Tıp Fakültesi Klinikleri Cilt 6 • Sayı 2 • Temmuz 2023 (153 - 158)

Atypical Paracentral Acute Middle Maculopathy: A Case Report

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Abstract

Paracentral acute middle maculopathy (PAMM) is a recently described SD-OCT finding that is characteristically observed as a hyper-reflective band in the inner nuclear layer (INL) and typically evolves into INL atrophy in the later stages. The mechanism associated with PAMM is still under investigation, but there is a consensus that it is mainly related to ischemic hypoxia of the middle retinal layers. The aim of this case report is to present an atypical PAMM case with foveal and parafoveal involvement on OCT who presented with the complaint of blurred vision. A 48-year-old male patient presented with painless blurred vision in the right eye that started 10 days ago. He said that he had never experienced a similar complaint in both eyes before. He had no known systemic or ocular disease. In the ophthalmological examination, the best corrected visual acuity was 7/10 in the right eye and 10/10 in the left eye. Fundoscopic examination revealed slight retinal pigment epithelial changes in the temporal fovea of the right eye, and left eye fundoscopic examination resulted in normal. In the OCT image, a hyperreflective band was observed in the parafoveal inner nuclear layers in the temporal right fovea, OCT image of the left eye was normal. A central visual field defect was observed in the right eye.

PAMM, which is defined as a variant of acute macular neuroretinopathy affecting the inner nuclear and outer plexiform layers, can be diagnosed more easily today thanks to the development of SD-OCT and multimodal imaging methods. SD-OCT is a useful imaging method in the diagnosis, progression follow-up and differential diagnosis of PAMM. The role of focal parafoveal ischemia in the retinal capillary plexus in the etiology of PAMM has been emphasized, and it is recommended to investigate vasopressor exposure and microvascular pathologies that may affect the retina in patients diagnosed with PAMM.

Keywords: *Hypoxia, Retina, Maculopathy*

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Atipik Bir Parasantral Akut Orta Makulopati Olgusu

Özet

Parasantral akut orta makülopati (PAMM), karakteristik olarak iç nükleer tabakada (INL) hiper-reflektif bir bant olarak izlenen ve ilerleyen aşamalarda tipik olarak INL atrofisine dönüşen yakın zamanda tanımlanmış bir SD-OCT bulgusudur. PAMM ile ilişkili mekanizma hala araştırma konusudur, ancak esas olarak orta retina tabakalarının iskemik hipoksisine bağlı olduğu konusunda bir fikir birliği vardır. Bu olgu sunumunun amacı, bulanık görme şikayetiyle gelen OCT de foveal ve parafoveal alanda tutulumu olan atipik bir PAMM olgusunu sunmaktır. 48 yaşında erkek hasta sağ gözde 10 gün önce başlayan ağrısız bulanık görme şikâyeti ile başvurdu. Anamnezinde daha önce buna benzer bir şikâyeti iki gözü için de yaşamadığını belirtti. Bilinen sistemik ve oküler hastalığı yoktu. Herhangi bir travma, oküler cerrahi, sistemik veya oküler ilaç kullanımı öyküsü mevcut değildi. Yapılan oftalmolojik muayenede en iyi düzeltilmiş görme keskinliği sağ gözde 7/10 sol gözde ise 10/10 düzeyindeydi. Fundus muayenesinde sağ gözde fovea temporalinde retina pigment epitel değişikliği izlenmekteydi, sol göz fundus muayenesinde özellik yoktu. Çekilen OCT görüntüsünde sağ göz fovea ve parafoveal alanda iç nükleer katlarda hiperreflektif bant izlenmekteydi. Sol göz OCT görüntüsünde özellik yoktu. Görme alanında sağ gözde santral görme alanı defekti olduğu görüldü.

PAMM, SD-OCT ve multimodal görüntüleme yöntemlerinin gelişmesi sayesinde günümüzde daha kolay teşhis edilebilmektedir. PAMM etiyolojisinde retinal kapiller pleksusta fokal parafoveal iskeminin rolü üzerinde durulmuştur ve PAMM tanısı alan hastalarda vazopressör maruziyeti ve retinayı etkileyebilecek mikrovasküler patolojilerin araştırılması önerilmektedir.

Anahtar Kelimeler: *Hipoksi, Retina, Makulopati*

Introduction

Paracentral acute middle maculopathy (PAMM) is a recently described SD-OCT finding that is characteristically observed as a hyper-reflective band in the inner nuclear layer (INL) and typically evolves into INL atrophy in the later stages (1). PAMM that initially appears in the outer plexiform layer (type 1 AMN) has been accepted as a variant of acute macular neuroretinopathy (AMN); however, PAMM is currently considered a different variant from AMN (2).

The mechanism associated with PAMM is still under investigation, but there is a consensus that it is mainly related to ischemic hypoxia of the middle retinal layers (3-7). OCT-A studies have reported changes in vascular structure in the intermediate (ICP) and deep capillary plexus (DCP) (8,9). Use of vasoconstrictors such as caffeine and epinephrine, use of oral contraceptives and microvascular diseases affecting the retina such as diabetes, hypertension, and sickle cell anemia may cause such change (10,11).

