Multimodal imaging of an idiopathic florid vascularised epiretinal membrane: Course, treatment, and outcome

Poornachandra B, Edwin James*, Aditya Aseem*, Chaitra Jayadev, Srinivasan Sanjay, Ashwini K T¹, Santosh G K Gadde, Naresh K Yadav

Idiopathic vascular epiretinal membrane is an extremely rare entity and the pathogenesis and clinical course is not

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Department of Vitreo-Retina, Narayana Nethralaya Eye Institute, ¹Kidwai Memorial Institute of Oncology, Bengaluru, Karnataka, India

*Both the authors have contributed equally.

Correspondence to: Dr. Poornachandra B, Narayana Nethralaya Eye Institute, 121/C, Chord Road, Rajajinagar, 1st 'R' Block, Bangalore, Karnataka - 560 010. India. E-mail: punith.poorna@gmail.com

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clearly understood. A 53-year-old hypertensive female patient presented with complaints of altered vision in the right eye. On examination, her vision was 20/30 and fundoscopy showed a vascularized epiretinal membrane (ERM), which was confirmed on spectral-domain optical coherence tomography. No primary cause was found after investigations. The symptoms and ERM showed slow progression over the next three years with a visual acuity of 20/60. She underwent surgery for removal of the ERM, which was subjected to histopathological evaluation. This is a unique case of a florid proliferative vascularisation of an ERM in the absence of any identifiable cause, which had a good visual outcome following surgery.

Key words: Metamorphopsia, spectral domain optical coherence tomography, vascularised epiretinal membrane

Metamorphopsia and blurred vision are common symptoms in patients with an epiretinal membrane (ERM). This is an abnormality of the vitreomacular interface due to fibrocellular proliferation on the inner surface of the retina. Epiretinal membranes have been classified as idiopathic, or secondary to inflammatory conditions such as posterior uveitis, vascular diseases like diabetic retinopathy and retinal vein occlusion,

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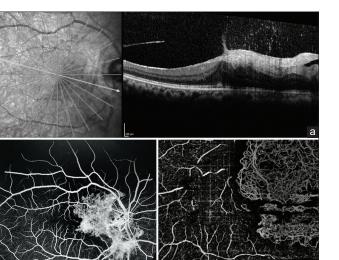


Figure 1: (a) Spectral-domain optical coherence tomography of the right eye showing the thickness and extent of the epiretinal membrane at the first visit. (b) Fundus fluorescein angiography of the right eye showing a vascular complex in the peripapillary and macular area increasing in intensity and size in the late phases with no capillary non-perfusion areas at the first visit. (c) Optical coherence tomography angiography of the right eye showing an abnormal vascular complex

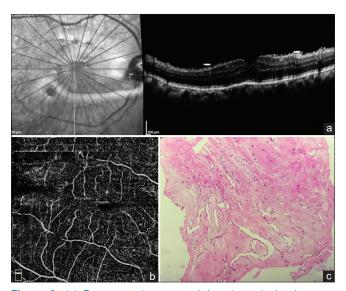


Figure 3: (a) Post-operative spectral-domain optical coherence tomography of the right eye showing a normal foveal contour. (b) Post-operative optical coherence tomography angiography of the right eye showing near normal morphology of the vasculature. (c) Histopathological examination with hemotoxylin-eosin stain showing hyalinised fibrocollagenous tissue containing proliferating blood vessels

cytomegalovirus retinitis, blunt trauma, retinal detachment and repair, argon laser retinal photocoagulation, and cataract surgery. Formation of fibrovascular membranes at the vitreoretinal interface has been classically associated with ischaemic retinopathies, as these areas in the retina release angiogenic factors. Surgery for idiopathic ERMs is a safe and effective procedure with good postoperative visual and anatomical improvement, and an overall low recurrence and complication rate.^[1]

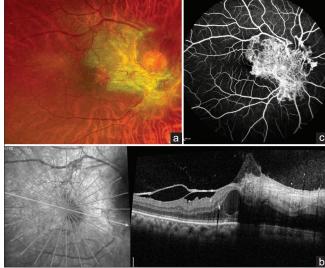


Figure 2: (a) Fundus photo of the right eye on follow up showing extension of the epiretinal membrane. (b) Spectral-domain optical coherence tomography of the right eye on follow up showing increased thickness and traction with cystoid changes. (c) Fundus fluorescein angiography of the right eye showing increased vascularity of the epiretinal membrane

We report a unique case of an idiopathic ERM complicated by neovascularisation, in the absence of ischaemic retinopathy, and the treatment course and outcome.

