

Non-ischemic central retinal vein occlusion and paracentral acute middle maculopathy: Unveiling the central scotoma phenomenon.

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Abstract

Background: Central Retinal Vein Occlusion (CRVO) is a vascular disorder causing vision impairment, often associated with macular edema. Paracentral Acute Middle Maculopathy (PAMM) is a novel condition observed in CRVO cases, resulting in paracentral scotomas. We present a case of non-ischemic CRVO with PAMM.

Case Presentation: A 60-year-old male presented with an acute-onset central scotoma in the right eye. Examination revealed non-ischemic CRVO, and subsequent evaluation confirmed the presence of PAMM. Despite one-month follow-up, visual acuity and the scotoma did not improve. Imaging and functional tests supported the diagnosis of CRVO coexisting with PAMM. Low-dose aspirin was initiated.

Discussion: Non-ischemic CRVO can cause scotomas, including PAMM. PAMM manifests as hyper-reflective lesions in the outer plexiform and inner nuclear layers, resulting from retinal ischemia. Early identification of PAMM is crucial for understanding vision loss in CRVO cases without macular edema. Visual field testing and microperimetry aid in diagnosis and monitoring.

Conclusion: PAMM should be considered in non-ischemic CRVO cases with vision loss. Integrating imaging and functional tests enhances accurate diagnosis and management of CRVO-associated complications, leading to improved outcomes.

Keywords: Retinal vein occlusion, Maculopathy, Vision disorders.

Introduction

Central Retinal Vein Occlusion (CRVO) is a prominent cause of visual impairment characterized by the obstruction of retinal veins at the optic nerve, resulting in retrograde venous stasis, optic disc swelling, and, in certain cases, elevated luminal gradient pressure that can lead to compromised arterial blood flow [1].

Vision loss in CRVO is frequently attributed to macular edema [2]. However, additional etiologies have been associated with CRVO, including cilioretinal artery occlusion and the emerging entity of Paracentral Acute Middle Maculopathy (PAMM) [3]. PAMM typically arises from deep capillary ischemia, giving rise to a central or paracentral scotoma [4]. Accurate diagnosis of PAMM is challenging but important, as it can serve as an indicator of an impending complete Retinal Artery Occlusion (RAO) [5]. Key diagnostic modalities such as Spectral Domain Optical Coherence Tomography (SD-OCT), Visual Field (VF) analysis, and microperimetry play pivotal roles in the evaluation process [5].

Herein, we present a compelling case of a 60-year-old patient who presented with a paracentral scotoma and received a

diagnosis of non-ischemic CRVO. Despite one month of follow-up, the central scotoma persisted. Utilizing SD-OCT, VF analysis, and microperimetry, we obtained comprehensive data that substantiated the presence of concomitant PAMM, further reinforcing the diagnosis.

Case Presentation

A 60-year-old male presented at the Emergency Department with a sudden-Onset Central Scotoma in his left eye (OS). The patient had a medical history of mild hyperlipidemia managed by atorvastatin, but no other systemic or ophthalmological conditions were reported. Upon examination, the Best-Corrected Visual Acuity (BCVA) was measured as 20/20 OD and 20/32 OS, with equal and reactive pupils. No Relative Afferent Pupillary Defect (RAPD) was detected. Confrontation visual field assessment revealed an inferior scotoma in the right eye. Anterior segment examination appeared normal. Fundus examination displayed central retinal vein occlusion, characterized by scattered hemorrhages in the posterior pole and periphery [Figure 1].

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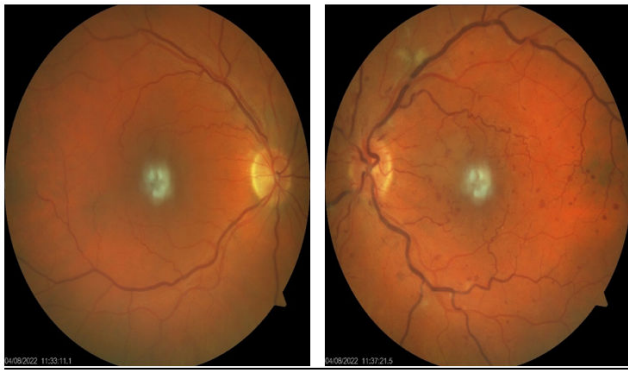


Figure 1: Colour Fundus photograph of the left eye showed multiple dot-blot hemorrhages in all four quadrants.

