Optical coherence tomography angiography of fleeting macroaneurysm in retinal racemose angioma

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We present a rare case of retinal racemose angioma complicated with fleeting macroaneurysm (MA). A 50-year-old female presented with diminution of vision in her right eye for 6 years. Fundus examination showed a racemose angioma with hemorrhagic MA temporal to the fovea in the right eye. On subsequent follow-ups, spontaneous thrombosis of MA was noted with the development of new MA inferior to the fovea, with intraretinal hemorrhage extending into the fovea. Focal laser to MA resulted in resolution of MA with improvement in vision. We report optical coherence tomography angiographic features of the fleeting MA in a case of racemose angioma.

Key words: Angiography, macroaneurysm, optical coherence tomography angiography, racemose angioma

Retinal racemose angioma (RRA), also called retinal arteriovenous malformations (AVMs), is a rare, congenital sporadic phakomatosis characterized by dilated and tortuous vessels frequently extending from the disc unilaterally.[1] Association of racemose angioma with AVM of the brain is known as Wyburn–Mason syndrome or Bonnet–Dechaume–Blanc syndrome.[2] RRA was once thought to be benign and follow a stable clinical course.[3] However, they can be complicated with vascular occlusions, retinal hemorrhages, and macular edema, leading to vision loss.[4] We present a rare case of RRA complicated with fleeting macroaneurysm (MA) and describe various optical coherence tomography angiography (OCTA) features.

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Case Report

A 50-year-old female presented with blurred vision in the right eye for 6 years. Family history and review of systems were unremarkable. Best-corrected visual acuity (BCVA) was 20/200 in the right eye and 20/20 in the left eye. Anterior segment examination was normal in both the eyes. Fundus examination of the right eye showed large dilated tortuous vessels emanating from disc with multiple arteriovenous shunting of vessels in all quadrants and sclerosed vessels nasally with multiple acute-angled bends. Arterial MA with intralesional hemorrhage was noted temporal to the fovea [Fig. 1a]. Fundus examination of the left eye was unremarkable [Fig. 1b]. OCTA (DRI Triton, Topcon Inc., Japan) showed multiple arteriovenous shunts and vessel of origin of MA with surrounding hemorrhagic shadow [Fig. 1c]. Optical coherence tomography through the MA showed involvement of inner retinal layers with underlying shadowing [Fig. 1d]. The patient was diagnosed with combined Type II and III RRA. The patient was treated conservatively, the lesion being away from the fovea. At 6-month follow-up, she had further diminution of vision to 20/600 in the right eye. At this follow-up, macroaneurysm noted in previous visit had thrombosed with development of new MA inferior to the fovea with intraretinal hemorrhage extending into the fovea [Fig. 2a and b]. Three-month postfocal laser BCVA was 20/400 with complete resolution of MA, hemorrhage, and

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macular edema [Fig. 2c]. Since MA caused foveal edema, focal laser was done using 532 nm and Nd: YAG laser to occlude the aneurysm [Fig. 2d]. Serial OCTA showed MA shadowed by overlying hemorrhage and patent angulated blood vessel at previous MA [Fig. 3a-c]. Grapevine-like creeping of capillaries around superior arcade vessels was noted [Fig. 4a]. Fundus fluorescein angiography (FFA) at 6-month follow-up demonstrated hemorrhagic MA with minimal leak in the late phase, acute-angled patent blood vessel at the site of previous MA temporal to the fovea with multiple nonleaking arteriovenous shunts, nasal attenuated vessels with surrounding staining and grapevine-like creeping of capillaries around superior major blood vessels [Fig. 4b].

