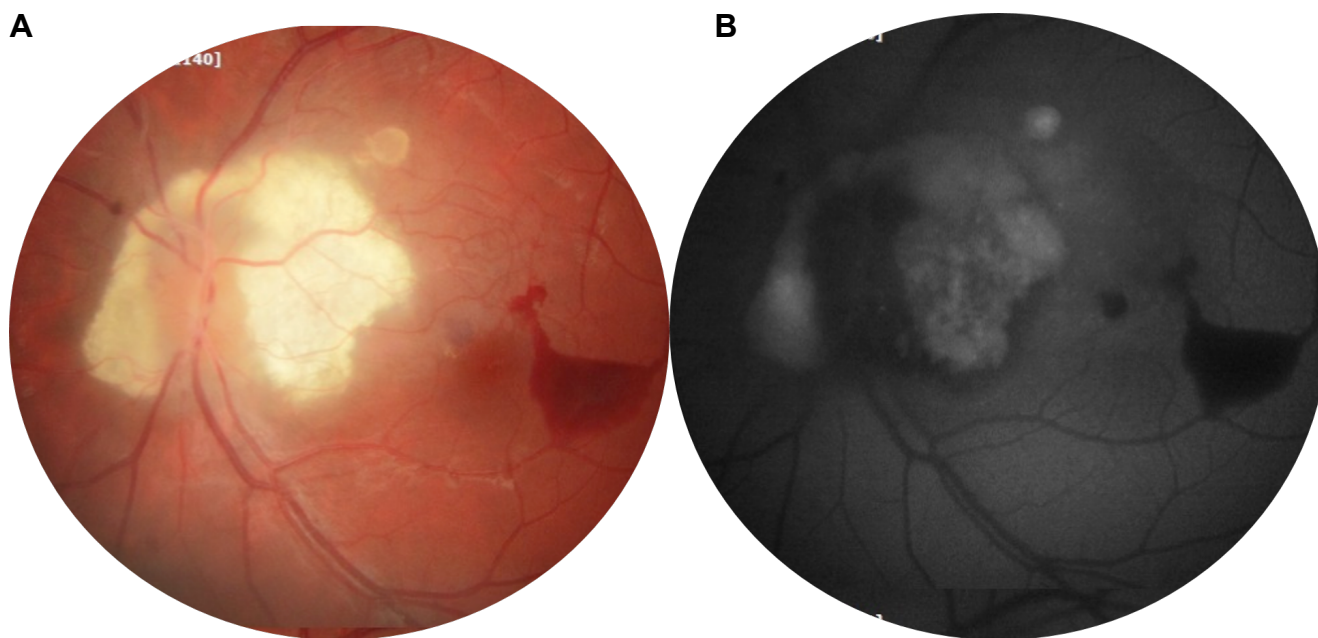


Figure 1. A: Retinography showed a well-circumscribed and elevated orange–yellow plaque around the optic disc and hemorrhages in the macular region; **B:** The fundus autofluorescence showed a hyperautofluorescent lesion throughout the tumor extension and fluorescent hypopigment in the hemorrhage area.

Color fundus photography, B-scan ultrasound, fundus fluorescent angiography (FFA), spectral domain optical coherence tomography (SD-OCT) (Heidelberg Engineering, Heidelberg, Germany), and optical coherence tomography angiography (OCTA) (Optovue, Inc., Fremont, CA, USA) were performed. FFA of the left eye revealed early hyperfluorescence and intense staining of the lesion associated with areas of blockage corresponding to the hemorrhage (Figure 2). The B-scan ultrasonography revealed a highly echogenic lesion with posterior acoustic shadowing (Figure 3).

The SD-OCT revealed a well-defined retinal lesion with optically empty spaces (OESs) and the presence of hyperreflective dots (Figure 4).

The OCTA demonstrated tumor vessels in the superficial vascular plexuses, and the preretinal hemorrhage was observed in the en-face OCT (Figure 5).

The patient received three consecutive monthly intravitreal injections of aflibercept (2.0 mg). After 1 month of the final intravitreal aflibercept injections, the OCTA revealed a reduction in the bleeding area.

