



Racemose hemangioma complicated with macular macroaneurysm rupture

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ABSTRACT

Purpose: To describe a rare case of racemose hemangioma which developed spontaneous macular macroaneurysm (MA) rupture and vitreous hemorrhage.

Observations: A 29-year-old healthy asian female visited our hospital and a racemose hemangioma was found in the left eye. At presentation, the best corrected visual acuity (BCVA) was 30/20 in her left eye. At 9 years after the first visit, MA-like lesion was noted in the macular area. After that, vitreous and subretinal hemorrhage appeared in the left eye. The patient underwent simultaneous vitrectomy and cataract surgery, but vitreous re-hemorrhage occurred two days after the operation. To avoid re-hemorrhage, silicone oil (SO) tamponade was added in the second vitrectomy. Two years after the second operation, SO was removed and postoperative BCVA in the left eye was 20/200 without re-bleeding in the vitreous.

Conclusions and Importance: Although retinal hemorrhages have been reported in the patients with a racemose hemangioma, in our case the macular MA rupture occurred at 9 years after the first visit. Congenital retinal arteriovenous anastomosis can show a change in vascular shape in some cases, thus it is important to observe carefully.

1. Introduction

A racemose hemangioma is a rare congenital retinal arteriovenous malformation that can occur as an isolated solitary lesion or component of Wyburn-Mason syndrome (retinal racemose hemangioma associated with intracranial vascular malformation). In cases of the latter, the patient may be affected by intracranial hemorrhage or seizures. Furthermore, the severity of ophthalmic findings can vary widely, from a singular well-defined anastomosis limited to one quadrant of the fundus to a tremendous tumor-like mass with dilated arteries and veins.¹

We recently encountered a patient with a racemose hemangioma who was complicated with a macroaneurysm (MA) rupture in the macula, as well as subretinal and vitreous hemorrhage.

1.1. Case report

When a 29-year-old female visited an ophthalmic clinic for treatment of hordeolum of the left eyelid, a racemose hemangioma was found in the left eye. The patient was referred to our hospital for further examinations.

The first visit to our hospital occurred in 2008 and visual acuity of

the left eye was 30/20, while the right eye was unremarkable. In the left eye, the tortuosity and dilation of retinal arteries and veins were severe, making it very difficult to discriminate between those blood vessel types (Fig. 1; Fig. 2A, B, C). For determination of Wyburn-Mason syndrome, a head MRI examination was performed, but the results were unremarkable.

Two years after the first visit to our hospital, when the patient was 31 years old, retinal dot hemorrhage findings were observed, with the appearance of mild branch retinal vein occlusion (BRVO) in the supratemporal area. Fluorescein angiography (FA) revealed absence of non-perfusion area, thus retinal photocoagulation therapy was not applied and the retinal hemorrhage disappeared naturally within a few months.

At the age of 32 and again at 34 years, the patient became pregnant and naturally delivered the baby without complications each time. During the associated observation periods, she was not complicated with general hypertension, hyper-lipidemia, or other noteworthy conditions. At 9 years after the first visit, MA-like lesion was noted in the macular area, though direct photocoagulation was not applied because it was thought to be unsafe due to proximity to the fovea (Fig. 2D, E, F). OCTA findings revealed macroaneurysm (MA) formation (Fig. 2-E, yellow arrow). However, because of its small size, we did not think that there

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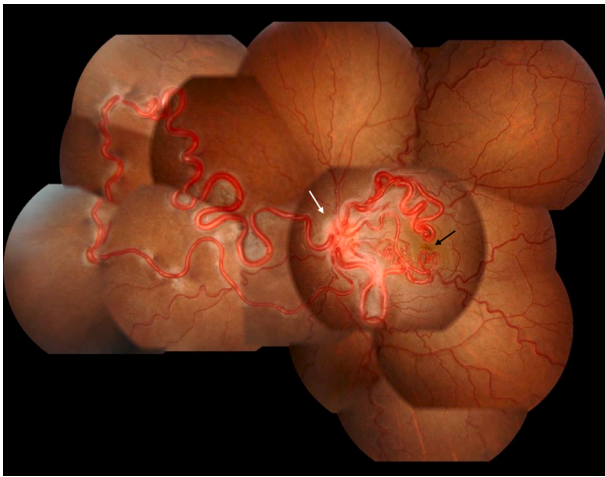