Discussion

Arteriovenous communications are congenital malformations of the vascular system. RRA is characterized by the presence of dilated vessels with or without communicating capillary network. As such Type I and II AVMs have stable clinical course without evidence of progression.[3] Common complications associated with AVMs include intraretinal hemorrhage, retinal vein obstruction, vitreous hemorrhage, and neovascular glaucoma.[4] This has been attributed to hyperdynamic flow through small caliber vessels and steal phenomenon caused by increased venous pressure and decreased arterial pressure.[5]

A literature review done by Qin *et al*. showed rare occurrence of MA in case of racemose angioma. They reviewed 167 cases of racemose angioma, among which only one case had MA.[5] There is a single case report by Tilanus *et al*. demonstrating fleeting MA in a case of racemose angioma over a course of 4 years.[6] In our case, we noticed Type II and III racemose angioma with development of new MAs at different sites. MAs resolved, leaving behind patent blood vessel with surrounding gliotic scar and focal progressive narrowing of the vessel at site of MA. In our case, OCTA was compared to conventional FFA. The grapevine-like appearance in racemose angioma was earlier reported by Biro.[7] This grapevine-like appearance was noted in our case which could be appreciated by both FFA and OCTA but was better seen on FFA. Unlike FFA, OCTA could identify the vessel of origin of MA. All the features appreciated in FFA such as AVM and patency of nasal vessels were equally appreciated in OCTA. Serial follow-up of the case with OCTA showed halo around the vessel at the site of MA corresponding to the gliotic tissue. Progressive kinking of vessels at the site of MAs was noted on serial OCTA scans. We noted that the angulation becomes more acute with time. This might explain the reason for multiple 90-degree bending of vessels nasally with gliotic tissue at the bending site.

Hence, OCTA is an effective, noninvasive, dye less, repeatable tool for studying the vascular malformations in a case of racemose angioma. The vessel of origin of MA and resolution of MA can be better studied with OCTA as the test can be repeated at each visit. It thus helps us to understand the natural course of MAs in racemose angioma. Limitation of OCTA is that it cannot demonstrate leakage of MA.

Conclusion

To best of our knowledge, there is only one published case report of MA in racemose angioma. This is the first case report on OCTA in case of racemose angioma. This study also throws light on natural course of MA in racemose angioma.

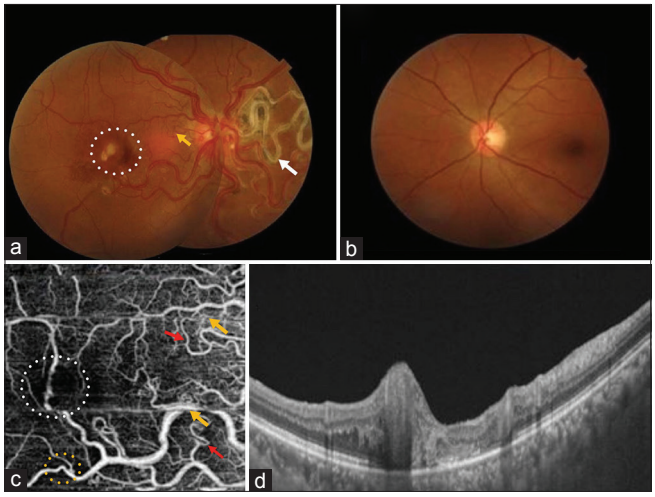

Figure 1: (a) Color photograph of the right eye showing dilated, tortuous vessels with arteriovenous malformations (yellow arrow), sclerosed looking vessels nasally (white arrow), and macroaneurysm temporal to the fovea (white-dotted circle). (b) Color fundus photograph of the left eye. (c) Optical coherence tomography angiography of the right eye showing abnormally dilated vessels with arteriovenous malformation (red arrows) and motion artifacts (yellow arrows) and macroaneurysm showing vessel of origin with halo due to masking by hemorrhage (white-dotted circle) and kinked vessel (yellow-dotted circle). (d) Optical coherence tomography angiography of the right eye passing through macroaneurysm showing hyperreflective inner retinal layers with backshadowing (white arrow)