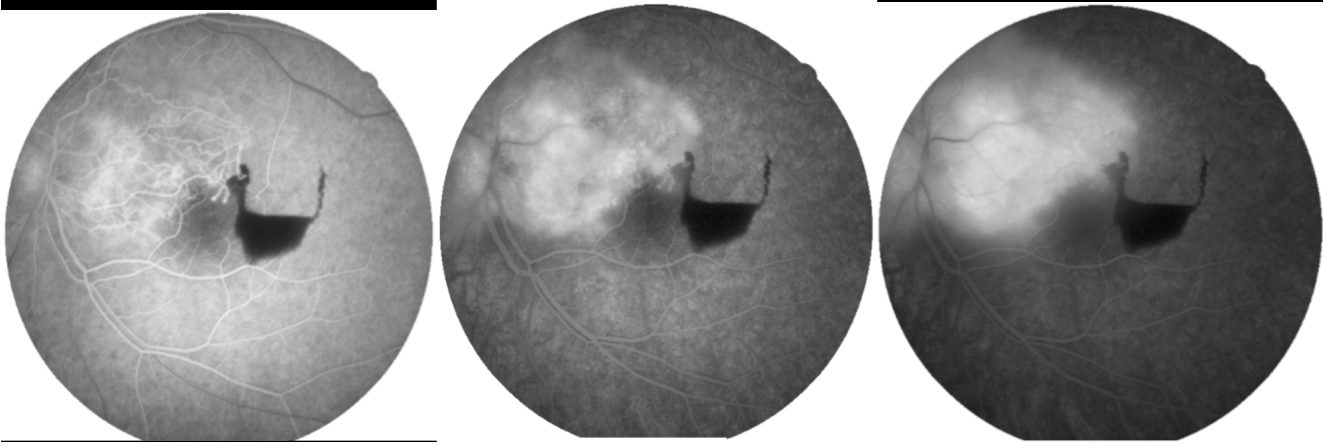


Figure 2. Fluorescent angiography of the left eye showed early hyperfluorescence and intense staining of the choroidal lesion associated with areas of blockage corresponding to preretinal hemorrhage.

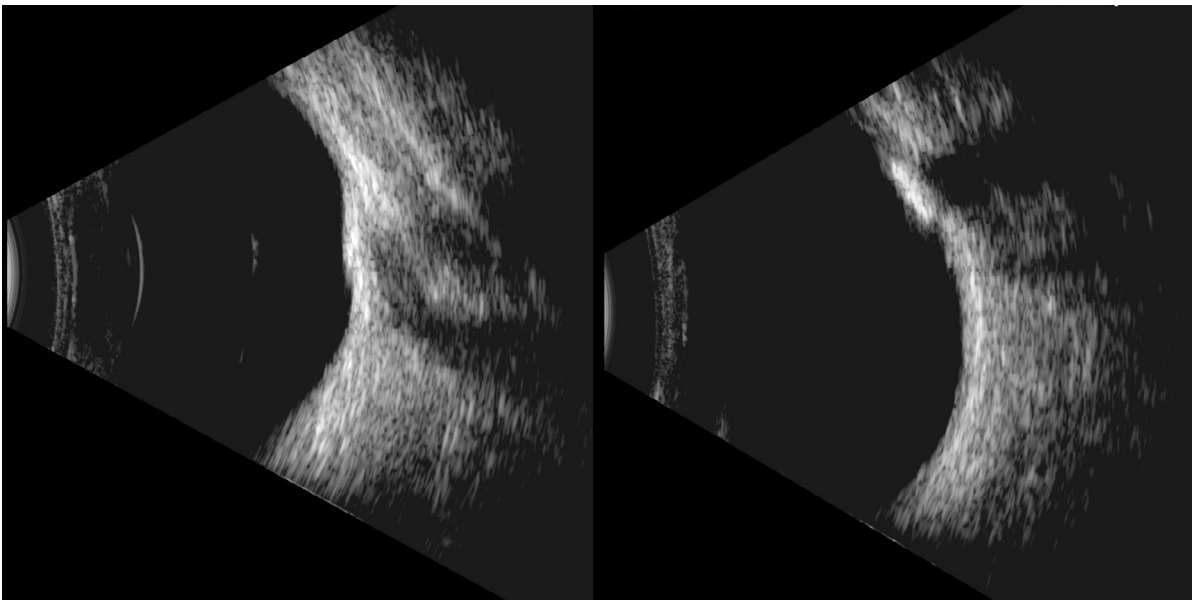


Figure 3. The B-scan ultrasonography revealed a highly echogenic lesion with posterior acoustic shadowing.

