DOI 10.5935/0034-7280.20200059

Choroidal hemangioma and the challenge of differential diagnosis

Hemangioma de coroide e o desafio do diagnóstico diferencial

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Abstract

Choroidal hemangioma is a fairly rare benign vascular tumor that can manifest in either circumscribed or diffuse type; the latter one is usually related to Sturge-Weber Syndrome. The circumscribed tumors have an insidious presentation and diagnosis is commonly made after the onset of secondary symptoms. Serious and potentially lethal lesions, such as choroidal melanoma and metastatic disease, may represent a differential diagnosis. In this report, we describe an advanced case of nodular hemangioma associated with hemorrhagic retinal detachment. This case highlights the challenge of differential diagnosis in intraocular tumors, due to their similar clinical and radiologic features.

Keyword: *Hemangioma; Choroidal neoplasms; Eye neoplasms; Benign tumor; Vascular tumor*

Resumo

O hemangioma de coroide é um tumor benigno relativamente raro, que se apresenta de forma circunscrita ou difusa, sendo esta última normalmente associada à Síndrome de Sturge-Weber. Os tumores circunscritos manifestam-se de forma insidiosa, com o diagnóstico realizado comumente após o aparecimento de sintomas secundários. Apresentam como diagnóstico diferencial lesões graves e potencialmente letais, como melanoma de coroide e doença metastática. Neste relato descrevemos o caso de um hemangioma intraocular nodular avançado associado a descolamento hemorrágico da retina, evidenciando o desafio do diagnóstico diferencial devido às semelhanças clínicas e radiológicas compartilhadas pelos tumores.

Descritores: Hemangioma; Neoplasias da coroide; Neoplasias oculares; Tumor benigno; Tumor vascular

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The authors declare no conflict of interest

Received for publication 6/8/2019 - Accepted for publication 6/10/2019.

Introduction

Choroidal hemangioma is an unusual benign vascular tumor (1 case for every 40 choroidal melanomas)[1] composed of large dilated choroidal vessels. It can occur in two different clinical and histopathological forms: as circumscribed tumor without extraocular associations, or in a diffuse way, when it is related to variations of the Sturge-Weber Syndrome.[2] The first form of it emerges from the second to the fourth decade of life, as red-orange well-circumscribed sessile elevation in the deeper layers of the retina. In most cases (95%), it is located posterior to the equator (most often in the macula or optic papilla) and can lead to exudative detachment of the retina, which leads to low visual acuity (LVA) when it affects the macular region.[3] On the other hand, the diffuse type is often diagnosed at birth as the manifestation of Sturge-Weber Syndrome - clinically, it appears as a poorly-defined red mass surrounding more than half of the choroid.[4] Caucasian patients account for more than 90% of the reported cases, but there are no difference regarding sex.[5]

Histologically, eye lesions are classified based on the type of vessels inside the tumor, including cavernous, capillary or mixed types. The cavernous type comprises large vessels separated by circumscribed connective tissue. The capillary type encompasses small-caliber vessels separated by loose connective tissue. The mixed type presents cavernous and capillary features. The cavernous and mixed types were the most prevalent forms of it in a series of circumscribed choroidal hemangiomas cases. The capillary type was observed in only a small number of cases. All diffuse hemangioma cases associated with Sturge-Weber Syndrome were of the mixed type.[6,7]

Diagnosis consists of biomicroscopic examination and of several complementary exams, such as ultrasound (US), angiofluoresceinography, magnetic resonance imaging (MRI) and optical coherence tomography (OCT); together they help differentiating choroidal hemangioma from other tumors.[8] Chordal hemangioma has a characteristic ultrasonographic pattern, it appears in the two-dimensional mode (B-scan) as a solid, high, dome-shaped acoustic mass, whereas, in the amplitude mode (A-scan), it is a high initial peak that corresponds to the surface of the anterior tumor. It shows high internal reflectivity (between 50% and 100%) due to several vascular channels within these tumors. Such features are important because they help differentiating choroidal hemangioma from choroidal melanoma, which is often acoustically concave and present medium to low internal reflection.[9,10] Choroidal hemangioma usually shows hyperintense signal in comparison to the vitreous in T1-weighted images, as well as hyperintense signal or isointensity in T2-weighted images; moreover, it shows important enhancement after paramagnetic agent (gadolinium) administration. Such findings are useful to differentiate choroidal hemangioma from choroidal melanoma, and from metastases evidencing high signal in T1-weighted images and low signal in T2-weighted images.[5,6] However, these features are not pathognomonic for choroidal hemangiomas; they have been found in some choroidal melanomas, and it makes differential diagnosis difficult.(10) Diagnostic difficulty can be seen in studies that have shown that 5% - 10% of eyes enucleated as choroidal melanoma had choroidal hemangioma.[11,12]