This case report aims to present a PAMM

case with the complaint of blurred vision and with no risk factors.

Case Report

A 48-year-old male patient presented with painless blurred vision in the right eye that started 10 days ago. He said that he had never experienced a similar complaint in both eyes before. He had no known systemic or ocular disease. There was no history of trauma, ocular surgery, or systemic or ocular drug use. He has no history of smoking. He had received 3 doses of the Biontech covid vaccine, the last dose of which was 5 months ago. He said he had never been diagnosed with Covid and had not experienced any Covid- like symptoms recently.

In the ophthalmological examination, the best corrected visual acuity was 7/10 in the right eye and 10/10 in the left eye. The intraocular pressure was 13 mmHg in the right eye and 16 mmHg in the left eye. In the slit-lamp examination, the anterior segment was bilaterally natural. Fundoscopic examination revealed slight retinal pigment epithelial changes in the temporal fovea of the right eye and left eye fundoscopic examination resulted in normal.

In the OCT image (Cirrus OCT Zeiss), a hyperreflective band was observed in the parafoveal inner nuclear layers in the temporal right fovea (Figure 1). OCT image of the left eye was normal. A central visual field defect was observed in the right eye. (Loss of fixation, false positive and false negative percentages are below 20%.) (Figure 2). In the color fundus image of the patient, retinal pigment epithelial changes were observed in the temporal fovea of the right eye (Figure 3).

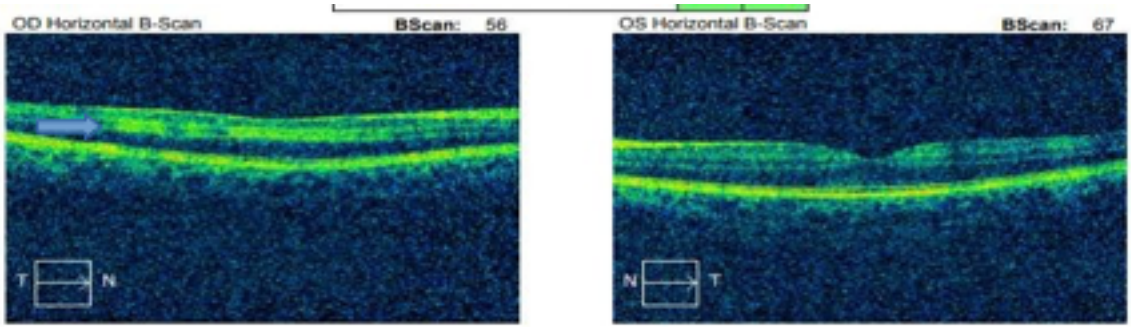

Figure 1. In the OCT image of the right eye, a hyperreflective area is observed in the area indicated by the arrow in the inner nuclear layer

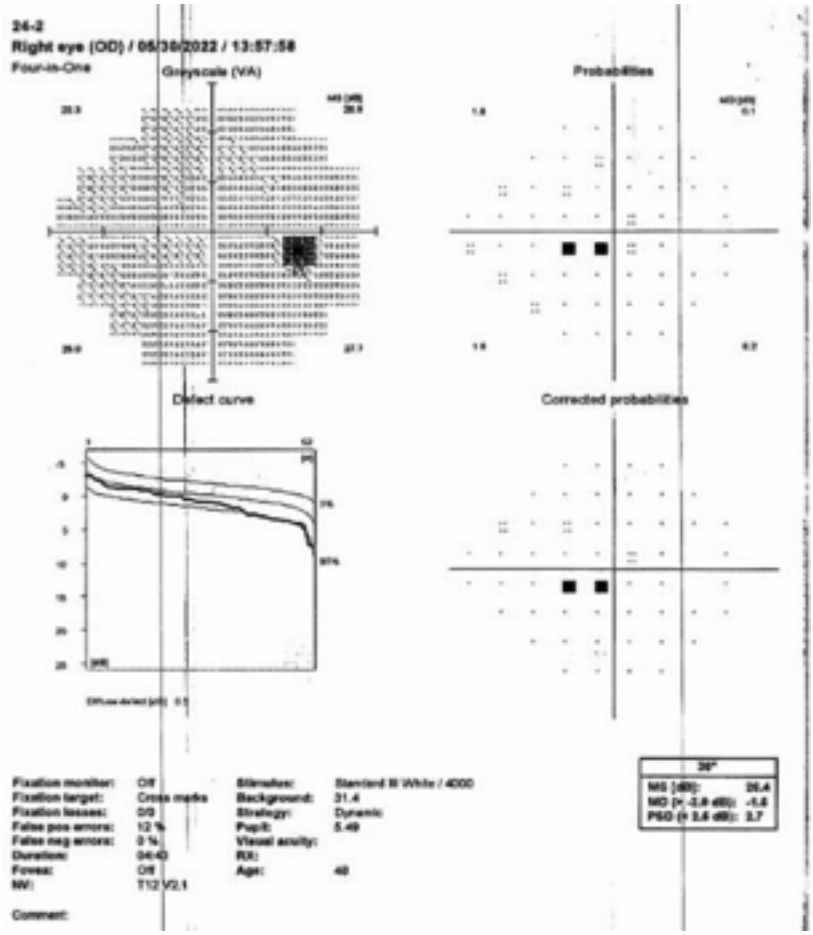

Figure 2. A central scotoma area is observed in the visual field of the right eye

