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Retinal racemose hemangioma presenting with a chorioretinal anastomosis

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ABSTRACT

Purpose: To report a case of neovascular glaucoma in an 8-year-old male, secondary to a racemose hemangioma without associated intracranial arteriovenous malformation, highlighting the challenges in management and novel findings on optical coherence tomography angiography (OCTA).

Observations: An 8-year-old male initially presented with pain, redness, and blurred vision in the right eye. The patient was diagnosed with secondary neovascular glaucoma due to a racemose hemangioma. Urgent interventions included intravitreal bevacizumab injection and tube shunt surgery for persistently high intraocular pressure. Pars plana vitrectomy and scatter laser photocoagulation were eventually performed to manage a tractional retinal detachment and peripheral ischemia, respectively. OCTA imaging revealed a racemose hemangioma with a unique chorioretinal anastomosis.

Conclusions and Importance: We present a rare pediatric case of neovascular glaucoma secondary to a racemose hemangioma with significant peripheral ischemia and an unusual chorioretinal anastomosis. The discovery of a chorioretinal anastomosis on OCTA suggests a potentially severe variant of racemose hemangioma.

1. Introduction

Racemose hemangioma (RH) of the retina, also known as a retinal arteriovenous malformation (AVM), is a rare, congenital vascular anomaly characterized by abnormal arteriovenous communications. Archer et al. described three groups of RHs: group 1, an artery and vein with an abnormal intervening capillary plexus; group 2, an artery and vein without an intervening capillary network; and group 3, complex and extensive large vessels without intervening capillaries.[1] RH can be associated with intracranial AVM's, termed Wyburn-Mason syndrome. The pathogenesis of Wyburn-Mason syndrome is presumed to be a local defect in the maturation of mesenchymal cells at the 7th week of gestation, affecting both the eye and the mesencephalon. Abnormal vascular development after 7 weeks gestation may result in an isolated RH without intracranial AVM.[2,3]

RH's can be asymptomatic or present with a range of complications, including retinal, vitreous, or subretinal hemorrhage, retinal vein occlusion (RVO), macular microaneurysms, and macular ischemia.[4] In severe cases, extensive retinal ischemia may develop, leading to neovascular glaucoma.[5] Three mechanisms for retinal ischemia in RH have been proposed. First, ischemia may result from a vascular steal phenomenon, where high flow through the RH draws critical blood flow away from retinal tissue. Second, partial thrombosis of the RH may occur, which would reduce flow to retinal tissue supplied by the RH. Finally, direct compression of venous outflow at the optic nerve head results in an RVO.

To our knowledge, involvement of the choroidal circulation by an RH has only been described by Stokes,[6] who observed, with meticulous indirect ophthalmoscopy, a diving artery and vein with a possible choroidal connection. In this article, we report a case of RH with several complications, including neovascular glaucoma, vitreous hemorrhage (VH), and tractional retinal detachment (TRD). Fundoscopic examination and fluorescein angiography showed evidence of peripheral ischemia and vein occlusion. Optical coherence tomography angiography (OCTA) showed a diving RH with an apparent connection to the choroidal circulation.

2. Case report

A healthy 8-year-old male presented to a local emergency room with progressively worsening pain, redness, and blurred vision in his right eye for three weeks. Ophthalmology reported a visual acuity (VA) of 20/200, a fixed and dilated pupil, relative afferent pupillary defect, and unreadable, high intraocular pressure (IOP) by handheld tonometry

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(Tono-Pen Indentation/Applanation Tonometer). The left eye was normal. There was a hazy view of the posterior segment with concern for vitritis and retinitis. The patient was transferred to our children's hospital, and initial management included intravenous acyclovir.

An examination under anesthesia (EUA) revealed an intraocular pressure of 55 mmHg by tonometry (Tono-Pen), and a handheld slit lamp examination revealed iris neovascularization. Handheld fundus photography revealed Frisen grade 4 optic disc edema and an Archer grade 2 superior-temporal RH extending from the optic disc into the macula (Fig. 1; camera: Retcam 3, Natus Medical, Inc., USA). There were scattered intraretinal hemorrhages both superior-temporally and directly inferior to the optic nerve head. Exudates were visible near the fovea, and venous sheathing was present in inferior vessels. Vitritis and retinitis were not present. Secondary neovascular glaucoma was diagnosed, and an intravitreal injection of bevacizumab was administered.