Treatment in asymptomatic cases that do not have subretinal fluid can be the simple observation of the lesion. However, different treatments, such as photodynamic therapy (PDT), plaque brachytherapy, external beam radiotherapy, stereotactic radiosurgery, transpupillary thermotherapy, laser photocoagulation, oral administration of propranolol and intravitreal antiangiogenic therapy are recommended for symptomatic lesions with the risk of visual impairment. Enucleation can be necessary in more advanced cases evidencing visual loss and neovascular glaucoma.[6]

A case of advanced nodular intraocular hemangioma, with hemorrhagic detachment of the retina and difficult diagnosis through imaging exams was herein reported.

Case Report

Male patient, aged 43 years, reported progressive LVA in the left eye (LE) for 1 year. It was related to moderate episodes of eye pain and ipsilateral conjunctival hyperemia, with sudden pain worsening and visual acuity for 1 week. The patient did not have other comorbidities or family history of eye disorders.

The ophthalmologic examination showed right eye (RE) visual acuity of 20/25 and absence of light perception in the LE. Biomicroscopy evidenced regular RE, with regular intraocular pressure and retinal mapping. LE had conjunctival hyperemia 1+/4, cornea with mild edema and iris rubeosis, in addition to intraocular pressure of 50 mmHg and the presence of a thin and mobile membrane on the anterior vitreous, which was compatible to detached retina (Figure1). LE fundoscopy showed dense vitreous hemorrhage, which made it impossible seeing the intraocular structures. LE US reported the presence of a high-reflectivity mobile membrane on the vitreous cavity, which suggested total detachment of the retina and the presence of an expansive, hyperechogenic solid dome-shaped lesion in the nasal region (9.1mm in height and 10.3 mm in anteroposterior length) (Figure 2). Systemic laboratory and imaging tests were requested for screening purposes; they showed normal results, MRI of the skull and orbits were requested to support the first diagnosis; MRI showed an expansive oval formation affecting the chorioretina of the left eye: hyposignal at T1, intense homogeneous impregnation by gadolinium (defined limits - 0.8 x 0.9 x 04 cm) and no signs of extraocular extension associated with massive retinal detachment; it presented high signal content at T1 (probable hematic content). Neoplastic lesions was one posssible diagnosis, including choroidal amelanotic melanoma or secondary lesions in the diagnosis (Figure 3).

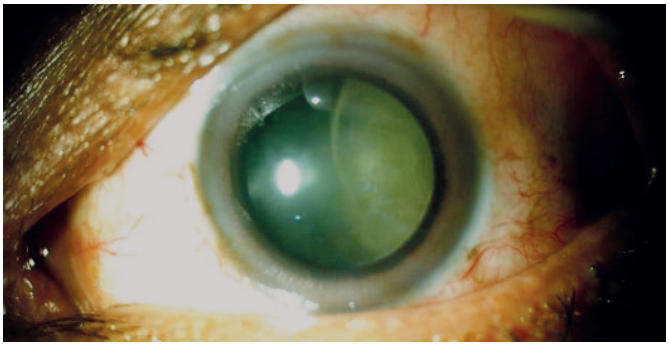

Figure 1: Biomicroscopy of the left eye showing anterior vitreous detached retina

Enucleation of the eyeball was chosen because the lesion likely had neoplastic origin and because there was low visual prognosis. The anatomopathological examination revealed expansive lesion with several vessels (different calibers) (Figure 4). The patient was instructed about his condition and referred for adaptation to ocular prosthesis.