Case Report

A 53-year-old Asian Indian woman, presented with a blurring of vision in the right eye of 4 months duration. She was a known hypertensive on treatment with no significant previous history of trauma or ocular treatment. Her corrected distance visual acuity (CDVA) was 20/30 in the right eye and 20/20 in the left eye. Her intraocular pressures were within normal limits. The anterior segment examination was normal in both eyes. The fundus examination of the right eye revealed a thick vascular ERM extending from the peripapillary area to the foveal region with no evidence of hypertensive retinopathy. The fundus examination was normal in the left eye. Spectral-domain optical coherence tomography (SDOCT; Spectralis™ Heidelberg Engineering, Germany) in the right eye revealed a hyper-reflective membrane on the surface of the retina measuring 177µ in its thickest part [Fig. 1a]. Fundus fluorescein angiography revealed a normal arm-retina time (14 sec), and the arterio-venous phase showed a vascular complex in the peripapillary and macular area which increased in intensity and size in the late phases. There was no evidence of capillary non-perfusion or neovascularization elsewhere [Fig. 1b]. Optical coherence tomography angiography (OCTA; AngioVue Imaging System, Optovue, Inc., Freemont, CA) showed an abnormal vascularization with high vascular flow in the ERM [Fig. 1c]. A detailed evaluation did not reveal any systemic vascular risk factors.

The patient was lost to follow-up for 3 years and presented with decreased vision in the right eye. The CDVA was 20/60 in the right eye and 20/20 in the left eye. Fundus examination of the right eye showed that the membrane had increased in size [Fig. 2a] and had increased in thickness on the SDOCT (305μ) with cystoid changes [Fig. 2b]. Fundus fluorescein angiography revealed an increase in the size and vascularity of the membrane [Fig. 2c]. Since the patient was now symptomatic, surgical intervention was planned with an intravitreal anti-vascular endothelial growth factor injection followed five days later by pars plana vitrectomy and ERM removal with silicone oil tamponade due to profuse intraoperative bleed. Post-operative follow-up at the end of 3 weeks showed the CDVA in the right eye had improved to 20/30, with an attached retina and normal foveal contour [Fig. 3a]. The morphology on OCTA was near normal [Fig. 3b].

Histopathological examination of the ERM revealed hyalinised fibrocollagenous tissue containing proliferating blood vessels with no granulomatous or malignant cells [Fig. 3c].

Discussion

Fibrovascular ERMs are a frequent complication of proliferative diabetic retinopathy and retinal vein occlusions.^[2] There has been one report of a patient with Terson syndrome presenting with an ERM complicated by preretinal neovascularization.^[3] One hypothesis has been that hypertensive changes may decrease retinal vascular perfusion, which creates a favorable environment for secondary neovascularization of the idiopathic ERM through vascular endothelial growth factor upregulation.^[4] An association of a vascularized ERM with a vasoproliferative has been reported and HPE revealed that apart from the vascular elements, the architecture and staining characteristics of this ERM were identical to a non-vascularized ERM and posterior hyaloid membrane.^[5] Our patient did not have any of the above risk factors or associations.

The treatment for visually significant ERMs remains surgical intervention. Innovations in technology, including instrumentation, visualization systems, enhanced light filters, high viscosity dyes, and small gauge forceps, have improved the outcomes of macular surgeries. The challenge with vascular ERMs is intraoperative hemorrhage at the time of membrane peeling. While we did give intravitreal anti-vascular endothelial growth factor injection prior to the surgery, there was significant bleed, which prompted us to use silicone oil as a tamponade. Completion of the ERM removal is also mandatory to prevent recurrence of the fibrovascular proliferation. This can be ensured during the surgical procedure by using an intraoperative optical coherence tomography.^[6] The post-operative SDOCT of our patient confirmed the complete removal of the ERM.

Conclusion

Idiopathic vascular ERM is a rare entity, the etiopathogenesis of which is yet to be understood. A better understanding of the same along with an assessment of intravitreal VEGF levels would help the course of such florid vascularisation and slow progression before and after surgery. Our case is unique in that it had florid proliferative vascularisation, which progressed over time and had a good visual outcome following surgery. Multimodal imaging helped to monitor progression and treatment planning.

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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Conflicts of interest

There are no conflicts of interest.

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