



Case report

Retinal racemose hemangioma with retinal artery macroaneurysm: Optical coherence tomography angiography (OCTA) findings

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ABSTRACT

Purpose: To describe a rare case of retinal racemose hemangioma (RRH) with retinal artery macroaneurysm (RAM) and its optical coherence tomography angiography (OCT-A) finding before and after treatment.

Observations: Congenital arteriovenous (AV) communications or RRH is a rare developmental anomaly associated with various ocular conditions. RRH alone is usually asymptomatic, and vision loss occurs when associated with other ocular complications like retinal vein occlusion, hemorrhage, macular edema and macroaneurysm. In this communication we describe a case of a 44-year-old female having RRH who presented with active, leaking RAM with subretinal hemorrhage. OCT-A through the lesion demonstrated an active aneurysm which on subsequent treatment with anti VEGF and focal laser photocoagulation showed involution of aneurysm.

Conclusions and importance: RRH is usually asymptomatic and may be associated with vision threatening ocular complications like RAM. OCT-A helps in giving additional dynamic blood flow information in RAM.

1. Introduction

Congenital arteriovenous communications or retinal racemose hemangioma (RRH) is a rare, unilateral, non hereditary and sporadic phacomatosis characterized by the appearance of dilated and tortuous retinal vessels frequently extending from the optic disc to the retinal periphery.¹ Archer et al.² have divided these AV communications into 3 groups depending on the size and location of vascular malformation. The first group is characterized by abnormal capillary plexus between the major vessels of the AV malformations and is usually asymptomatic. The second group is characterized by AV malformations and absence of any intervening capillary bed between artery and vein. These are usually associated with retinal complications due to retinal decompensation and have low risk for intracranial AV malformations. The third group is characterized by extensive AV malformations with dilated and tortuous vessels and no distinction between artery and vein. There is a high risk for vision loss due to retinal decompensation leading to complications like retinal vein occlusion, vitreous hemorrhage, macular edema, macroaneurysms and are associated with intracranial AV malformations.

Retinal arterial macroaneurysm (RAM) is an acquired, isolated dilatation of a retinal arteriole usually associated with hemorrhage, edema, and exudation. High risk factors include older age, woman

gender, systemic hypertension and arteriosclerotic disease.³ Although most of the RAMs may involute spontaneously, treatment may be needed in patients with associated macular edema, retinal/sub retinal hemorrhage or exudation. In this communication we describe a rare combination of RRH with RAM; we also describe the optical coherence tomography angiography (OCT-A) features before and after treatment.

2. Case report

A 44-year old female presented with painless sudden diminution of vision in her left eye since 5 days. Her best-corrected visual acuity was 20/20, N6 in the right eye and 20/30, N6 in the left eye. Both eyes anterior segment was within normal limits. Left eye fundus showed multiple dilated and tortuous retinal vessels emerging from the optic disc and extending along the supero-temporal arcade with no distinction between the artery and the vein (Fig. 1A). There was a small RAM with surrounding yellowish subretinal hemorrhage in the supero-temporal quadrant (Fig. 1A, arrowhead). Fundus fluorescein angiography (FFA) in the late phase showed blocked fluorescence corresponding to the subretinal hemorrhage with a central focal leakage corresponding to the fusiform leaking aneurysm. In the temporal periphery communication of the arteriovenous malformations was noted without any intervening capillary bed. The optical coherence tomography (OCT)

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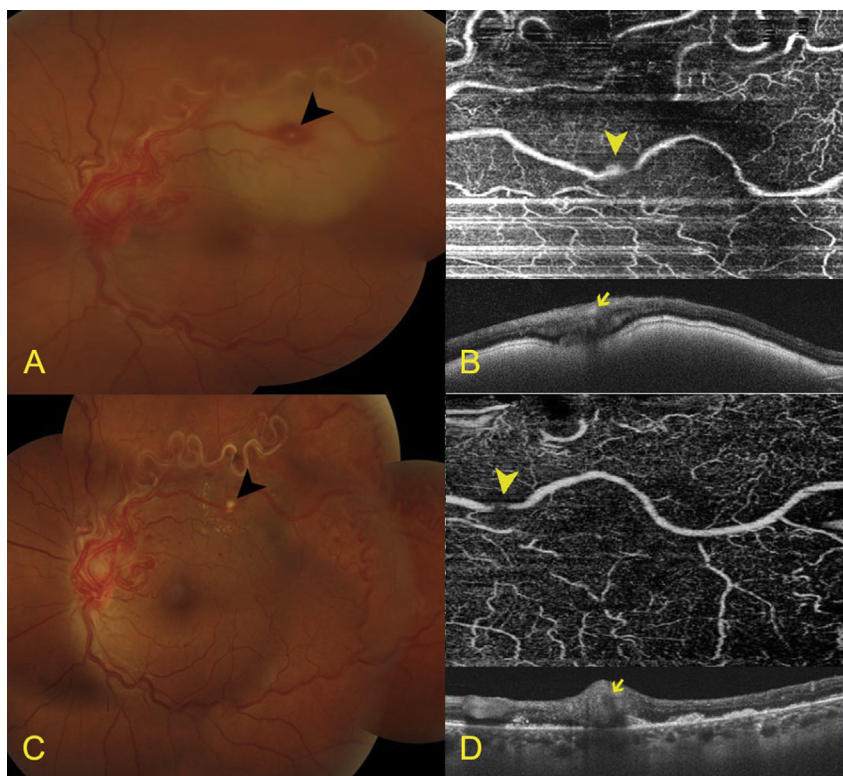