Fig. 1. Color montage fundus image of the left eye at first visit to our hospital (2008) showing retinal racemose hemangioma. No hemorrhage was found at that time. White and black arrows indicate the optic disc and the macula, respectively. . (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

was a risk of rupture. Thereafter, vitreous hemorrhage appeared in the left eye and subretinal hemorrhage at the macula was observed through the vitreous hemorrhage.

The patient underwent simultaneous vitrectomy and cataract surgery procedure. Intra-operative observation showed a subretinal hemorrhage around the macular MA as the bleeding origin of the vitreous

hemorrhage. Two days after the operation, vitreous hemorrhage recurred and a second vitrectomy was performed. There was no other bleeding origin in the retina except for the MA. In order to avoid re-bleeding in the vitreous space, a silicone oil tamponade was added during the second vitrectomy. During the SO tamponade, visual acuity of the left eye remained at 4/200 (Fig. 2G, H, I). Following the rupture, no evidence of MA formation was noted by OCTA observation (Fig. 2-I).

Nearly 2 years after the second surgery, the MA gradually changed to a white color and dry condition, thus a third vitrectomy for removal of the SO was performed. During the procedure, epiretinal membrane peeling was done, though part of the membrane remained due to its tight attachment to the macula (Fig. 2J, K, L).

Postoperative visual acuity in the left eye was 20/200 without re-bleeding in the vitreous. Vascular tortuosity and dilation of the retina observed in the former examination partially disappeared along with the appearance of vascular sheathing (Fig. 3A, B, C).

2. Discussion

Principal vascular tumors of the retina include retinal capillary hemangioma, cavernous hemangioma of the retina, racemose hemangioma of the retina, and retinal vasoproliferative tumor. Even though these lesions are classified as benign, retinal detachment caused by exudation, hemorrhage, or fibrovascular tissue proliferation can occur, resulting in severe and permanent visual impairment.²

A racemose hemangioma is a rare congenital retinal arteriovenous anastomosis that does not pass through any capillaries. It is often an incidental finding in an asymptomatic patient, with affected cases showing multiple dilated vessels in funduscopy results. This tumor has