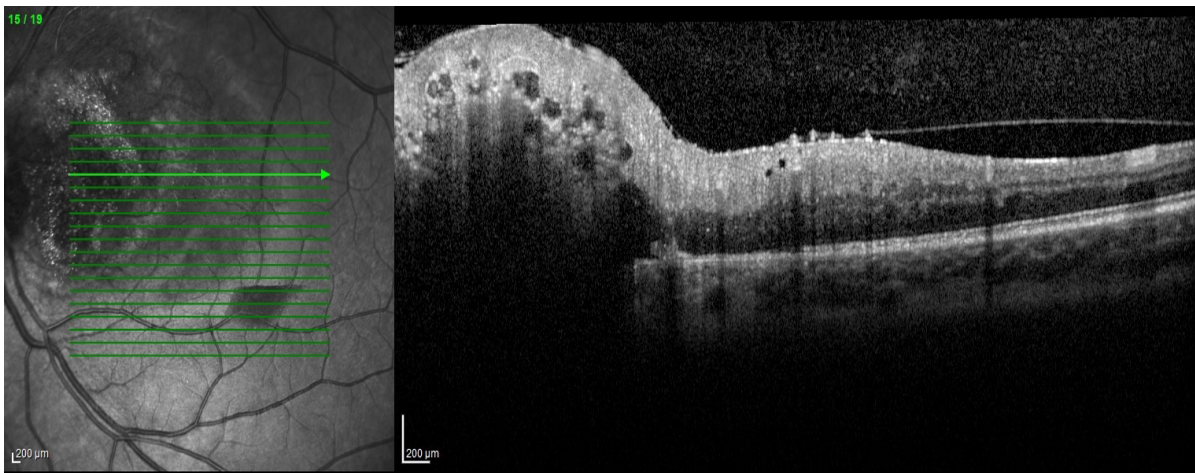


Figure 4. Spectral domain optical coherence tomography showed a well-defined retinal lesion with optically empty spaces and the presence of hyperreflective dots shadowing the choroid.