Optical Coherence Tomography (OCT) did not present macular edema, but it revealed an unnoticed hyper reflective band in the deep retinal layers [Figure 2].

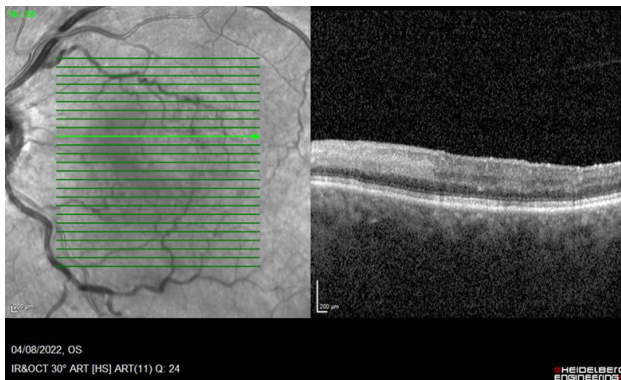


Figure 2: SD-OCT of the macula of the left eye demonstrating patchy perifoveal hyperreflectivity the inner plexiform and nuclear layers.

The patient was diagnosed with non-ischemic CRVO based on the absence of RAPD and retinal whitening in the periphery. At one-month follow-up, the visual acuity remained unchanged (20/20 OD and 20/32 OS), and the paracentral scotoma persisted. Reassessment of the OCT findings during the acute episode confirmed the presence of a hyper reflective lesion in the Outer Plexiform Layer (OPL) and Inner Nuclear Layer (INL), consistent with PAMM. Subsequent OCT performed at the one-month follow-up revealed retinal thinning in specific areas, suggestive of ischemia secondary to PAMM [Figure 3].

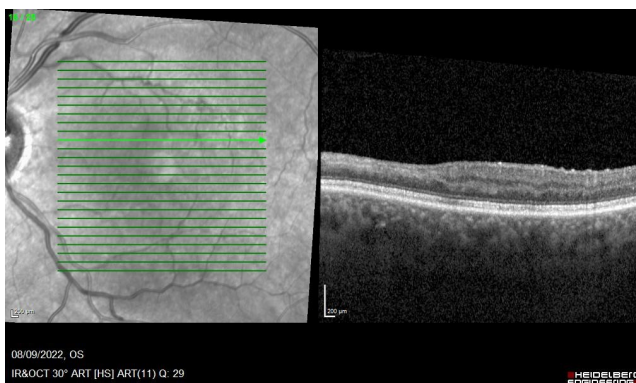


Figure 3: SD-OCT of the macula showing retinal thinning and atrophy in the areas consistent with the PAMM.

Visual field testing exhibited a scotoma extending from the right optic nerve blind spot to the foveal region [Figure 4].

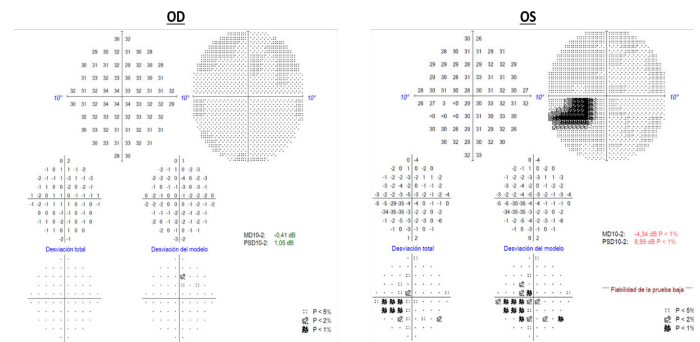


Figure 4: Central 10-2 Humphrey visual field testing of showing a pronounced nasal deficit consistent with the clinical findings.

Microperimetry results confirmed the reduced retinal sensitivity in regions corresponding to the observed retinal lesions on OCT [Figure 5].