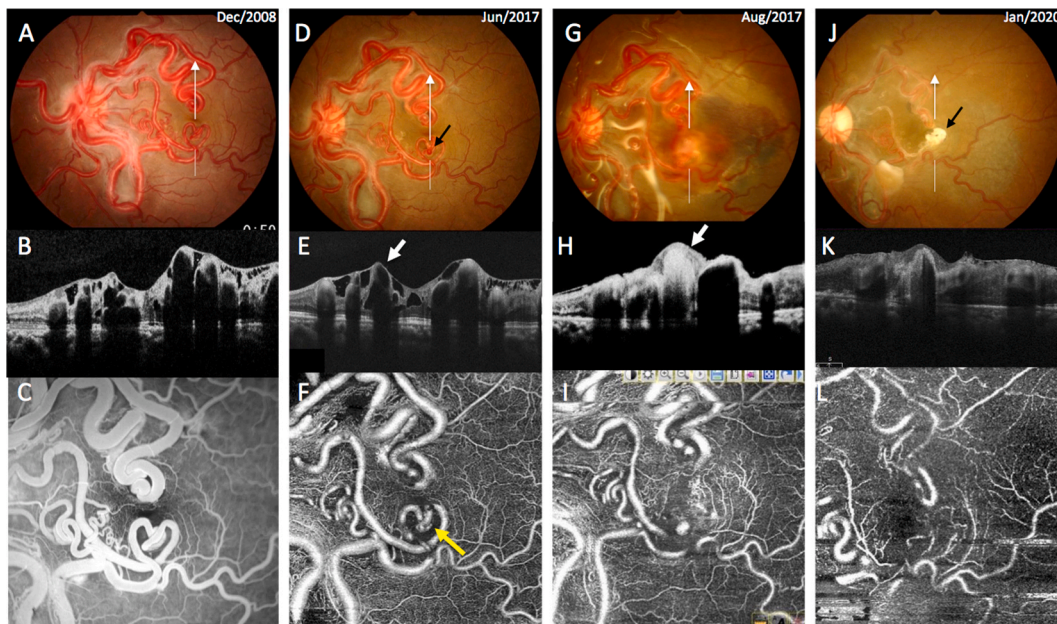


Fig. 2. A. Color fundus image showing vessel tortuosity and dilation in the vascular arcade (2008). White arrow indicates OCT scanning line. B. OCT vertical section showing elevation of retinal surface by dilated vessels (2008). C. FA in the early phase showing excessively dilated vessels near the macula (2008). D. An MA was observed (black arrow) prior to its rupture (2017). E. OCT showing partial elevation by MA formation (white arrow) prior to its rupture (2017). F. MA-like lesion observed inferior to the fovea by OCTA (black arrow) prior to MA rupture (2017). G. Subretinal hemorrhage around MA formation observed through SO tamponade following MA rupture (2017). H. OCT image showing subretinal stromal signal in area of MA rupture (white arrow) (2017). I. No strong signals similar to MA were shown by OCTA following MA rupture (2017). J. MA color changed to white (black arrow). Vessel density around the macula was decreased at 2 years after MA rupture (2020). K. OCTA findings showed decreased vascular density at 2 years after MA rupture (2020). L. Decreased vascular caliber of retinal arteries and veins was shown by OCTA at 2 years after MA rupture (2020). . (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

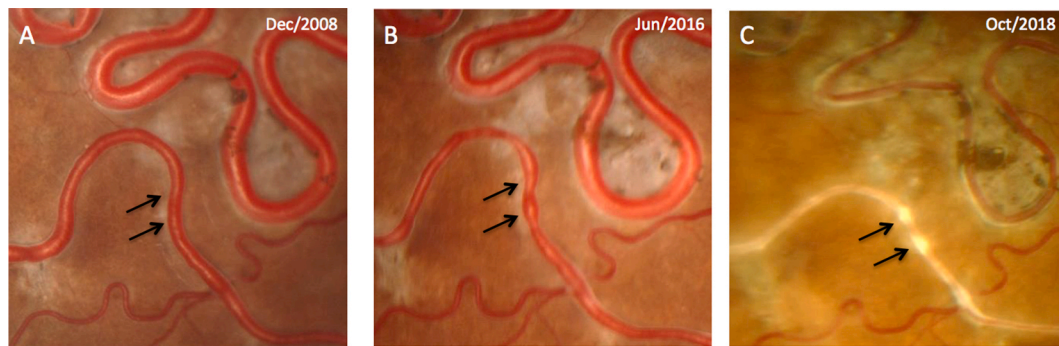


Fig. 3. A, B, C: Color fundus images of nasal area revealed that dilated vessels had decreased caliber in some areas. MA-like lesions (black arrows) were observed between the narrowed parts of the vessels. Finally, the color of these lesions changed to white, the same as noted with the MA. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

been classified into 3 groups.¹ Group 1 is characterized by an abnormal capillary plexus between arterioles and venules, and may not cause any clinical symptoms. Group 2 tumors have a direct arteriovenous communication without interposition of capillary or arteriolar elements. The dilated vessels may superficially resemble those of a capillary hemangioma, though no tumor, exudation, or retinal detachment is present. Cases classified as Group 3 have severely dilated tortuous blood vessels that are often visible throughout the whole fundus area, making it impossible to distinguish arteries from veins. Patients in this group most frequently show coincident changes in the central nervous system, which constitutes Wyburn-Mason syndrome.¹

Although some racemose hemangioma cases have no symptom,³ others have been reported to be complicated by retinal, vitreous, or subretinal hemorrhage,^{4,5} central retinal vein occlusion (CRVO) or CRVO-like retinal hemorrhage,^{6–9} a retinal MA in the macula,^{10,11} macular ischemia without retinal neovascularization or iris rubeosis,¹² or extensive retinal ischemia resulting in rubeotic glaucoma.^{13,14} Soliman reported a case of bilateral racemose hemangioma complicated with macular edema or exudation.¹⁵ Macular edema in the left eye of that case was treated with photocoagulation, while that in the right eye spontaneously healed.

