

Paracentral Acute Middle Maculopathy: A Case Report

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ABSTRACT

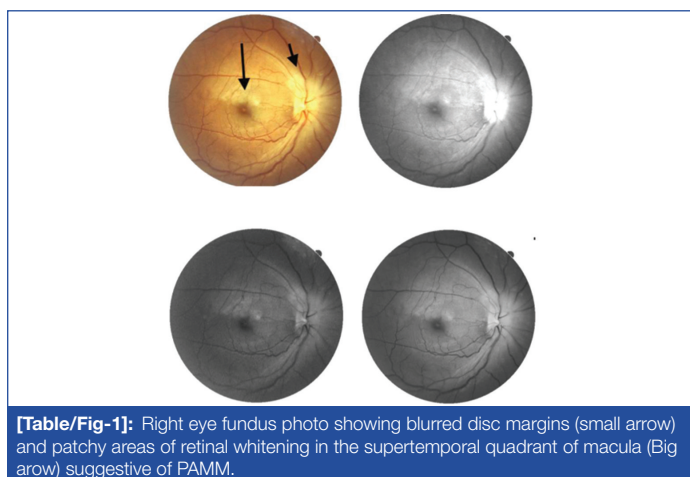
Paracentral Acute Middle Maculopathy (PAMM) is an optical coherence tomography finding observed in patients with retinal capillary ischaemia. It is a type of ischaemic maculopathy that affects the intermediate and deep retinal capillary plexuses. PAMM has been associated with various retinal vascular disorders such as retinal artery and venous occlusions, diabetic retinopathy, sickle cell anaemia, and Purtscher retinopathy. It can also occur following a flu-like illness and transorbital compression. The authors present a case of a 37-year-old healthy female patient who exhibited signs and symptoms suggestive of PAMM a few days after experiencing a self-limiting flu-like illness.

Keywords: Acute macular neuropathy, Disc drusen, Ischemic maculopathy, Macular optical coherence tomography, Parafoveal lesion

CASE REPORT

A 37-year-old woman presented with blurred vision in her right eye for the past two days, which was painless and sudden in onset. Upon detailed history-taking, it was revealed that she had experienced a mild fever and sore throat eight days before the ocular symptoms, for which she received symptomatic treatment. The illness subsided without the need for hospital admission. There was no significant medical or family history, and no previous episodes of a similar nature were reported.

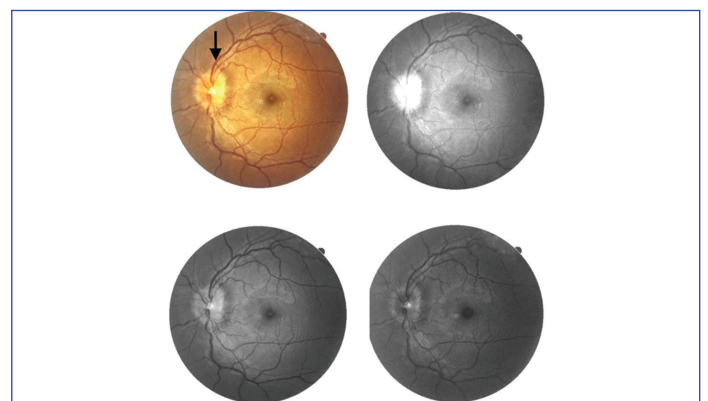
Her best corrected visual acuity was 6/9 in the right eye and 6/6 in the left eye. On slit lamp examination, the anterior segment was unremarkable in both eyes, with no anterior vitreous cells or flare noted. Fundus examination of the right eye showed blurred disc margins due to the presence of disc drusen and a patchy area of retinal whitening in the superotemporal quadrant of the macula [Table/Fig-1]. The left fundus photo revealed blurring of disc margins due to disc drusen, with a normal foveal reflex and retinal vasculature [Table/Fig-2].



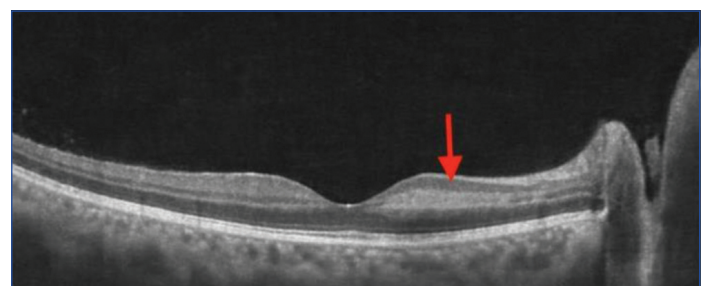
[Table/Fig-1]: Right eye fundus photo showing blurred disc margins (small arrow) and patchy areas of retinal whitening in the superotemporal quadrant of macula (Big arrow) suggestive of PAMM.

The Spectral Domain Optical Coherence Tomography (SD-OCT) of the right eye revealed a hyperreflective band at the middle retinal layers of the retina [Table/Fig-3]. The patient was advised to undergo fundus fluorescein angiography, but she refused.

The patient was conscious, well-oriented, and had a completely unremarkable systemic examination. There was no history of jaw claudication or any pulsatile artery upon examination. The patient did not have a history of Coronavirus Disease-2019 (COVID-19)-related



[Table/Fig-2]: Left eye fundus photo showing blurred disc margin due to disc drusen.



