Racemose hemangioma presenting as a vitreous hemorrhage: case report

Hemangioma racemoso apresentando-se como hemorragia vítrea: relato de caso

Osias Francisco de Souza[1], Lais Bomediano de Souza[2], Antonio Marcelo Barbante Casella[3], Acacio Muralha Neto[4], José Paulo Cabral de Vasconcellos[1]

[1] Department of Ophthalmology, Faculty of Medical Sciences, Universidade Estadual de Campinas, Campinas, SP, Brazil.
[2] Faculty of Medicine, Pontifícia Universidade Católica de Campinas, Campinas, SP, Brazil.
[3] Department of Ophthalmology, Universidade Estadual de Londrina, Londrina, PR, Brazil.
[4] Department of Ophthalmology, Hospital de Olhos de Niterói, Niterói, Rio de Janeiro, RJ, Brazil.

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Corresponding author:
Osias F. de Souza
Rua Engenheiro Carlos Stevenson, 66 – Cambuí
Zip code: 13092132 Campinas, SP, Brazil
E-mail: osiasfs@uol.com.br

Institution:
Department of Ophthalmology, Faculty of Medical Sciences, Universidade Estadual de Campinas, Campinas, SP, Brazil.

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ABSTRACT

This report describes a case of retinal racemose hemangioma that first presented as a vitreous hemorrhage. The authors present the case of a 47-year-old woman with a sudden 5-day painless visual loss in her left eye. At the first visit, the best-correct visual acuities were 20/20 in the right eye and hand motions in the left eyes. Ultrasonography showed an attached retina and a massive vitreous hemorrhage. Pars plana vitrectomy was performed and a dilatation of large vessels was detected bulging from the optic disc. The best-correct visual acuities on day 30 postoperatively was 20/25 in the left eye. Fundus angiography and spectral-domain optical coherence tomography angiography showed anomalous arteriovenous communications with no intervening capillaries. The diagnosis was racemose hemangioma, an arteriovenous malformation of group 2 retina based on the Archer classification.

RESUMO

Este relato descreve um caso de hemangioma racemoso da retina que se apresentou inicialmente como hemorragia vítrea. Os autores apresentam o caso de uma mulher de 47 anos com perda visual súbita e indolor 5 dias antes no olho esquerdo. Na primeira visita, a melhor acuidade visual corrigida foi de 20/20 no olho direito e movimentos das mãos no olho esquerdo. A ultrassonografia mostrou uma retina aderida e uma hemorragia vítrea maciça. Foi realizada vitrectomia *pars plana*, sendo detectada proliferação de grandes vasos salientes do disco óptico. A acuidade visual no dia 30 de pós-operatório foi de 20/25 no olho esquerdo. A angiografia de retina e a angiotomografia de coerência óptica de domínio espectral mostraram comunicações arteriovenosas anômalas sem capilares intermediários. O diagnóstico foi hemangioma racemoso, uma malformação arteriovenosa da retina do grupo 2 com base na classificação de Archer.

INTRODUCTION

Racemose hemangioma of the retina or arteriovenous malformation of the retina is a nonhereditary disorder that presents with multiple other arteriovenous malformations that predominantly affect the face and brain. The disorder, also referred to as the Bonnet-Dechaume-Blanc syndrome, is classified as a nonhereditary congenital phakomatosis.[1] Bonnet, in 1937, and Wyburn-Mason, in 1943, reported the vascular abnormalities associated with this condition, and the syndrome is referred to frequently as Wyburn-Mason syndrome. The disease consists of an abnormal communication between the arterial and venous circulations with no intermediary capillaries. It can vary widely from barely visible connections to an extensive vascular network of enlarged vessels that resembles a bag of worms.[1,2]

In 1973, Archer et al.[2] classified this condition according to three distinctive groups. Group 1 is characterized by an abnormal capillary communicating with the circulations that is usually isolated to one ocular quadrant and difficult to visualize. Group 2 is comprised of communications without intervening capillaries that can be either medium or small in size. Group 3 includes large and complex vascular communications that typically compromise visual acuity (vessels like bag of worms).