Recently, optical coherence tomography (OCT) and OCT angiography (OCTA) have been shown useful as multimodal imaging modalities for diagnosis.^{11,16} An MA in racemose hemangioma cases can be observed with OCTA.¹¹ Cases of macular edema due to a racemose hemangioma have been treated with intra-vitreous injection of anti-vascular endothelial growth factor.^{9,17,18} Additionally, some cases that showed spontaneous regression of dilated vessels within a few months or years have also been reported.^{19,20}

Follow-up observation of the present case have been continuing for 11 years. During that period, retinal hemorrhage similar to BRVO, appearance of a paramacular MA, an MA rupture, vitreous and subretinal bleeding, and premacular fibrous membrane have been observed. Our patient does not have general hypertension, and has experienced 2 pregnancies, both with a natural delivery. Those pregnancy events are thought not to have had effects on disease complications because of the time period.

We speculated that the vascular dilations disappeared gradually in a segmental manner over several years. During that period, the area in which vascular caliber was not decreased remained as a dilated vascular segment and resulted in MA formation. Those changes are presented in Fig. 3-A, B, C. However, we do not think any of those triggered MA rupture. In the future, the details of this mechanism will gradually become revealed based on findings obtained with noninvasive multimodal imaging, such as OCT, OCTA, and laser speckle flowgraphy.

3. Conclusions

Congenital retinal arteriovenous anastomosis can show a change in

vascular shape in some cases, thus it is important to carefully observe and control the health condition of affected patients, who can be complicated with hypertension, diabetes, hyperlipidemia and other conditions.

Patient consent

This research has received ethic approval from The Research Ethics Committee in Hirosaki University Graduate School of Medicine. The research adhered to the tenets of the Declaration of Helsinki. Written informed consents were obtained from the patient for publication of this case report and any accompanying images.

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Conflicts of interest

The following authors have no financial disclosures: KY, YS, TT-G, KA, NM and MN.

Authorship

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

Declaration of competing interest

None of the authors have any financial/conflicting interests to disclose.

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References

- Archer DB, Deutman A, Ernest T, Krill AE. Arteriovenous communications of the retina. *Am J Ophthalmol.* 1973;75(2):224–241. [https://doi.org/10.1016/0002-9394\(73\)91018-0](https://doi.org/10.1016/0002-9394(73)91018-0).
- Knutsson KA, De Benedetto U, Querques G, Del Turco C, Bandello F, Lattanzio R. Primitive retinal vascular abnormalities: tumors and telangiectasias. *Ophthalmologica.* 2012;228:67–77. <https://doi.org/10.1159/000338230>.
- Knecht PB, Bosch MM, Helbig: fibrotic racemose haemangioma of the retina. Das fibrosierte razemöse hämangiom der Retina. *Klin Monatsbl Augenheilkd.* 2008;225(5):495–496. <https://doi.org/10.1055/s-2008-1027350>.
- Bernth Petersen P. Racemose haemangioma of the retina. Report of three cases with long term follow-up. *Acta Ophthalmol.* 1979;57(4):669–678. <https://doi.org/10.1111/j.1755-3768.1979.tb00516.x>.
- Papageorgiou KI, Ghazi-Nouri SMS, Andreou PS. Vitreous and subretinal haemorrhage: an unusual complication of retinal racemose haemangioma. *Clin Exp*