Within the next few days, a dense VH developed. IOP was persistently elevated at 40 mmHg, despite maximal topical therapy (atropine drops three times daily, iopidine drops three times daily, timolol twice daily, and latanoprost once at night), intravenous mannitol (1 gm per kg daily), and oral acetazolamide (5 mg per kg, three times daily). Urgent tube shunt surgery was performed with an Ahmed implant. Magnetic resonance angiography of the brain revealed no associated intracranial AVM. After the surgery, the patient was discharged with a VA of hand motion (HM) and IOP of 8 mmHg.

After two months, a pars plana vitrectomy was performed for non-clearing VH. A tractional retinal detachment above the optic nerve was released. A repeat EUA with fluorescein angiography revealed early arterial filling of the RH during the choroidal phase (Fig. 2a), and significant macular and superotemporal nonperfusion of the choroidal and retinal circulations (Fig. 2b, c, d; camera: Retcam 3, Natus Medical, Inc., USA). The mid phase of the FA showed diffuse areas of capillary leakage particulary in the inferior macula (Supplementary Fig. 1). Meticulous indirect ophthalmoscopy was performed, and the RH appeared to dive deep toward the choroid. This examination finding and the early filling of the RH during the choroidal phase of the FA led us to suspect a possible chorioretinal anastomosis (CRA). Intraoperatively, scatter laser photocoagulation was applied in the peripheral ischemic retina of the right eye. The left eye had normal perfusion and vasculature on both indirect ophthalmoscopy and fluorescein angiography.

Fig. 1. Handheld fundus photograph of the right eye during the first exam under anesthesia showed a dilated retinal artery and a vein exiting the superior temporal aspect of the optic disc. An anastomosis is present without an intervening capillary plexus, indicating an Archer grade 2 superior-temporal racemose hemangioma. Frisen grade 4 optic disc edema and intraretinal hemorrhages are present, as well as exudates in the fóvea. Vascular sheathing is also observed in the inferior arcade.

Ultra-wide field photography, performed on post-procedure day 1, showed laser burns in the temporal and supero-temporal periphery. (Fig. 3; camera: Ultra-wield field retinal image, Optos, Scotland). There was sheathing of vessels in the periphery of the supero-temporal quadrant. To investigate the possibility of a CRA, OCTA was performed (camera: Cirrus 6000 AngioPlex OCTA, Zeiss, Germany). The enface view of the superficial slab showed the RH exiting the superotemporal aspect of the optic disc and curving towards the fovea (Fig. 4a). The enface view of the outer-retina to choriocapillaris (ORCC) slab showed that the tip of the RH lied deep, where typically retinal vasculature would not be present (Fig. 4b, cyan and magenta crosshairs). An OCT cross-section with OCTA overlay in red showed thinning of the inner retinal layers, a break in Bruch's membrane, and the tip of the RH adjacent to the choroid (Fig. 4c, white arrow), confirming the CRA. A fly-through of the OCT and OCTA volume is provided (Supplementary Video 1). Finally, to better visualize the entire RH complex, we three-dimensionally rendered the OCTA volume using Imaris (Imaris Viewer 10.1.0, Oxford Instruments), as shown in Fig. 4d. The rendering enabled us to visualize the diving nature of this RH towards the choroid layer and the CRA.

Supplementary video related to this article can be found at https://doi.org/10.1016/j.ajoc.2024.102188

3. Discussion

We present a pediatric case of neovascular glaucoma secondary to RH with significant macular and peripheral ischemia. The peripheral ischemia likely resulted from a vascular steal phenomenon, whereas the macula non-perfusion likely resulted from a retinal vein occlusion. The associated optic nerve swelling and scattered intraretinal hemorrhages were also suggestive of RVO. An arterial thrombosis seemed unlikely as there was complete perfusion of the RH on fluorescein angiography. There was no associated intracranial AVM to suggest Wyburn-Mason syndrome.

Anti-VEGF agents for central RVO in RH have been used with demonstrated recovery of macular edema.[7] However, our case was unusually complicated by a non-clearing VH that occurred after the injection of bevacizumab. Hardy et al. reported a severe RH with long-term follow-up with peripheral retinal neovascularization on FA and a VH.[8] We attributed the VH in our case to bleeding from retinal neo-vessels. These neo-vessels likely regressed after anti-VEGF injection and were potentially associated with the TRD noted at the later surgery. After vitrectomy, the scatter laser photocoagulation was performed to treat the peripheral ischemia, which has been used in prior reports with observed regression of the iris neovascularization.[9]