This study was approved by the local committee at Faculdade de Medicina de Jundiaí, Brazil, as well as the consent form (CAEE 30925020.9.0000.5412).

CASE REPORT

A 47-year-old woman complained of sudden visual loss in her left eye with 5-day duration. She described the episode as spontaneous and painless with no other symptoms. She denied any previous ophthalmologic disorder. Her medical history was unremarkable and she also denied any comorbidities.

On examination, the best-corrected visual acuity (BCVA) was 20/20 in the right eye and hand motions in the left eye. Biomicroscopy findings and the intraocular pressure values were normal bilaterally. Fundus examination of the right eye showed no abnormalities. In the left eye, the Fundus was not visualized due to a massive vitreous hemorrhage.

Ocular ultrasonography of the left eye showed an attached retina, and no other abnormalities, except for the diffuse punctate echogenicity, was diagnosed as vitreous hemorrhage. An exploratory PPV was proposed in order to clean the vitreous cavity, improve vision, and possibly determine the etiology.

The surgery was uneventful. After cleaning the vitreous and removing the posterior hyaloid, a dilatation of large vessels was detected bulging from the optic disc. No active bleeding was observed. On day 30 postoperatively, the BCVA improved from hand motion to 20/25 in the left eye (Figure 1). Fundus color and fluorescein angiography (FA) showed anomalous arteriovenous communications with no intervening capillaries; and the later phases showed no leakage (Figure 2). OCTA visualized the arteriovenous communication that appeared as a protuberance on the optic disc and the absence of capillaries (Figure 3).

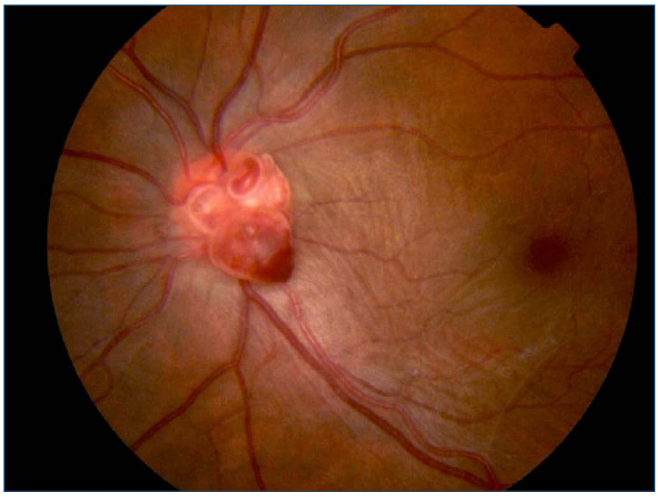

Figure 1. A color fundus retinography of the left eye on day 30 after PPV. An arteriovenous malformation is seen bulging from the optic disc.

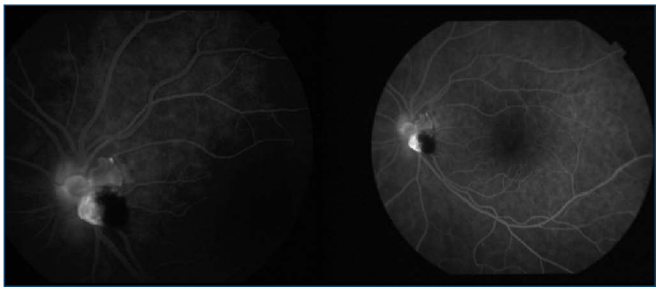

Figure 2. FA shows arteriovenous communications bulging from the optic disc with no capillaries intervening and no leakage during the late phases.

The diagnosis was hypothesized to be a racemose hemangioma, a group 2retinal arteriovenous malformation according to the Archer classification. To rule out the Wyburn-Mason syndrome, the patient was screened and referred to the Neurology Department and general specialists. The results showed no systemic abnormalities related to vascular malformations; magnetic resonance