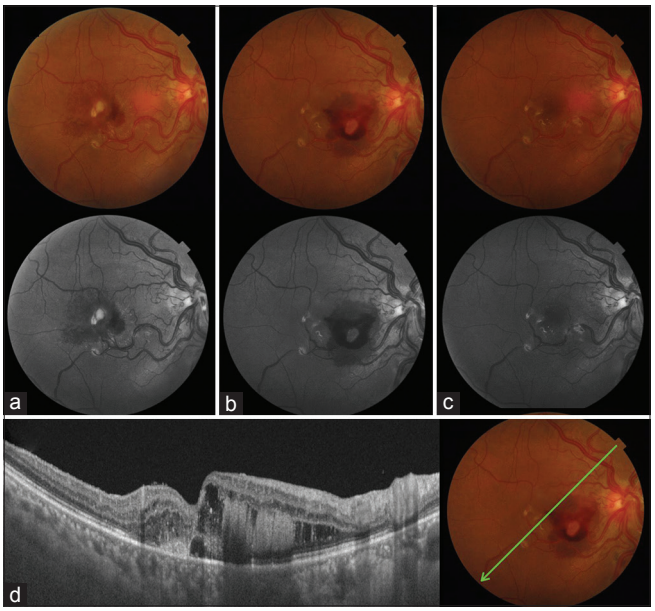

Figure 2: Serial color fundus photographs and corresponding red-free image of the right eye. (a) At presentation showing macroaneurysm temporal to the fovea with surrounding hemorrhage. (b) At 6 months showing spontaneous resolution of macroaneurysm with appearance of surrounding gliotic tissue and appearance of new macroaneurysm inferior to the fovea with intraretinal hemorrhage involving fovea. (c) At 9 months showing resolution of macroaneurysm with glial scar at sites of previous macroaneurysm. (d) Coherence tomography angiography scan of the right eye showing intraretinal cystoid spaces at the fovea and intraretinal hyperreflectivity corresponding to intraretinal hemorrhage

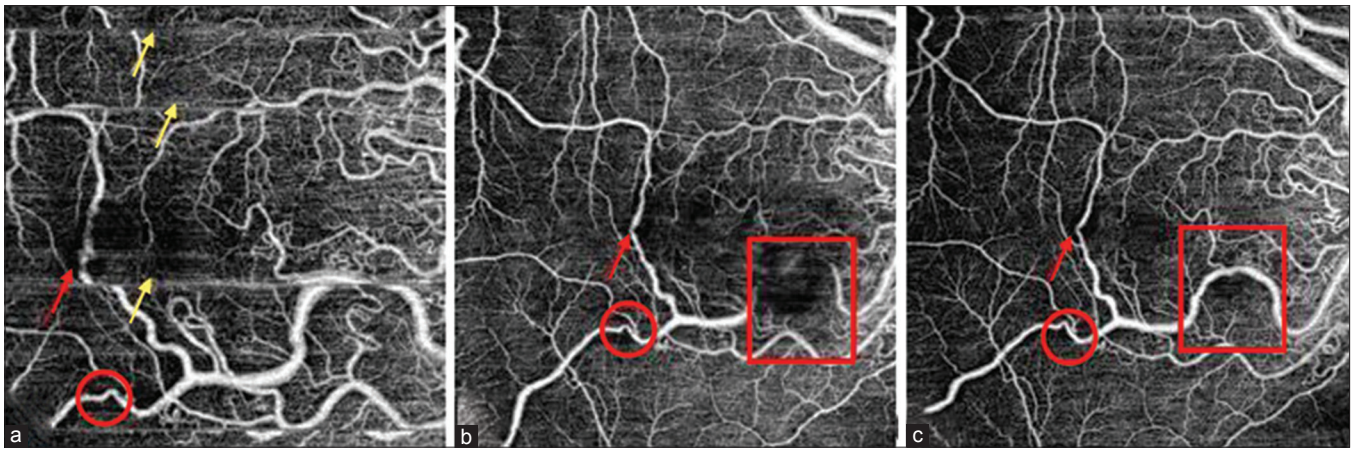

Figure 3: Optical coherence tomography angiography of the right eye showing abnormally dilated vessels with arteriovenous malformation: (a) 6 mm × 6 mm slab with motion artifacts (yellow arrow) at site of macroaneurysm showing vessel of origin with halo due to masking by hemorrhage (red arrow), kinked vessel with halo (red circle), (b) 9 mm × 9 mm slab at 6-month follow-up showing acute angulation of vessel (red arrow), increased angulation of vessel (red circle), and blockage at site of new macroaneurysm (red square), (c) 9 mm × 9 mm optical coherence tomography angiography at 9 months showing acute angulation of vessel at the site of previous macroaneurysm (red arrow) and progressive kinking (red circle) and resolution of macroaneurysm with development of halo