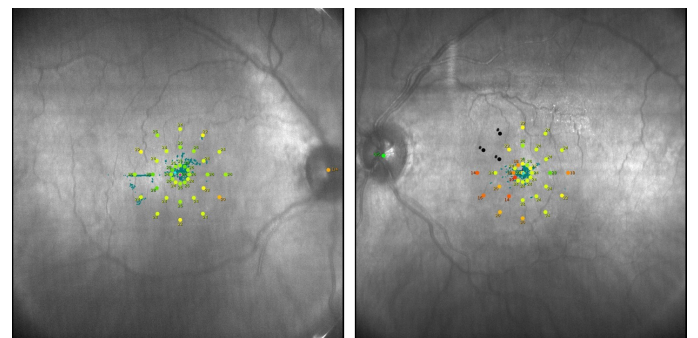


Figure 5: Microperimetry showing reduced retinal sensitivity in the superonasal area of the left eye

Based on these findings, the patient was diagnosed with CRVO coexisting with PAMM. Low-dose aspirin treatment was initiated, along with lifestyle recommendations.

Results and Discussion

Central Retinal Vein Occlusion (CRVO) is a vaso-occlusive event characterized by the presence of superficial and deep intraretinal hemorrhages, along with variable venous engorgement and optic disc swelling [6]. In certain cases, the occlusion of retinal veins can lead to an elevation in luminal pressure within the retinal capillary bed, surpassing that of the arterial system, which can subsequently result in ischemia and arterial occlusion [7].

While macular edema is the most common cause of vision loss in CRVO [2], it is essential to consider other potential etiologies, such as Retinal Artery Occlusion (RAO), Cilioretinal Artery Occlusion (CRAO), or the emerging entity known as Paracentral Acute Middle Maculopathy (PAMM) [8].

PAMM, initially described by Sarraf et al., is identified as a hyper-reflective band at the level of the Outer Plexiform Layer (OPL) and Inner Nuclear Layer (INL) on Optical Coherence

Tomography (OCT) [9]. These hyper-reflective lesions are believed to be a response to retinal ischemia in the deep capillary plexus. Previous reports have suggested that PAMM can be present in up to 5% of cases of non-ischemic CRVO [9].

In our patient's case, several factors were noteworthy. Firstly, the absence of cardiovascular risk factors, except for well-controlled hyperlipidemia, raised the possibility of alternative mechanisms contributing to CRVO [10]. Secondly, the patient's relatively young age was atypical for a CRVO presentation. Lastly, extensive systemic investigations yielded negative results.

It is important to note that PAMM lesions can be evanescent and may resolve before being clinically detected. Hence, early identification of PAMM at the time of diagnosis can provide valuable insights into the underlying cause of vision loss in non-ischemic CRVO cases. In challenging situations where OCT findings may be overlooked, particularly in cases with preserved best-corrected visual acuity or the absence of macular edema, Visual Field Analysis (VF) and microperimetry play crucial roles in diagnosing and monitoring the progression of PAMM.

Conclusion

Central Retinal Vein Occlusion (CRVO) is a vascular disorder affecting the eye, causing reduced vision or even sudden blindness. It occurs when the central vein that carries blood out of the retina becomes blocked, leading to a buildup of pressure and fluid leakage. The condition's exact cause isn't always clear, but risk factors include hypertension, diabetes, and glaucoma. Symptoms range from painless blurred vision to sudden vision loss, often in one eye. Early diagnosis through a comprehensive eye examination is important. Treatment aims to manage underlying conditions and prevent complications. Therapies such as anti-VEGF injections, laser treatment, or steroid injections can help alleviate symptoms and preserve vision. Prevention involves managing risk factors, maintaining a healthy lifestyle, and regular eye check-ups. Timely intervention and lifestyle modifications play an important role in managing CRVO, highlighting the significance of early detection and prompt treatment.

Consent

Verbal informed consent was obtained from the patient for publication of this case report and any accompanying images.

Acknowledgment

None

Conflict of Interest

The authors declare that they have no conflict of interest.

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