Our indirect ophthalmoscopy exam under anesthesia suggested that the RH dived and connected to the choroid. In 1934, Stokes described a case of an asymptomatic 10-year-old girl with a similar arteriovenous malformation in the left eye. With keen ophthalmoscopic examination, he determined there was an unusual artery that "had the appearance of communicating directly with the choroidal circulation."[6] To our knowledge, this was the first potential description of a case of a RH with a CRA. In our patient, the FA also suggested a functional CRA, as the choroidal phase showed bright filling of the RH, before full filling of the arterioles. One limitation of our analysis is that we were unable to perform indocyanine angiography (ICGA), which could of potentially confirmed a functional CRA with true flow. First, our handheld camera (Retcam) does not have an ICGA filter function. Second, the final visual acuity of our pediatric patient was HM. Given poor visual acuity with limited fixation and cooperation, ICGA could not be performed in the clinic. Nevertheless, careful review of the OCT and OCTA B-scans revealed a clear break in Bruch's membrane, and the visualization of the connection on both en-face ORCC slab and 3-D rendering lend plausibility to a functional CRA. Given our patient's severe presentation, we

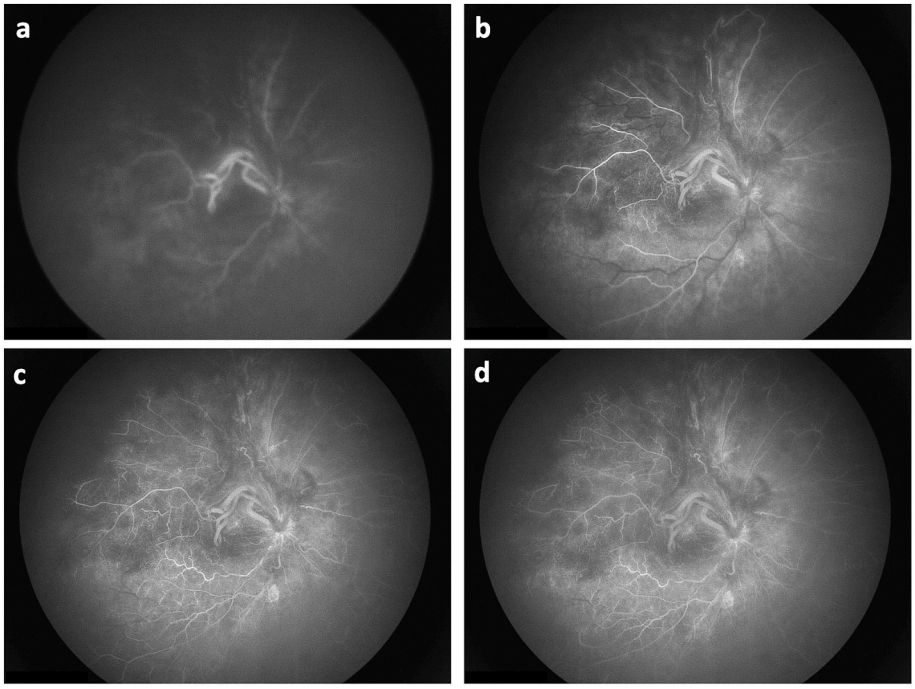

Fig. 2. Fluorescein angiogram from the repeat exam under anesthesia 2 months after presentation. (A) Choroidal phase shows bright, complete filling of the RH, before complete filling of the arterioles. There is significant choroidal non-perfusion, particularly in the supero-temporal quadrant. (B) The arterial phase shows capillary non-perfusion in the fovea and supero-temporal quadrants. (C) The arterio-venous phase again shows peripheral ischemia and macular ischemia. (D) Venous phase shows poor perfusion of the peri-foveal capillaries.

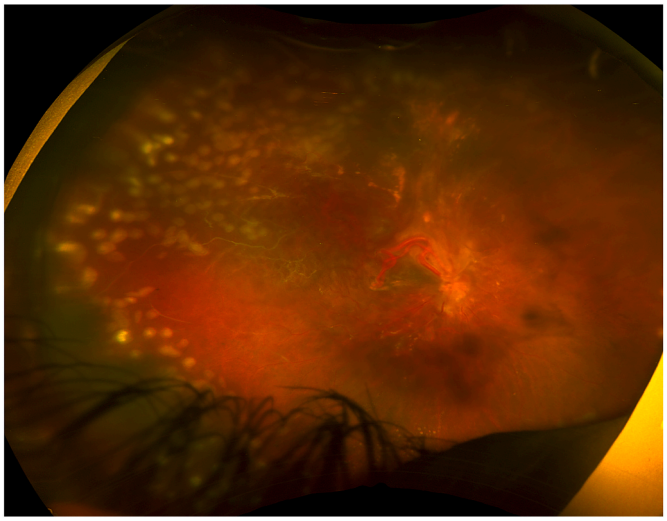

Fig. 3. Ultrawide field photograph on post-scatter laser photocoagulation day 1 shows laser treatment to areas of capillary non-perfusion in the supero-temporal and temporal periphery.