- Ophthalmol.* 2006;34:176–181. <https://doi.org/10.1111/j.1442-9071.2006.01178.x>.
6. Yang C, Liu YL, Dou HL, Lu XR, Qian F, Zhao L. Unilateral hemi-central retinal vein obstruction associated with retinal racemose angioma. *Jpn J Ophthalmol.* 2009;53(4):435–436. <https://doi.org/10.1007/s10384-009-0673-8>.
 7. Stoffelns BM. Ungewöhnliche Präsentation eines isolierten razemösen Hämangioms der Netzhaut mit Involution nach Zentralvenenverschluss. Unusual presentation of a racemose haemangioma of the retina with involution after central retinal vein occlusion. *Klin Monatsbl Augenheilkd.* 2014;231(4):436–438. <https://doi.org/10.1055/s-0034-1368249>.
 8. Qin X-J, Huang C, Lai K. Retinal vein occlusion in retinal racemose hemangioma: a case report and literature review of ocular complications in this rare retinal vascular disorder Qin et al. *BMC Ophthalmol.* 2014;14:101. <http://www.biomedcentral.com/1471-2415/14/101>.
 9. Winter E, Elsas T, Austeng D. Anti-VEGF treating macular oedema caused by retinal arteriovenous malformation – a case report. *Acta Ophthalmol.* 2014;92:192–193. <https://doi.org/10.1111/aos.12019>.
 10. Bech K, Jensen OA. Racemose haemangioma of the retina. Two additional cases, including one with defects of the visual fields as a complication of arteriography. *Acta Ophthalmol.* 1958;36:769–781. <https://doi.org/10.1111/j.1755-3768.1958.tb02287.x>.
 11. Vishal R, Avadesh O, Srinivas R, Das Taraprasad D. Retinal racemose hemangioma with retinal artery macroaneurysm: optical coherence tomography angiography (OCTA) findings. *American Journal of Ophthalmology Case Reports.* 2018;11:98–100. <https://doi.org/10.1016/j.ajoc.2018.06.018>.
 12. Panagiotidis D, Karagiannis D, Tsoumpris I. Spontaneous development of macular ischemia in a case of racemose hemangioma. *Clin Ophthalmol.* 2011;5:931–932. <https://doi.org/10.2147/OPTH.S21925>.
 13. Bloom PA, Laidlaw A, Easty DL. Spontaneous development of retinal ischaemia and rubeosis in eyes with retinal racemose angioma. *Br J Ophthalmol.* 1993;77(2):124–125. <https://doi.org/10.1136/bjo.77.2.124>.
 14. Soliman W, Haamann P, Larsen M. Exudation, response to photocoagulation and spontaneous remission in a case of bilateral racemose haemangioma. *Acta Ophthalmol.* 2005;84:429–431. <https://doi.org/10.1111/j.1600-0420.2005.00644.x>.
 15. Efron L, Zakov ZN, Tomsak RL. Neovascular glaucoma as a complication of the Wyburn-Mason syndrome. *J Clin Neuro Ophthalmol.* 1985;5(2):95–98.
 16. Mota SHE, Hernandez-Delgado AG, Gonzalez-Cortes J, Bejar-Macouzet L, Mennel S, Olvera-Barrios A. Retinales razemöses Hämangiom und multimodale Bildgebung: fallbericht Retinal Racemose Hemangioma and Multimodal Imaging: case Report. *Klin Monatsbl Augenheilkd.* 2020;237(1):87–89. <https://doi.org/10.1055/a-0898-3734>.
 17. Barreira Jr AK, Nakashima AF, Takahashi VK, Marques GA, Minelli T, Santo AM. Retinal racemose hemangioma with focal macular involvement. *Retin Cases Brief Rep.* 2016;10:52–54. <https://doi.org/10.1097/icb.0000000000000159>.
 18. Pangtey BPS, Kohli P, Ramasamy K. Wyburn–Mason syndrome presenting with bilateral retinal racemose hemangioma with unilateral serous retinal detachment. *Indian J Ophthalmol.* 2018;66:1869–1871. https://doi.org/10.4103/ijo.IJO_455_18.
 19. Dekking HM. Arteriovenous aneurysm of the retina with spontaneous regression. *Ophthalmologica.* 1955;130:113–115.
 20. Elizalde J, Vasquez L. Spontaneous regression in a case of racemose haemangioma Archer's type 2. *Retin Cases Brief Rep.* 2011;5:294–296. <https://doi.org/10.1097/ICB.0b013e3181f66a97>.