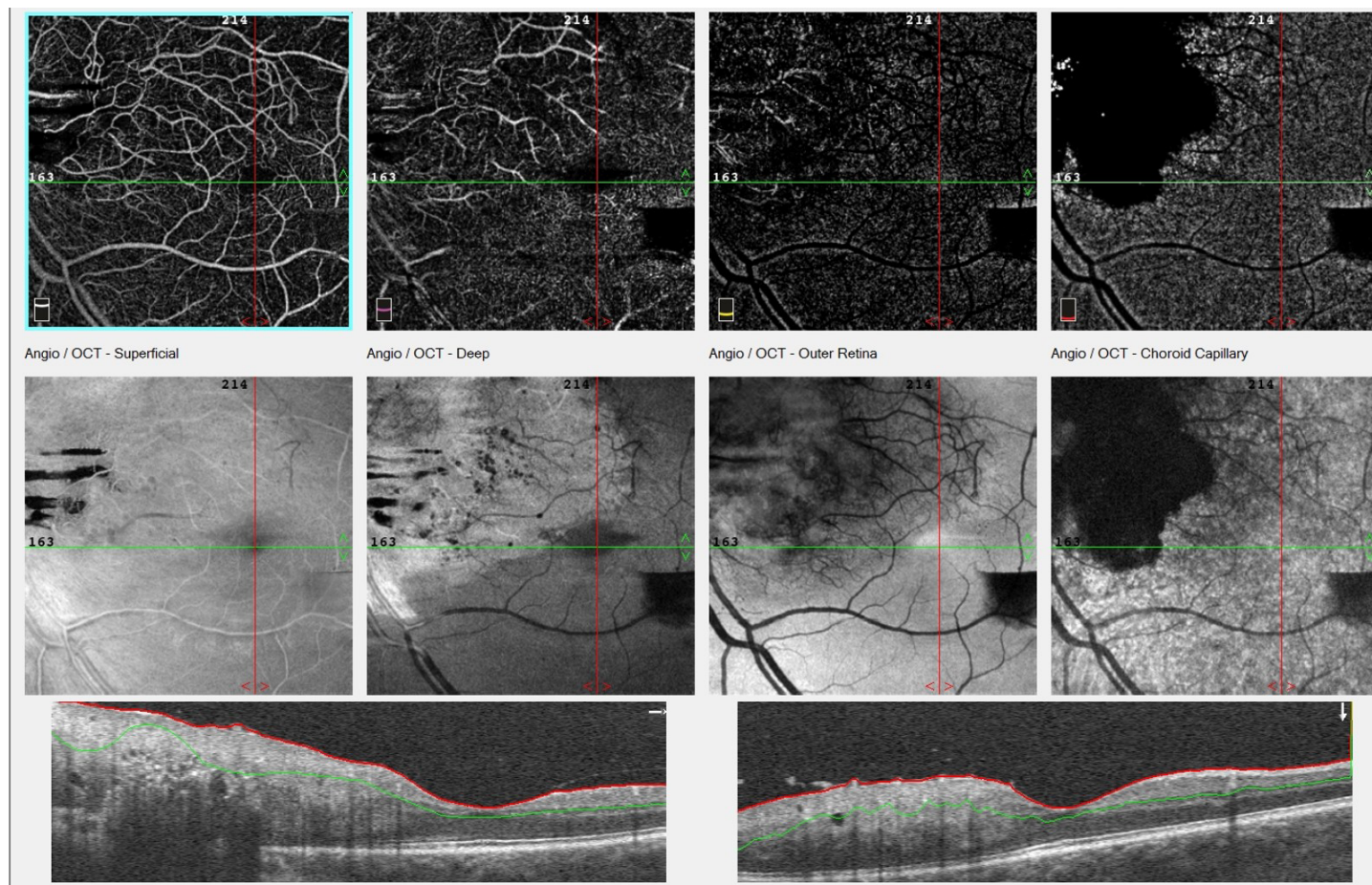


Figure 5. Optical coherence tomography angiography revealed tumor vessels in the superficial vascular plexuses, and the preretinal hemorrhage was detected in the en-face optical coherence tomography.

In addition, yellow laser photocoagulation was performed at the temporal margin of the tumor (Figure 6).

The final visual acuity of the patient was 20/20, and there was improvement in the visual field defect in the left eye after 2 months of treatment.

DISCUSSION

The unique, peripapillary, yellowish lesion associated with the macular hemorrhage led us to differentiate two tumors that could be calcified, which, in this topography, are CO and RAH⁵.

The patient did not present systemic alterations such as tuberous sclerosis complex. Ocular ultrasound revealed a calcified lesion and a macular hemorrhage that could be caused due to a neovascular membrane secondary to CO. However, in the OCT, the lesion was located in the retina, and it was not possible to evaluate the choroid due to the shadowing caused by the tumor.

Analysis of the CO using magnified EDI-OCT images reveals a typical sponge-like pattern comprising dense hyperreflective dots spread into the hyporeflective matrix and a multilayer structure, probably because of the presence of different degrees of calcification within the tumor. It is believed that SD-OCT scanning of eyes with an amelanotic lesion in the fundus can facilitate clinicians to differentiate COs from other conditions such as sclerochoroidal calcifications, choroidal melanomas, choroidal metastasis, and choroidal lymphoma⁶⁻⁸.

Regarding EDI-OCT, Pichi et al. described type I as flat and generally in the nerve fiber layer, type II with slight elevation of the nerve fiber layer and retinal traction, type III with “moth-eaten” lucent areas suggestive of calcification involving the inner and outer retina, and type IV with optically empty intralesional cavities⁹.