postulate that an RH with a CRA may indicate a more severe variant of RH. Potentially, high flow from the choroidal circulation would further exacerbate the hypothesized vascular steal phenomenon that contributes to retinal ischemia in this condition. We did not have longitudinal imaging before the patient's presentation; thus, we cannot be certain whether the CRA was congenital or secondary to the RH. However, given the severe retinal thinning on OCT, we postulate that the CRA may be secondary with a connection forming after severe macular ischemia. To our knowledge, this is the first case of an RH with CRA imaged using OCTA. With the growing availability of OCTA and ICG, future attention should be turned to evaluating the choroid in cases of RH to look for the possibility of a CRA.

CRediT authorship contribution statement

Brian T. Soetikno: Writing – review & editing, Writing – original draft, Methodology, Formal analysis, Data curation, Conceptualization. **Hashem Ghoraba:** Writing – review & editing, Methodology. **Arthur Brant:** Writing – review & editing, Methodology. **Charles DeBoer:** Methodology. **Lucie Guo:** Methodology. **Ann Shue:** Supervision, Methodology. **Mary Elizabeth Hartnett:** Writing – review & editing, Supervision, Conceptualization.

Patient consent

Consent to publish this case report was obtained from the patient. This report, however, does not contain any personal information that could lead to the identification of the patient.

4. Claims of priority

After conducting a literature review on racemose hemangioma and chorioretinal anastomosis with optical coherence tomography angiography utilizing PubMed, Google Scholar using the key words "racemose hemangioma", "chorioretinal anastomosis", and "optical coherence tomography angiography", we did not find prior reports other than what is mentioned in our manuscript.

Authorship

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

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No funding was received for this study.

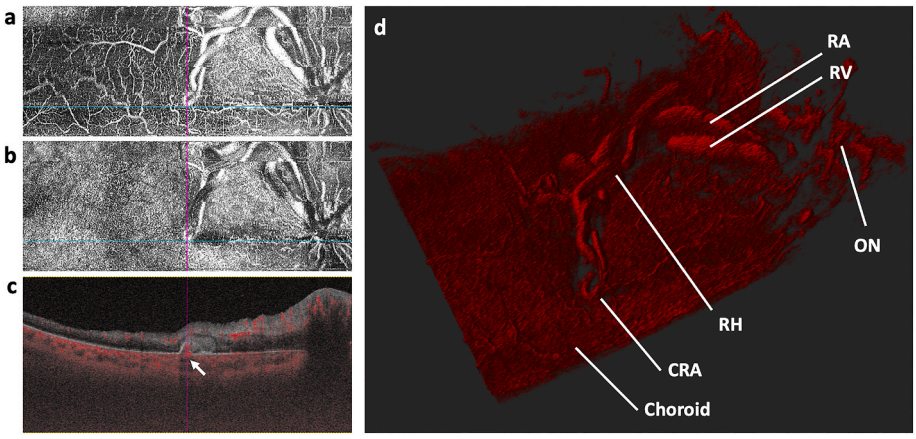

Fig. 4. Optical coherence tomography (OCT) angiography (OCTA) on post-scatter laser photocoagulation day 1 was performed to investigate the chorioretinal anastomosis. (A) En-face of the superficial plexus from the OCTA scan, taken post-operative day 1 after scatter laser photocoagulation. (B) En-face of the outer retina to choriocapillaris from the OCTA scan. (C) OCT and OCTA overlay (red) B-scan through the turquoise line in C, revealing a break in Bruch's membrane (white arrow) with RH angiographic signal adjacent to the choriocapillaris and choroid. (D) Three-dimensional rendering of the OCTA volume shows the diving portion of the RH connected to the choroidal circulation. Abbreviations: ON – optic nerve, RA – retinal artery, RV- retinal vein, RH – racemose hemangioma, CRA – chorioretinal anastomosis. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

Declaration of competing interest

The authors have no conflict of interest.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ajoc.2024.102188.

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