

Venovenous anastomosis in macular telangiectasia type 2: an unusual presentation

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DESCRIPTION

A 55-year-old woman presented with reduced vision in both eyes (BE). Her visual acuity was 20/100 in the right eye (RE) and 20/320 in the left eye (LE). Anterior segment was normal except for early cataract. Fundus evaluation showed pigment alteration at the macula with subretinal neovascular membrane (SRNVM) and venovenous anastomosis of major veins around the fovea temporally in BE (figure 1A). Fundus fluorescein angiography (FFA) and indocyanine green confirmed SRNVM and venovenous anastomosis in BE (figure 1C–F; *).

Optical coherence tomography (OCT) in the RE showed posterior vitreous detachment and epiretinal membrane with intraretinal hyporeflective cystoid spaces, mild retinal thickening, hyperreflective dots in outer retina and disrupted subfoveal ellipsoid zone. In addition, OCT in the LE showed subretinal hyperreflective material. (figure 2A). Optical coherence tomography angiography (OCTA) en-face projection images showed the venovenous anastomosis (arrows figure 2B). OCTA also showed telangiectasia of parafoveal vessels in the deep capillary plexus.

The patient received intravitreal ranibizumab in BE. After 1 month, her vision improved to 20/60 in the RE and remained stable in the LE. OCT showed reduced macular thickness.

MacTel type 2 is a neurodegenerative disorder involving Müller cells. It is characterised by parafoveal greying with crystalline deposits, pigment clumps and right-angled venules.^{1 2} SRNVM is a known complication of MacTel type 2 and is

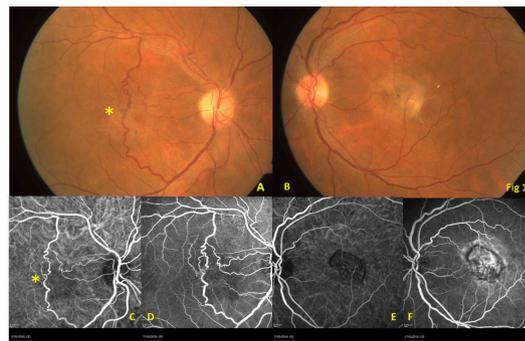


Figure 1 (A and B) Fundus evaluation showed pigment alteration at macula with subretinal neovascular membrane (SRNVM) and vascular anastomosis of major arcades veins around the fovea temporally in both eyes (BE). (C–F; *) Fundus fluorescein angiography and indocyanine green confirmed SRNVM and venovenous anastomosis in BE.

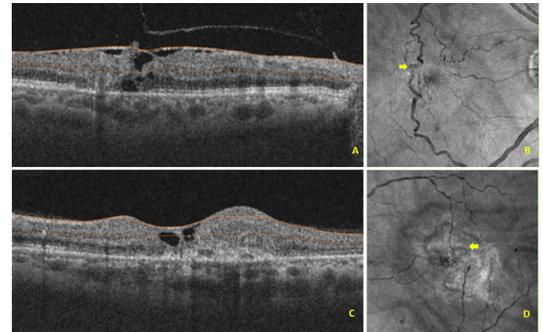


Figure 2 (A and C) The optical coherence tomography (OCT) in the right eye showed posterior vitreous detachment and epiretinal membrane with intraretinal hyporeflective cystoid spaces, mild retinal thickening, hyperreflective dots in outer retina and disrupted subfoveal ellipsoid zone. In addition, OCT in the left eye showed subretinal hyperreflective material. (B and D) Optical coherence tomography angiography en-face projection images showed the venovenous anastomosis (yellow arrows).

estimated to occur in 10% of patients with this disease.³

Venovenous anastomosis or collaterals of retinal vessels is a rare entity, commonly associated with retinal venous occlusion.⁴

In this patient, anastomosis of main arcade vessels is a novel finding in the setting of MacTel, which alerted us to carefully look for any vascular changes. On carefully reviewing the FFA images, flow restriction in superior vein was evident and probably, it was an old venous occlusion leading to formation of collaterals. The learning from this case was the fact that comorbidities do exist, and one needs to be clinically vigilant. This case also highlights the role of non-invasive en-face imaging in diagnosis of vascular anomalies.



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Learning points

- ▶ En-face scan on optical coherence tomography angiography is a useful non-invasive modality to view vascular anomalies.
- ▶ Novel finding of anastomosis of main arcade vessels gives a clue that comorbidities can occur, and that one needs to be clinically vigilant.

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