Optical Coherence Tomography Angiography and Multifocal Electroretinogram Findings in Paracentral Acute Middle Maculopathy

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Abstract:
Paracentral acute middle maculopathy (PAMM) is an optical coherence tomography (OCT) finding seen in patients with retinal capillary ischemia. In this case report, we present a case of PAMM after a transient central retinal artery occlusion and the multifocal electroretinogram (mfERG) and other multimodal imaging findings. Clinical examination, OCT angiography, OCT en face, fluorescein angiography, and visual fields were performed at the baseline and follow-up examinations. As a result, we identified in this PAMM case evidence of hypoperfusion in both the choriocapillaris as well as the deep capillary plexus. To the best of our knowledge, the involvement of choriocapillaris has not been reported previously in the literature. Moreover, we concluded that mfERG constitutes a useful investigation in PAMM and this is the first mfERG findings to be presented for a PAMM case specifically.

Keywords:
Multifocal electroretinogram, optical coherence tomography angiography, paracentral acute middle maculopathy, transient central artery occlusion

Introduction
Paracentral acute middle maculopathy (PAMM) is a newly introduced term, confirmed by the advancements in ophthalmological investigations. It is an optical coherence tomography (OCT) finding seen in patients with retinal capillary ischemia in vascular-related diseases.[1,2] The affected retinal component in PAMM is the intermediate (ICP) and deep capillary plexus (DCP).[1,2] at the level of outer plexiform and inner nuclear layers (INLs) were the OCT shows hyperreflectivity and the OCT angiography depicts hypoperfusion in the DCP.[1,3]

In this case of PAMM, we report evidence of the involvement of choriocapillaris as well as the DCP on the OCT angiography and the multifocal electroretinogram (mfERG) findings.

Case Report
A 48-years-old male was referred to our department complaining of transient sight loss of his right eye for 12 h. He had no previous medical or ophthalmic history.

At the initial presentation, the patient had best-corrected visual acuity (BCVA) <1/20 and 20/20 in the right and left eye, respectively. There was an evident relative afferent pupillary defect (RAPD) in his right eye. Clinical examination and several
investigations performed, including blood tests, CT and MRI of the head, and orbits, which were all within the normal limits. His visual acuity improved gradually and his BCVA became 20/20 in the affected eye at the 5th day, the RAPD, however, persisted.

He had a reliable Humphrey’s visual field test, Central 30-2 threshold test, (Humphrey Field Analyzer II, Carl Zeiss Meditec, Dublin, Calif., USA), which revealed a central scotoma in the right eye. He also had a short wavelength autofluorescence imaging (Heidelberg Spectralis, Heidelberg, Germany) which showed perimacular hypoauflorescent areas with a petaloid pattern. The fluorescein angiography (FA) (Heidelberg Spectralis, Heidelberg, Germany) revealed a delay in the retinal arterial circulation with a delay in the arterial phase of 20 s after the choroid circulation, the filling of the vessels was complete without indication of specific occlusion in the arterial tree.

The macular OCT images (DRI OCT Triton Plus; Topcon, Tokyo, Japan) of the affected eye showed hyperreflective areas at the level of INL [Figure 1a]. Similarly, the en face OCT images showed fern-like perimacular lesions at the DCP and outer retina with gradual improvement [Figure 1b]. Interestingly, the OCT angiography images showed hypoperfusion in both the choroidal and DCP, which was significantly improved in the re-examination after 6 days [Figure 2b].

The patient had electrodiagnostic tests and a (mfERG Flash FOK program) which were performed to identifying any underlying causes. The multifocal electroretinogram (mERG) was recorded with the VERIS III (Visual Evoked Imaging System, Tomey, Nagoya, Japan). The findings of the mfERG showed decreased retinal responses. Both trace array and three-dimensional response density plot revealed macula dysfunction within the 5° and 10° zone without demonstrating a characteristic retinal artery occlusion pattern [Figure 2a]. Up to our knowledge, there has not been any mfERG findings description in the literature specifically for PAMM.

Discussion

The causes of PAMM are mainly vascular related.\(^2,3\) The pathogenesis is attributed to ischemia in the ICP and DCP.\(^1,4\) It is believed that the deep location of these plexuses is the reason why FA cannot depict the ischemic lesions adequately,\(^5\) and thus FA is not always diagnostic of PAMM. It has been proposed in PAMM cases, that OCT can better demonstrate and evaluate the grade and the level of the retinal ischemia.\(^1,6\) In our case, the multimodal imaging results, specifically the OCT images, and advocated the diagnosis of PAMM.

In our case, we believe that FA has a significant role in terms of the pathophysiology of PAMM. The delayed filling of the retinal arteries indicates an occlusive disorder of the retinal arteries, which we describe as a transient central retinal artery occlusion (CRAO). Yu et al., proposed that the combination of the characteristic OCT findings in PAMM and the lack of angiographic findings of arterial occlusive disorder, maybe attributed to reperfusion of a previous CRAO or vasospasm.\(^4\) They also suggested that DCP may be more susceptible to ischemia than other parts of the retina due to its location in the watershed zone with limited oxygen supply.\(^4\)

OCT angiography plays a crucial role in the diagnosis of PAMM. It is known so far that PAMM indicates ischemia in the ICP and DCP.\(^1,3\) Interestingly, in our case, OCT angiography shows ischemia in the choriocapillaris as well as DCP.

It has been reported, that the OCT en face images were used for follow-up and classification and a characteristic fern-like pattern was described, in correlation with central retinal vein occlusion causes only.\(^3\) In our case, which is of arterial occlusion, the OCT en face, Interestingly, did reveal the same fern-like pattern.

Finally, we believe that mfERG is useful in the diagnosis of PAMM, showing macular involvement with distorted macular potentials and an abnormal non-specific pattern. To the best of our knowledge, there have not been any descriptions of mfERG in PAMM in literature. Yusuf et al. proposed that mfERG may contribute in the diagnosis of localized macular ischemia, especially in the suspected transient retinal artery occlusion without other evidence.\(^6\) They suggest that retinal ischemia after vasospasm or thromboembolism episode of the retinal artery may remain after the initial insult and can be recorded by mfERG. Hence, in our case, the decreased mfERG macular responses without a characteristic pattern suggest transient CRAO, which is supported by the delay in the retinal arterial filling time in the FA and the presence of RAPD.
In conclusion, in this PAMM case report, apart from the typical OCT hyperreflective bands and involvement of the DCP, we noticed additionally the interesting involvement and hypoperfusion of the choriocapillaris shown on OCT angiography, suggesting the involvement of two separate layers in the pathophysiology of PAMM.

This case also suggests that the use of mfERG is useful in the diagnosis of PAMM and its possible cause alongside FA, which to our knowledge is the first description and reported results of mfERG in a case of PAMM.

The need for more PAMM cases with mfERG is required to establish specific characteristics and comparative studies are needed subsequently to determine its specificity and sensitivity.

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**Conflicts of interest**
There are no conflicts of interest.

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