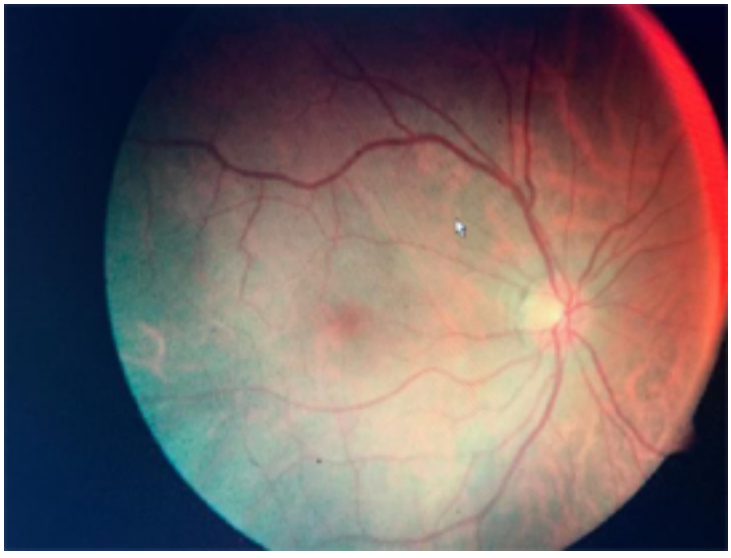

Figure 3. In the right eye color fundus image, retinal pigment epithelial changes are observed in the temporal fovea.

DISCUSSION

Sarraf et al. first described a PAMM lesion that is characterized by a hyper-reflective band in the INL in the acute phase, which can be seen on SD-OCT images, and progresses to atrophy of the INL in the chronic phase (12). The retinal capillary system supplies the neurosensory retina via 3 plexuses: superficial (Superficial Capillary Plexus, SCP), intermediate (Intermediate Capillary Plexus, ICP) and deep (Deep Capillary Plexus, DCP) capillary plexus. SCP is located within the ganglion cell layer while ICP and DCP are located at the inner and outer borders of the INL. Fundus fluorescein angiography (FA) can only show SCP; cannot demonstrate occlusion in ICP or DCP (12). SCP occlusions are associated with soft exudates, while ICP and DCP occlusions appear as a deep, grey-white lesion with more prominent edges. The presence of a hyperreflective band in the INL visible on SD-OCT suggests that a PAMM lesion may have developed as a result of ICP and/or DCP ischemia.

Although we could not detect any etiological factor in our case, in patients with PAMM lesions, the presence of microvascular diseases such as diabetes, hypertension, and sickle cell anemia in the etiology should be evaluated regarding their impact on the retina. In other words, PAMM may be a warning sign of systemic microvascular disease (13).

PAMM lesions have been associated with the use of sympathomimetics such as epinephrine, norepinephrine, ephedrine, and caffeine (14). Patients diagnosed with PAMM should also be asked about vasopressor exposure. Sympathomimetics may cause ischemia in the retinal middle and deep capillary plexuses due to their vasopressor impacts and may be responsible for the hyperreflective lesion in the inner nuclear and outer plexiform layers observed in SD-OCT. In a case series study by Ishibashi et al., four patients presented with BRAO-related visual field defects, one patient PAMM, and one patient AMN after receiving the Pfizer BioNTech COVID-19 vaccine. Although the Pfizer-BioNTech COVID-19 vaccine has not been shown to be associated with an increased risk of arterial embolism and thrombosis (15). The inner nuclear layer thinning that is experienced by some patients with chronic PAMM supports the ischemia theory. Paracentral scotoma may be permanent in patients whose inner nuclear layer is thinned in the chronic period.

In conclusion, PAMM, which is defined as a variant of acute macular neuroretinopathy affecting the inner nuclear and outer plexiform layers, can be diagnosed more easily today thanks to the development of SD-OCT and multimodal imaging methods. SD-OCT is a useful imaging method in the diagnosis, progression follow-up and differential diagnosis of PAMM. The role of focal parafoveal ischemia in the retinal capillary plexus in the etiology of PAMM has been emphasized, and it is recommended to investigate vasopressor exposure and microvascular pathologies that may affect the retina in patients diagnosed with PAMM. There is currently no cure for PAMM lesions, but etiological investigations are important to identify systemic vascular risk factors.

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