Fig. 1. A. Colour fundus photograph of the left eye: Dilated and tortuous arteriovenous malformations (retinal racemose hemangioma) in the superotemporal quadrant with retinal artery macroaneurysm (RAM, black arrowhead) and sub-retinal hemorrhage.

B. Top OCT-A in the superficial slab showing active RAM lesion with increased blood flow (yellow arrowhead), Bottom OCT shows intact inner retinal layers with hyperreflectivity within the lumen of artery (yellow arrow).

C. Colour fundus photograph of the left eye following treatment showing involution of aneurysm (black arrowhead) and resolution of hemorrhage.

D. Top OCT-A in the superficial slab showing involuted RAM lesion with decreased blood flow (yellow arrowhead), Bottom OCT shows hyporeflectivity within the lumen of artery (yellow arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

scan through the disc showed multiple large intraretinal vessels with back shadowing and without associated macular edema. OCT through the RAM lesion showed intact inner retinal layers with hyperreflectivity within the lumen of vessel with back shadowing suggestive of active leaking aneurysm (Fig. 1B, arrow). OCT-A of the disc showed dilated and tortuous vessels in the superficial and deep retinal layers and increased blood flow through the well-delineated RAM lesion in superficial retina (Fig. 1B, arrowhead). On the basis of FFA, OCT and OCT-A a diagnosis of type 3 RRH with leaking RAM in the left eye was made. An intravitreal injection of bevacizumab (1.25 mg in 0.05 mL) was administered. The vision at 1 month improved to 20/20, N6 with resolution of subretinal hemorrhage. In view of leaking aneurysm, focal laser (double frequency ND-YAG; power-100 mw, energy-80J and spots-25) was applied directly to the aneurysm and the area surrounding it. At the last follow up (5 months), vision remained stable at 20/20, N6 with involution of aneurysm and resolution of subretinal hemorrhage (Fig. 1C, arrowhead). The OCT through the aneurysm showed hyporeflectivity within the lumen of the vessel (Fig. 1D, arrow) and OCT-A in the superficial layer showed reduced flow signal through the RAM lesion (Fig. 1D, arrowhead). Systemic examination was unremarkable and ruled out risk factors like hypertension and atherosclerotic disease; computed tomography scan of the brain ruled out any intracranial arteriovenous malformations.

3. Discussion

Congenital arteriovenous communications are rare, unilateral developmental anomalies of the vascular network.^{4–6} Most of the cases are asymptomatic. The cause for loss of vision is due to late ocular complications like retinal vein occlusion (45%), hemorrhage (33%), macular edema or rare congenital vascular malformations like morning glory disc anomaly, Sturge-Weber syndrome and macroaneurysm.¹

Our patient had a rare combination of unilateral type 3 RRH, RAM with presence of active leaking aneurysm and subretinal hemorrhage. Various hypothesis postulated related to development of ocular complications associated with AV malformations are hyperdynamic blood

flow through small-caliber vessels, and steal phenomenon caused by increased venous pressure with decreased arterial pressure.⁷

Currently FFA and OCT are used in the diagnosis and follow-up of patients with RAM.^{8–10} The FFA produces a characteristic late leakage within the areas of hemorrhages. The OCT-A helps visualization of the inner and outer retinal vascular plexus and the choriocapillary layer.¹¹ We also observed the ability of OCT-A demonstrating the reduction in flow signals through the RAM lesion before and after treatment, thus indicating involution of aneurysm.

In most instances RAMs involute naturally.¹² Treatment is indicated in instances of leaking aneurysms, extensive hemorrhage and macular involvement threatening vision.¹³ Pichi F et al.¹³ have reported the benefit of intravitreal bevacizumab in improving the vision and reducing the macular edema in complicated RAM. We decided for initial intravitreal anti VEGF injection in our case as presence of large subretinal hemorrhage prevented direct laser to aneurysm. Once the hemorrhage cleared, a direct focal laser was done for complete regression of aneurysm and faster recovery of visual acuity.

4. Conclusion

RRH is usually asymptomatic and may be associated with vision threatening ocular complications like RAM. OCT-A helps in giving additional dynamic blood flow information in RAM. Possibly the OCT-A could be used as an alternative non-invasive tool for understanding the morphology and activity of RAM.

Patient consent

Written consent to publish case details obtained from the patient.

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Authorship

“All authors attest that they meet the current ICMJE criteria for Authorship”.

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