RAH manifests characteristic features on an OCT, including a gradual transition from a normal retina to an optically hyperreflective mass with retinal disorganization, characteristic moth-eaten spaces, and posterior shadowing¹⁰⁻¹¹.

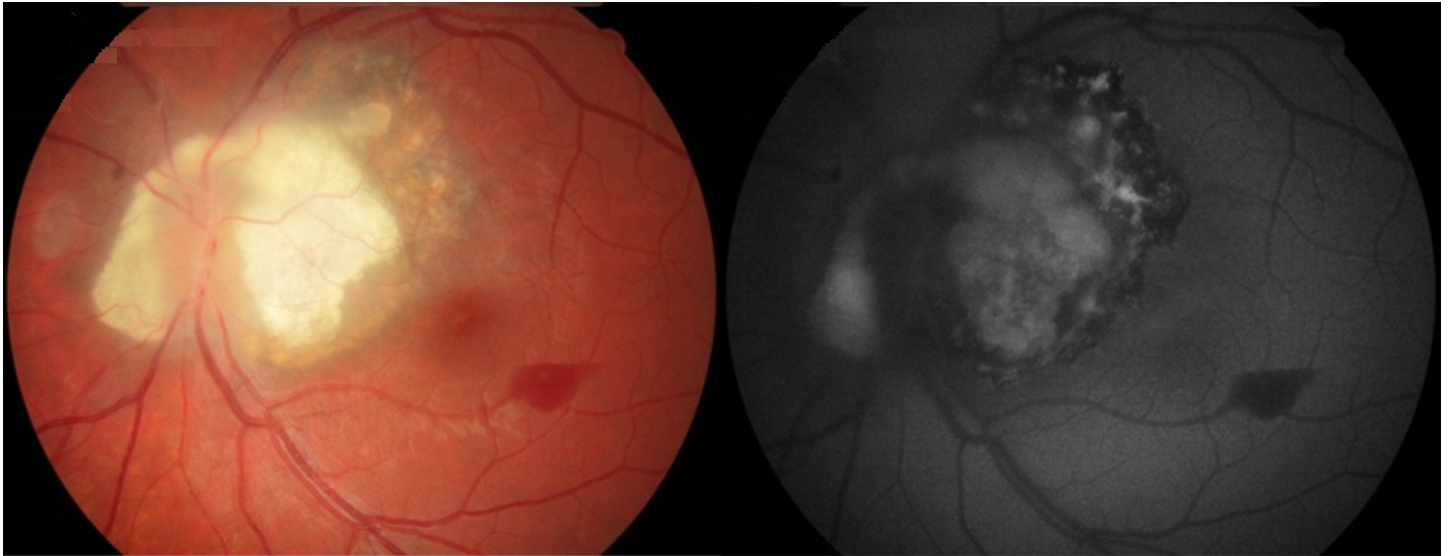


Figure 6. Retinography and fundus autofluorescence showed the tumor after yellow laser photocoagulation in the temporal margin.

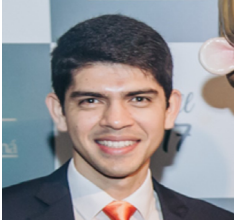
RAH is associated with tuberous sclerosis but may appear as a sporadic tumor, and intralesional calcification occurs in about half of the cases, in addition to hemorrhages and subretinal fluid that are more common in acquired astrocytomas¹².

The complications related to the presence of retinal hamartomas include vitreous hemorrhage, retinal vascular abnormalities (including telangiectasia, neovascularization, and exudation), and vitreous seeding, which is characteristically described in association with optic nerve head or epipapillary astrocytic hamartomas. According to a clinicopathologic report, vitreous hemorrhage may arise from the substance of retinal or optic nerve astrocytomas. A myriad of small vascular channels is confirmed to be pathologic¹³.

EDI-OCT and a cytological evaluation through fine needle biopsy could be useful tools to clarify the etiological diagnosis of this case.

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