Retinal Racemose Hemangioma

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Case Presentation:

A 15-year-old male presented to clinic with complaints of central blurry vision in the right eye. Symptoms began one week prior and did not improve. He denied any other ocular or systemic review of symptoms. He had no significant ocular history. His past medical history consisted of anxiety and allergies treated with Concerta and Singular. His visual acuity was 20/80 in the right eye and 20/20 in the left eye. His intraocular pressures were 16 in both eyes. Pupils were equally round and reactive to light without afferent pupillary defect. Confrontational visual fields, extraocular movements, and anterior segment examination were normal. Posterior examination demonstrated preretinal foveal hemorrhage in the right eye. (Figure 1) Additionally there was dilated tortuous anomalous vasculature originating off the optic disc. The peripheral examination was found to be normal. The left eye funduscopic examination was normal. Optical coherence tomography of the right eye confirmed preretinal hemorrhage in the macula without subretinal fluid or edema and normal retinal anatomy in the left eye.

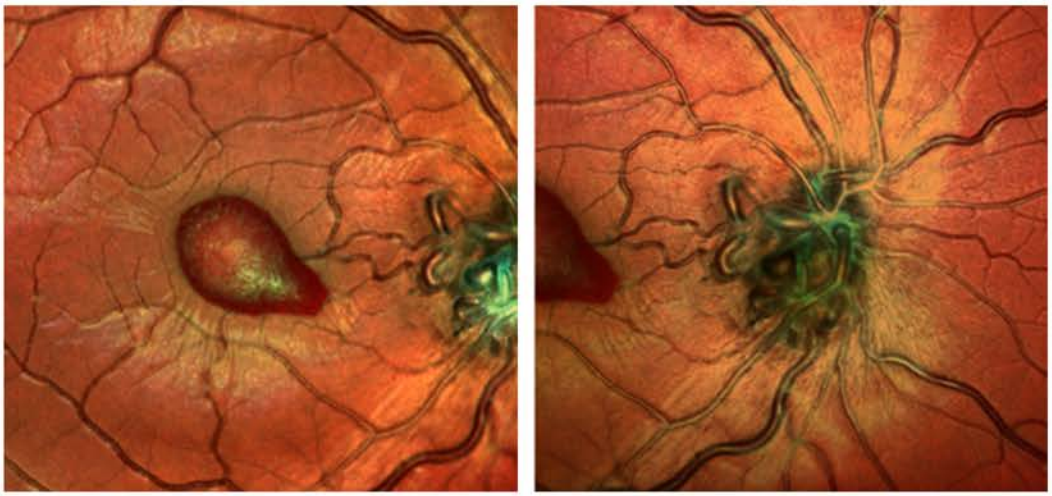

Figure 1A (left) – Fundus photo demonstrating preretinal hemorrhage.
Figure 1B (right) – Disc photo demonstrating dilated anomalous vasculature.

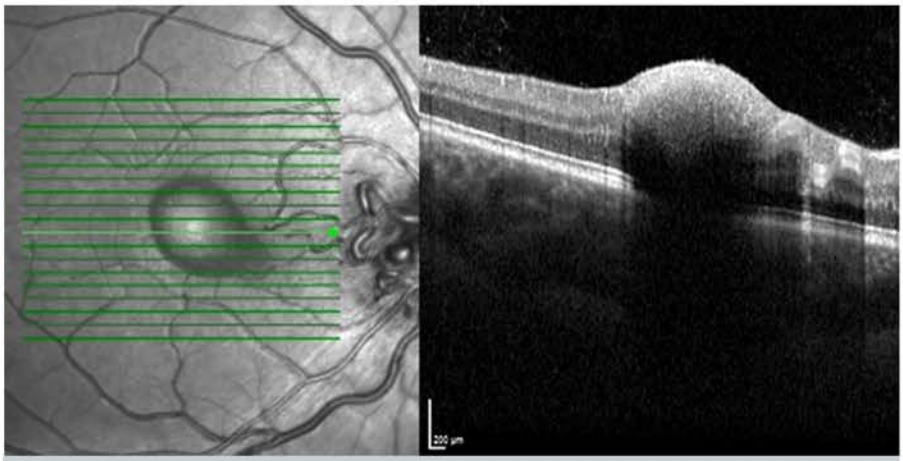

Figure 2 – OCT of the right eye showing preretinal hemorrhage on presentation.

(Figure 2) Fluorescein angiography of the right eye demonstrated arteriovenous communications without leakage and blockage from the preretinal hemorrhage (Figure 3).

Differential Diagnosis:

The differential diagnosis for anomalous arteriovenous malformations is relatively short. It can present as an isolated congenital arteriovenous malformation without any other lesions or associations. When the lesion is specific of the fundus vessels it as known as a retinal racemose hemangioma. When these findings are associated with systemic malformations as well,

this combination is known as Wyburn-Mason syndrome, or Bonnet-Dechaume-Blanc syndrome. Other vascular lesions that can mimic an AVM include hemangiomas and hemangioblastomas.

