Multimodal Imaging of an Idiopathic Vascularized Epiretinal Membrane: A Case Report

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Patient: Male, 70-year-old
Final Diagnosis: Vascularized epiretinal membrane
Symptoms: Asymptomatic
Clinical Procedure: —
Specialty: Ophthalmology

Objective: Rare disease

Background: Idiopathic epiretinal membranes (ERMs) are commonly associated with fibrovascular tissue, primarily observed in ischemic retinopathies. However, idiopathic vascularized ERMs (IVEM) are exceedingly rare, and their pathogenesis and clinical course remain poorly understood. This report aims to contribute to the limited literature on IVEM, shedding light on its characteristics and potential implications for patient management.

Case Report: We present the case of a 70-year-old man diagnosed with idiopathic ERM in the left eye, revealing a neovascular complex within the membrane. Despite the absence of ocular symptoms and medical history, multimodal imaging using the Nidek Mirante, including spectral domain optical coherence tomography (SD-OCT) and optical coherence tomography angiography (OCT-A), revealed a thick pre-retinal hyper-reflective line with a partial posterior vitreous detachment and an abnormal vascular complex resembling a pruned-vascular-tree pattern. Notably, fluorescein angiography confirmed hyperfluorescence and leakage corresponding to the observed vessels. Despite the rarity of IVEM, the patient remained asymptomatic, and observation was deemed appropriate.

Conclusions: IVEM poses a rare challenge in clinical practice, necessitating a comprehensive understanding of its features and potential complications. While the etiopathogenesis remains unclear, hypertension has been proposed as a contributing factor. This case adds valuable insights to the growing literature on IVEM, emphasizing the importance of multimodal imaging in diagnosis and decision-making. Given the limited reports and varied treatment outcomes, managing IVEM requires careful consideration of observation and various therapeutic approaches, highlighting the need for further research to optimize patient care.

Keywords: Epiretinal Membrane • Multimodal Imaging • Neovascularization, Pathologic

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Background

Idiopathic epiretinal membrane (ERM) consists of fibrocellular tissue caused by pre-retinal proliferation of myofibroblastic cells on the surface of the internal limiting membrane [1]. Although fibrovascular ERMs are commonly found in ischemic retinopathies such as proliferative diabetic retinopathy and retinal vein occlusion, idiopathic vascular ERMs have been rarely described in the literature [2].

An idiopathic vascularized epiretinal membrane (IVEM) without a clear association with ischemic retinopathy is extremely rare and its pathogenesis and clinical course remain poorly elucidated [2]. The incorporation of multimodal imaging in the examination of these membranes provides a greater understanding of IVEM, allowing for a more precise characterization of the vascular alterations and their impact on retinal architecture.

We present the case of a patient diagnosed with an IVEM with no associated ischemic retinopathy, evaluated with different imaging modalities.

Case Report

A 70-year-old man diagnosed with idiopathic ERM in his left eye (LE) was referred to our hospital for further examination as a neovascular complex was identified within the ERM. He was non-diabetic and non-hypertensive, with no ocular symptoms and no past medical history.

At presentation, his best-corrected visual acuity (BCVA) was 20/25 in both eyes. Slit lamp examination of the anterior segment on both eyes was unremarkable. Fundus examination was normal in his right eye (RE). On fundus examination of the LE, a semi-translucent membrane was observed in the superior temporal vascular arcade with an associated flat vascularization complex of thin vessels underneath. Associated subtle overlying folds were observed with no visible signs of vitreoretinal traction (Figure 1A, 1B). Multimodal imaging was obtained using the Nidek Mirante (https://usa.nidek.com/mirante/) (NIDEK Co., Ltd., Gamagori, Japan). Spectral domain optical coherence tomography (SD-OCT) showed a thick pre-retinal hyper-reflective line with partial posterior vitreous detachment (PVD), no vitreoretinal traction, and a subtle irregularity in the inner and intermediate retina layers (Figure 1C, 1D). Fluorescein angiography revealed hyperfluorescence with leakage corresponding to the thin vessels observed clinically. No areas of capillary closure were identified (Figure 1E). Optical coherence tomography angiography (OCT-A) showed a high-flow abnormal vascular complex with unbranched long dilated filamentous vessels similar to pruned-vascular-tree pattern in the superficial capillary plexus (Figure 1F, 1G). No anomalies were found on his RE. Considering his BCVA and lack of ocular symptoms, we decided on observation only. At his last follow-up, he remained asymptomatic and his bilateral BCVA continued to be 20/25.

