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REVIEW

Circumscribed choroidal hemangioma: A case report and literature review

Morgan Berry, Linda J.H. Lucas*

Marion VA Medical Center, Marion, IL, USA

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KEYWORDS

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PALABRAS CLAVE

Hemangioma circunscrito de coroides;
Ultrasonografía;
Terapia fotodinámica;
Melanoma coroidal

Abstract Choroidal hemangioma is a rare congenital ocular tumor that can present as either circumscribed or diffuse. Circumscribed choroidal hemangioma (CCH) typically manifests as a red-orange mass within the posterior pole and appears similar to other ocular conditions, such as choroidal melanoma and choroidal metastasis. Proper diagnosis is crucial and is aided by the use of ancillary testing. CCH itself is benign but can cause secondary complications such as subretinal fluid accumulation and subsequent retinal detachment. If these conditions should arise, several treatment options are available.

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Hemangioma circunscrito de coroides: informe de un caso y revisión de la literatura

Resumen El hemangioma coroidal es un raro tumor ocular congénito que puede presentarse de manera circunscrita o difusa. El hemangioma circunscrito de coroides (CCH) se manifiesta típicamente como una masa rojizo-anaranjada en el interior del polo posterior, y parece similar a otras situaciones oculares, tales como el melanoma coroidal y la metástasis coroidal. El diagnóstico adecuado es esencial, y viene asistido por el uso de pruebas complementarias. El CCH en sí mismo es benigno, aunque puede originar complicaciones secundarias tales como la acumulación de fluidos sub-retinianos y el subsiguiente desprendimiento de retina. En caso de producirse dichas situaciones, existen diversas opciones de tratamiento.

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* Corresponding author at: VA Medical Center, 2401 W Main St, Marion, IL 62959, USA.
E-mail addresses: Linda.Lucas2@va.gov, jalismom@hotmail.com (L.J.H. Lucas).

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Choroidal hemangioma is a benign vascular tumor.^{1,2} It is considered a congenital condition but can continue to develop into early adulthood.^{2,3} Choroidal hemangiomas are classified into two subtypes, circumscribed and diffuse, a distinction made based on the extent of choroidal involvement. Diffuse choroidal hemangiomas involve more than one quadrant of the choroid and are typically associated with systemic conditions such as Sturge–Weber syndrome.^{1–4} Circumscribed choroidal hemangioma (CCH) typically presents as a unilateral orange-red choroidal mass without correlation to systemic conditions. CCH may appear similar to other choroidal tumors, some of which are malignant, such as choroidal melanoma and choroidal metastasis.^{2,3} For this reason, it is important to be able to differentiate between these lesions. This can be accomplished by employing ancillary testing such as ultrasonography, fluorescein angiography (FA), indocyanine green angiography (ICGA), magnetic resonance imaging (MRI), and optical coherence tomography (OCT).^{2,3} Once the diagnosis has been made, referral to a retinal specialist is usually warranted. Treatment options include laser photocoagulation, thermotherapy, radiotherapy, photodynamic therapy (PDT), and anti-vascular endothelial growth factor (VEGF) therapy.^{1,5–9}

Case report

Relevant history

A 49-year-old Caucasian male presented to clinic with complaints of blurred vision at distance that was worse in his left eye. His medical history included hypertension, hyperlipidemia, sleep apnea, and post-traumatic stress disorder. His ocular history was unremarkable.

Clinical findings

The patient's best-corrected visual acuities (BVA) were 20/20 OD and 20/100 OS, and he consistently missed letters on the left side of the chart with his left eye. Pupils were equal, round, and reactive to light OU, and there was no afferent pupillary defect. Confrontation fields were full, and extra-ocular muscles had full range of motion. Amsler grid was unremarkable OD and revealed left-sided metamorphopsia OS. Slit lamp examination was unremarkable OU, and intraocular pressures were within normal limits. Dilated fundus exam of the left eye (Fig. 1) revealed a 2DD orange-red elevated lesion superior temporal to the optic nerve head with overlying retinal pigment epithelium change and pigmentation. An associated 3–4 DD central serous retinal detachment was also noted. Several ancillary tests were performed. Ultrasonography showed a 2 mm × 8 mm solid choroidal mass on B-scan with high internal reflectivity on A-scan. OCT revealed a large choroidal mass with overlying subretinal fluid. Fundus photos were also taken.

Management/follow-up

The patient was diagnosed with CCH and referred to a retinal specialist. Fluorescein angiography was performed, and it showed hyperfluorescence in all stages. The lesion was treated with argon laser photocoagulation over the entire



Figure 1 Fundus photo shows a circumscribed choroidal hemangioma superior temporal to the optic nerve in the left eye.

mass. The patient was seen two weeks later, and BVA was 20/50 OS. Dilated fundus exam revealed an improvement in subretinal fluid, but a serous detachment was still present. FA was repeated as well as another session of argon laser photocoagulation.

Over the next 15 years, the patient was seen every three to six months depending on the status of the condition. After three treatments of argon laser photocoagulation, all of the subretinal fluid resolved. The CCH did not shrink in size. Pigment later developed over the lesion. BVA remained around 20/30 at each follow-up visit. The subretinal fluid eventually returned 11 years after the last treatment session, but it did not affect the patient's vision. Therefore, observation was chosen as the treatment at that time. No changes in the amount of subretinal fluid or BVA have been noted at any follow-up visits since.

Literature review

Background

CCH is a rare vascular tumor.^{1,3} It is considered to be congenital in nature; however, it often does not cause symptoms until the fourth to sixth decade of life.^{2,3} It is observed more frequently in Caucasians and equally between the sexes. CCH is a very uncommon tumor that is likely to go undiagnosed unless the patient becomes symptomatic.

Unlike diffuse choroidal hemangiomas, which are associated with phakomatoses such as Sturge–Weber syndrome, CCH has no correlation with systemic disease.^{3,4} CCH is composed of dilated choroidal vessels but does not involve the choriocapillaris. The specific cause of the blood vessel malformation is unknown. CCH is different from other “true hemangiomas” in that it does not show cellular proliferation. In other words, it shows slow to no progression in size over time.^{2,3}

CCH presents clinically as an orange-red mass in the posterior pole, usually within one to three disk diameters of the macula.^{2–4} They typically range in size from 3 to 19 mm in diameter and 1 to 8 mm in thickness. The

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