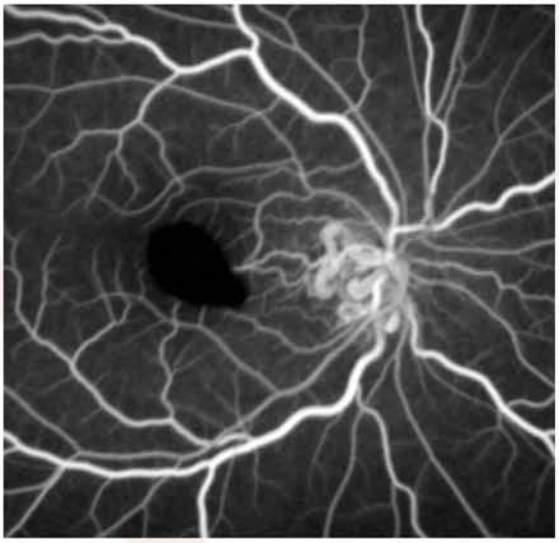

Figure 3 – FA of the right eye demonstrating anomalous vasculature without leakage.

Discussion:

Congenital arteriovenous malformations are rare. Retinal racemose hemangioma is generally non-hereditary without gender or race predilection and usually present unilaterally. Presentation can vary widely from subtle capillary changes to anastomoses involving large portions of the fundus. The most common complication is retinal vein occlusion and vitreous hemorrhage which make up 45% and 33% of the total complications. It has been theorized that the high flow into the veins cause endothelial damage leading to thrombosis.

About 30% of patients with racemose hemangiomas can have cerebral arteriovenous malformations in the brain, a condition known as Wyburn Mason Syndrome. These cerebral hemangiomas can cause severe malformations anywhere along the visual axis from the optic nerve to the midbrain to the occipital lobes. Other areas of involvement include the mandible, maxilla, and facial bones presenting as gingival bleeding and epistaxis. Rupture of intracranial lesions can lead to devastating fatal hemorrhagic strokes therefore it is important to screen all patients with AV malformations with an MRI as intracranial lesions can be treated with radiosurgery or embolization. Retinal lesions are non-progressive and rarely require treatment, but cryotherapy and laser have been used.

Our patient underwent an MRI brain which demonstrated no lesions suspicious for vascular malformations. After two months, visual acuity improved to 20/40 with near complete resolution of the intraretinal and preretinal hemorrhage. (Figure 4)

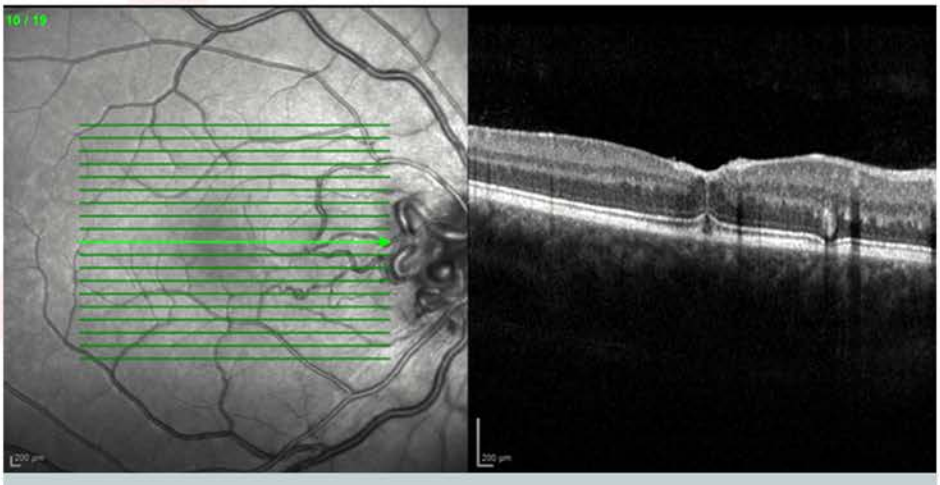

Figure 4- OCT of the right eye demonstrating resolution of the hemorrhage with inner and outer retinal changes after 2 months.

References:

Xue-jiao Qin, Chao Huang and Kun Lai. Retinal vein occlusion in retinal racemose hemangioma: a case report and literature review of ocular complications in this rare retinal vascular disorder. BMC Ophthalmology 2014 14:101.

J.J. Bhattacharya, C.B. Luo, D.C. Suh, H. Alvarez, G. Rodesch, and P. Lasjaunias. Wyburn-Mason or Bonnet-Dechaume-Blanc as Cerebrofacial Arteriovenous Metameric Syndromes (CAMS) A New Concept and a New Classification Interv Neuroradiol. 2001 Mar; 7(1): 5–17.

Shields CL, Shields JA. Phakomatoses. In: Ryan SJ, Schachat AP, Murphy RP (eds). Retina, Vol. III: Section 1, Chapter 132. Tumors of the Retina. 5th ed. St. Louis: Mosby; 2013:2170-83.

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