Discussion

An idiopathic vascularized epiretinal membrane (IVEM) is a rare entity that can have a significant impact on vision; it has been mainly described in the posterior pole of the retina [2,3]. It has been reported that even avascular membranes can develop abnormal vascularization, since hypoxia-inducible factor-1α (HIF-1α), vascular endothelial growth factor (VEGF), and tumor necrosis factor-α (TNF-α) have been identified in idiopathic ERMs [3,4]. Although its etiopathogenesis is not well understood, Anguita et al proposed hypertension as a primary cause of IVEM, as it can increase VEGF and decrease retinal perfusion [3]. This theory was supported by Toffoli et al, who presented a case of neovascularized ERM in a patient with Terson syndrome, which they hypothesized was secondary to systemic hypertension, vasoconstriction, and ischemia due to cocaine use [5]. Nevertheless, in our patient diagnosed with IVEM, no cause or risk factor was identified, including hypertension.

Gueunoun et al described the first case of idiopathic ERM complicated by neovascularization on OCT-A [6]. The authors emphasized the relevance of detecting an abnormal vascular complex inside an idiopathic ERM before proposing retinal surgery. This was the most comparable case report to ours as it shares various characteristics such as age, similar BCVA, and no past medical history, and it remained stable over time.

Giachos et al declared that it is feasible that intraretinal neovascularization in an idiopathic ERM occurred because of, or near, preexisting ERMs. Initially, they considered a perimacular branch retinal vein occlusion, a congenital retinal anomaly, an age-related macular degeneration (AMD) lesion, and macular telangiectasia as differential diagnoses [7]. However, they were ultimately ruled out until IVEM was the final diagnosis.

Considering the rarity of IVEM and the absence of well-established risk factors, it becomes pertinent to explore potential triggers and contributors that might influence their development. While the following suggestions are speculative and require further investigation, they may provide avenues for future research: (1) age-related factors, such as alterations in retinal microarchitecture or cumulative cellular damage; (2) ERM mechanical forces, as ERMs can harm macular capillaries through tangential and vertical forces over the retina; (3) VEGF regulation in ERMs, as Anguita et al proposed that abnormal vascularization can occur even in avascular membranes; (4) ocular microenvironment, like changes in retinal blood flow; (5) metabolic factors; (6) environmental exposures; and (7) genetic predisposition [1,3].
Managing IVEM can be challenging. Since it is rare, the only published case reports had different follow-up periods and treatment approaches that incorporated observation, anti-VEGF therapy, vitrectomy, and combined treatment [2,3,5-7]. Therefore, treatment outcomes may vary. Multimodal imaging could contribute to decision-making, especially when initiating anti-VEGF therapy, considering the potential risk of fibrovascular complex contraction, and monitoring the response to treatment over time [3].

Conclusions

This article adds valuable insights by presenting the case of a man with IVEM without identifiable risk factors, challenging existing hypotheses. The absence of hypertension or other known causes prompts a reevaluation of the current understanding of IVEM’s pathogenesis. Additionally, we propose potential triggers and risk factors, which require further research on this rare ocular condition to be confirmed. Their characterization by multimodal imaging could enable predicting their clinical course, thereby facilitating improved patient management.

Declaration of Figures’ Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

References:

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Figure 1. Multimodal imaging of the left eye. (A, B) Multicolor imaging shows a semi-translucent membrane in the superior temporal vascular arcade with an associated flat vascularization complex of thin vessels (white arrow). (C, D) Spectral domain optical coherence tomography reveals a thick pre-retinal hyper-reflective line with no vitreoretinal traction and a subtle irregularity in the inner and intermediate retina layers. See horizontal (C) and vertical (D) cross-section. (E) Fluorescein angiography exposes localized hyperfluorescence with leakage corresponding to the thin vessels observed (asterisk). No areas of capillary closure were identified. (F, G) Optical coherence tomography angiography shows a high-flow abnormal vascular complex with unbranched long dilated filamentous vessels similar to pruned-vascular-tree pattern in the superficial capillary plexus. Note how OCT-A en-face superficial cross-section (F) shows that the borders of the vascular complex have more fine vessels and how it exactly correlates with the high-flow observed in the OCT-A B-scan (G).