[Table/Fig-3]: SD-OCT showing hyperreflective band at the middle retinal layers (as shown by the arrow).

illness and had been vaccinated two years prior. Haematological and serological investigations were negative for dengue, malaria, and typhoid. Complete blood count, Liver Function Test (LFT), Kidney Function Test (KFT), blood sugar, and lipid profile were all within normal limits. Tests for Human Immunodeficiency Virus (HIV), Hepatitis B surface antigen (HBsAg), Venereal Disease Research Laboratory (VDRL), and Treponema Pallidum Haemagglutination Assay (TPHA) were all negative. A detailed cardiological work-up, including carotid doppler and echocardiography, revealed normal results. A diagnosis of PAMM was made, and since the patient was stable systemically, it was decided to observe the patient and schedule a review after four weeks. However, the patient was lost to follow-up thereafter.

DISCUSSION

The PAMM, as first described by Sarraf in 2013 is a variant of Acute Macular Neuropathy (AMN) with a possibility of vascular aetiology

affecting intermediate and deep retinal capillaries [1,2]. PAMM shows hyperreflective bands which affects the Inner Plexiform Layers (IPL), Inner Nuclear Layer (INL), and Outer Plexiform Layer (OPL) on SD-OCT [3].

The PAMM is caused by ischaemia and is associated with various retinal vascular disorders such as diabetic retinopathy, central retinal vein occlusion, and retinal artery occlusion, as described in the literature [4]. The literature has also reported a role of inflammation and infection in its pathogenesis [5]. Therefore, besides retinal vascular disorders, other causes of PAMM include flu-like upper respiratory tract infection, Purtscher's retinopathy, inflammatory occlusive retinal vasculitis, post-H1N1 vaccine complications, migraine disorders, and blunt trauma causing ocular ischaemia. The inability of the deep capillary plexus and choriocapillaris to autoregulate vascular flow changes in the settings of a sudden blood pressure change is suggested as the pathophysiology behind the focal inner retinal atrophy seen in patients with acute macular neuroretinopathy and PAMM. Findings on Optical Coherence Tomography Angiography (OCTA) in patients with acute macular neuroretinopathy have shown flow loss in the deep capillary plexus and choriocapillaris [6].

Although earlier considered to be an inner retinal disease, the aetiology of PAMM is likely complex and not completely understood. Infection, inflammation, and ischaemia have all been considered in the pathogenesis of PAMM. After reviewing the published literature on PAMM and acute macular neuropathy, which are considered to be variants of the same spectrum associated with vascular aetiology [7], it appears that PAMM should actually be considered a subtle sign that can only be seen on imaging. With the advent of advanced imaging technologies like OCTA, PAMM can now be more easily appreciated as it affects a particular layer of retinal vasculature that was difficult to diagnose with earlier imaging modalities like fundus fluorescence angiography.

There is no treatment for PAMM as it has been associated with vasculopathies. However, it is important for patients with PAMM to get screened for other local and systemic diseases. Careful follow-up is also important. As mentioned earlier, various rare causes of PAMM have been reported in the literature, such as birdshot chorioretinopathy, post-viral Purtscher-like retinopathy, retinal vasculitis due to Behçet's disease, and post-H1N1 vaccination [8]. The authors report a case of PAMM in a healthy middle-aged female patient secondary to a possible nonspecific flu-like illness with associated bilateral disc drusen appearing as pseudo disc oedema. They evaluated the patient in detail, but no systemic cause was identified. So, PAMM can be secondary to a mild flu-like illness. The patient was treated symptomatically and did not undergo any hospital admission, indicating that the illness was of a very mild nature, yet it caused significant changes in the retinal vasculature. Unfortunately, as with retinal artery occlusion, there is

no effective treatment for PAMM, and it is difficult to completely recover vision [9]. The present case is also associated with disc drusen, which gives a false appearance of disc oedema. Earlier, a single case report of PAMM with bilateral disc swelling secondary to meningitis has been described in the literature [10]. The patient was lost to follow-up, and it would be helpful if the authors could have OCTA and fundus fluorescein angiography images for proper documentation.

CONCLUSION(S)

The present case report concludes that PAMM can occur in healthy young individuals with no significant systemic and ocular vascular causes. Being a subtle finding, it can easily be missed on examination, and hence we need to be more vigilant in diagnosing this condition to avoid other unnecessary ocular investigations. Unfortunately, there is no treatment for this condition, and treatment of the systemic factors does not result in a reversal of vision. Still, the available literature on PAMM is limited, and more studies with histopathological correlation are needed to define its exact pathophysiology.

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AUTHOR DECLARATION:

- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

PLAGIARISM CHECKING METHODS: [Jain H et al.]

- Plagiarism X-checker: Mar 25, 2024
- Manual Googling: Apr 29, 2024
- iThenticate Software: May 16, 2024 (16%)

ETYMOLOGY: Author Origin

EMENDATIONS: 6

Date of Submission: **Mar 24, 2024**
Date of Peer Review: **Apr 27, 2024**
Date of Acceptance: **May 17, 2024**
Date of Publishing: **Jul 01, 2024**