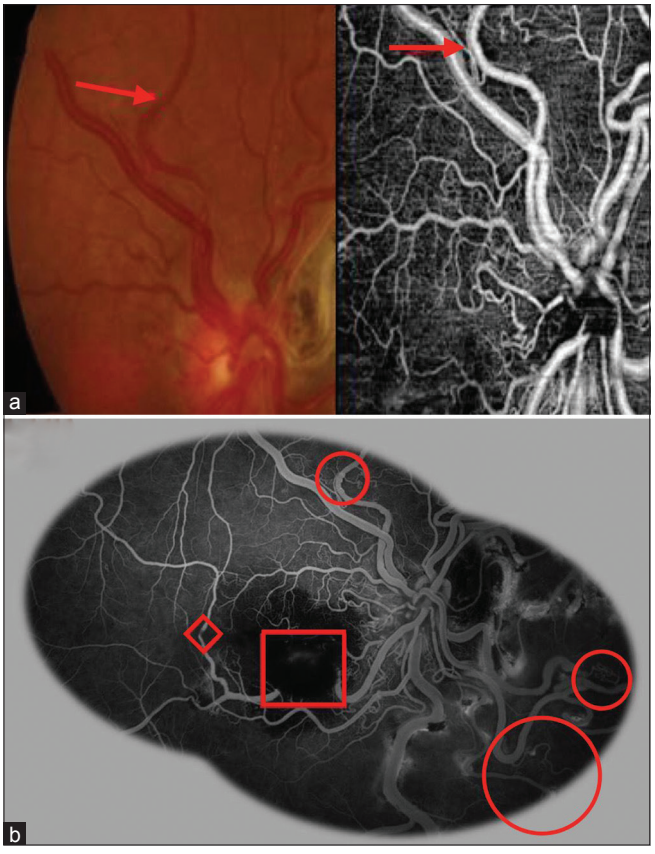

Figure 4: (a) Fundus image and corresponding optical coherence tomography angiography showing grapevine-like twining of vessels (red arrow). (b) Late phase of fluorescein angiography at 6-month follow-up showing dilated tortuous vessels, nonleaking arteriovenous communications (circle), macroaneurysm with leakage (square), and obliterated macroaneurysm with patent lumen (diamond)

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Archer DB, Deutman A, Ernest JT, Krill AE. Arteriovenous communications of the retina. Am J Ophthalmol 1973;75:224-41.
2. Wyburn-Mason R. Arteriovenous aneurysm of midbrain and retina, facial Nevi and mental changes. Brain Dev 1943;66:40.
3. Leitão Guerra RL, Leitão Guerra CL, Guerra M, Guerra Neto AS, Leitão Guerra AA. Retinal racemose hemangioma (Wyburn-Mason syndrome) – A patient ten years follow-up: Case report. Arq Bras Oftalmol 2009;72:545-8.
4. Mansour AM, Wells CG, Jampol LM, Kalina RE. Ocular complications of arteriovenous communications of the retina. Arch Ophthalmol 1989;107:232-6.
5. Qin XJ, 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. BMC Ophthalmol 2014;14:101.
6. Tilanus MD, Hoyng C, Deutman AF, Cruysberg JR, Aandekerk AL. Congenital arteriovenous communications and the development of two types of leaking retinal macroaneurysms. Am J Ophthalmol 1991;112:31-3.
7. Biro I. Racemose retinal aneurysm. Ophthalmologica 1951;121:201-7.