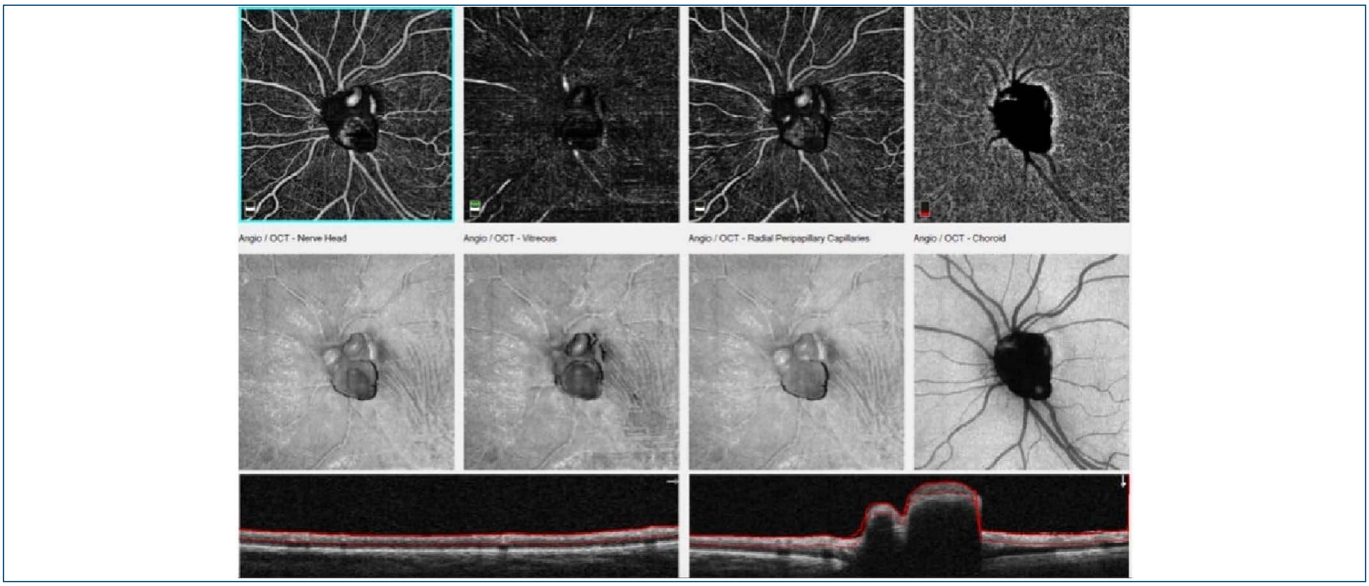

Figure 3. OCT angiograms of the optic nerve show dilated vasculature with no intervening capillaries decorrelation signals throughout different segmentations, suggesting direct arteriovenous communication.

imaging of the central nervous system showed no abnormalities. No hemorrhages have recurred over the subsequent 24 months.

DISCUSSION

The present study reported a racemose hemangioma in which the first finding was massive vitreous hemorrhage in a previously asymptomatic patient. Unlike previous reports, our diagnosis was established after vitreous surgery.

Using multimodal analyses, we found an anomalous protrusion of arteriovenous communication without intermediate capillaries: We performed an OCT B-scan, optic nerve angiograms and analyses with OCTA of signs of decorrelation along the different segmentations. With these data, it was possible to classify the findings as an isolated AVM of group 2 according to Archer's classification.[2]

A review of the literature showed that vitreous hemorrhages in anomalous arteriovenous communications are reported, as well as other complications such as rhegmatogenous retinal detachment,[3] but none of these cases of isolated vitreous hemorrhage were treated surgically, as the vitreous hemorrhage described was not massive and may resolve spontaneously.[4-10] In the present report, Pars Plana Vitrectomy was performed and allowed post-surgical observation and diagnosis.

In the current report of anomalous arteriovenous communication, group 2 according to Archer's classification, we emphasized that the first symptom was visual loss due to a vitreous hemorrhage, and after a favorable postoperative recovery, the diagnosis was established using multimodal analysis. The search for systemic pathologies did not identify other vascular anomalies. The important thing is that AVM of the retina without macular involvement can remain silent for extended periods and a vitreous hemorrhage can be the first manifestation. We suggest that this may be another pathology to be considered in cases of vitreous hemorrhage with unknown cause. Unfortunately, no treatment is available for this vascular anomaly, and dangerous vascular lesions can develop throughout life, making multidisciplinary systemic follow-up crucial after an ophthalmologic diagnosis.

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