Discussion

Choroidal hemangioma is a rare intraocular tumor; however, it is important differentiating it from other intraocular tumors. Choroidal hemangioma diagnosis is challenging, oftentimes patients have initial choroidal melanoma or metastatic lesion diagnoses.[13] Auxiliary exams, such as US and MRI, help differentiating

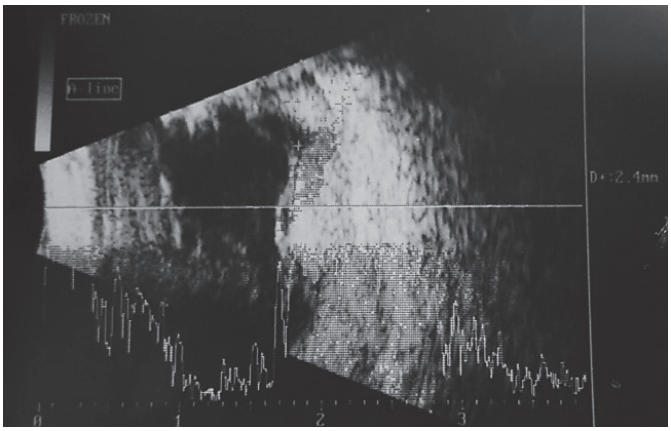

Figure 2: Ultrasound of the left eye showing hyperechogenic dome-shaped solid lesion.

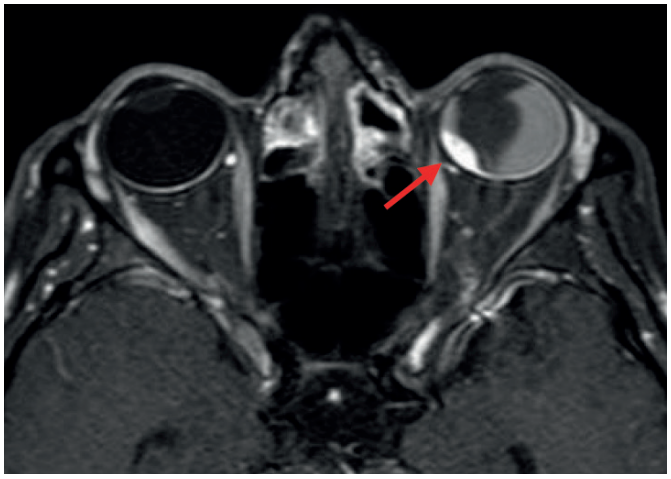

Figure 3: Magnetic resonance imaging of the orbits, in post-gadolinium T1 sequence, showing expansive lesion with intense contrast impregnation in the medial aspect of the choroid to the left (red arrow). It is related to large hemorrhagic retinal detachment to the same side, characterized by slight hypersignal in T1

this tumor from similar injuries; however, it is challenging to have an accurate diagnosis due to clinical and radiological similarities shared by such lesions.[14]

Peculiarities of the herein reported case come together because, in addition to present a more aggressive lesion – that has evolved to amaurosis, total retinal detachment and neovascular glaucoma (unlike most cases, which have more indolent evolution) -, it was also not possible stating that it was not a malignant tumor based on imaging exams.

Enucleation of the eyeball and ocular prosthesis were chosen due to the impossibility of excluding the possibility of a malignant lesion, as well as to the low visual prognosis and eye pain. Currently, the patient is clinically well and reports adequate post-operative and aesthetic adaptation, without any complaint.

Histopathological analysis was compatible to capillary hemangioma of the tunica media, and this finding evidenced another singularity of this case, since cavernous and mixed are the most common histological types of nodular hemangiomas. According to Witschel and Font, the capillary type represents only a small percentage of cases (3%).[7]

The reported case is important because it can shine light and guide professionals on investigations and on the difficulties reaching to accurate choroidal hemangioma diagnosis due to it different clinical and radiological aspects. Therefore, the reported case helps ophthalmologists managing such challenging cases.

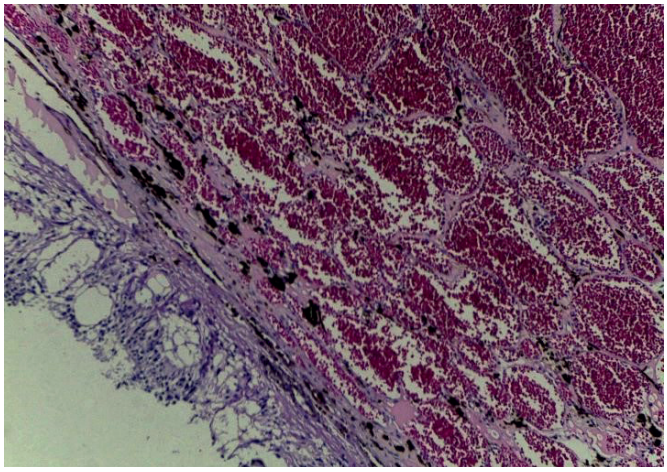

Figure 4: Histological study showing a lesion with several vessels (different calibers), which confirmed the diagnosis of capillary hemangioma.

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