Paracentral Acute Middle Maculopathy: a novel clinical finding

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The advent of retinal multimodal imaging and the rapid advances in high-resolution imaging systems have not only improved the understanding of the health and diseased retina, but also allowed for the recognition of new clinical findings, such as paracentral acute middle maculopathy (PAMM)¹-³.

PAMM was first described in 2013 by Sarraf et al.¹ as a new variant of acute macular neuroretinopathy (AMN) associated with retinal capillary ischemia. In the original report, all patients presented with the typical findings of AMN such as an acute paracentral scotoma associated with a gray wedge-shaped paracentral lesion in near-infrared imaging (NIR). The differentiating features were the spectral-domain optical coherence tomography (SD-OCT) findings. While some patients showed hyperreflective bands in the outer plexiform layer (OPL)/inner nuclear layer (INL) region with subsequent INL thinning and sparing of the outer retina - a novel finding - and were classified as having type 1 AMN, also referred to as PAMM, others had hyperreflective bands in the OPL/outer nuclear layer (ONL) region with subsequent ONL thinning and ellipsoid disruption, and thus were termed type 2 AMN (classic AMN). These two different tomographic presentations (above or below the ONL), were hypothesized to be related to the occlusion of the superficial capillary plexus in type 1 AMN/PAMM or deep capillary plexus in type 2 AMN.⁴ The PAMM variant was also noted to occur more frequently in older male patients with vasculopathic risk factors⁵,⁶.

However, after the recent description of PAMM as a subtype of AMN characterized by a hyperreflective band-like SD-OCT lesion affecting the middle layers of the retina at the level of the INL because of superficial capillary -lary plexus ischemia,⁷,⁸ it has now been recognized that the lesion instead relates to ischemia of the intermediate and deep retinal capillary plexuses (deep capillary ischemia), anatomically located at the inner and outer zones of the INL, respectively, and represents a nonspecific finding of retinal ischemia⁹-¹⁴.

Indeed, it has been shown that PAMM can also occur in the setting of various retinal vascular diseases, such as diabetic retinopathy, retinal vein occlusion, retinal artery occlusion, sickle cell retinopathy and Purtsher’s retinopathy.⁵,⁷,¹¹. In this context, SD-OCT is an invaluable tool as it improves the in vivo visualization of the retinal microcirculation and allows for a more precise location of the retinal ischemia. It also permits the differentiation between the “cotton-wool spot”, a lesion of superficial capillary plexus ischemia, and PAMM, consequence of intermediate and deep retinal capillary plexuses ischemia. This latter assumption has been recently confirmed by Sarraf and associates using OCT angiography. Nevertheless, the exact physiopathological mechanism underlying PAMM lesions is still not completely understood. It could be that the deep capillary plexus is located in an oxygen supply watershed zone and therefore more vulnerable to ischemia or that the paucity of superficial capillaries in the perifovea is responsible.⁵,⁷

As the clinical manifestations of PAMM can be subtle, the diagnosis should be made by NIR reflectance and SD-OCT.⁵,⁶ Fundus autofluorescence can also be helpful (Fig.1A, D and G). NIR allows for the visualization of a sharply demarcated dark wedge-shaped lesion, that fades with serial follow-up imaging (Fig. 1B, E and F). SD-OCT demonstrates the typical hyperreflective lesion in the acute phase that resolves with thinning of the INL (Fig.1 C, F and I). This thinning is usually accompanied by persistent scotomas. It should be noted that conventional fluorescein angiography is usually unremarkable, as it cannot properly assess the intermediate and deep capillary plexus. There is no current treatment for PAMM, except for the recognition and treatment of possible associated vasculopathic risk factors.

In conclusion, although just recently described, PAMM is now widely accepted to be a novel SD-OCT sign of retinal deep capillary plexus ischemia, which may occur in isolation or complicating an
Patient with an isolated paracentral acute middle maculopathy (PAMM) lesion in the right eye. Baseline autofluorescence (AF) and near infra-red (NIR) reflectance images showed a paracentral dark wedge-shaped lesion (arrowhead), respectively (A) and (B). Same visit spectral-domain optical coherence tomography (SD-OCT) revealed a characteristic hyperreflective lesion (arrowhead) at the level of the inner nuclear layer (C), consistent with PAMM. Follow-up imaging at 1 month demonstrated attenuation of the lesion in both AF and NIR, respectively (D) and (E), with subsequent resolution at 3 months, (G) and (H). SD-OCT at 1 and 3 months, showed gradual resolution of the PAMM lesion (F) with following severe INL thinning (I) (between arrowheads).

underlying retinal vascular disorder. Ophthalmologists should be aware of PAMM as it is more common than previously believed and may be responsible for non-specific visual complaints, especially when the examination is unremarkable.

**REFERÊNCIAS**


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DE INJEÇÕES FREQUENTES NO TRATAMENTO DO EMD CRÔNICO 1,2

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* Em média, doentes com EMD crónico receberam 1,3 implantes no período de 3 anos. 2

1. NORE DE MEDICAÇÃO LUVEN é um medicamento indicado na efeitos colaterais comuns de medicamentos.
2. INFORMAÇÕES CLÍNICAS 6 é indicado como um tratamento de escolha em doentes com a principal queixa associada ao EMD crónico diabético crónico, que não respondem adequadamente à terapia de treinamento com luz de alta intensidade (LIT). O tratamento com a solução LUVEN deve ser administrado por um profissional de saúde qualificado e treinado. O princípio ativo do medicamento é o 4-hidroxi-4-(2-hidroxi-3-fenil-3-fenilpropano) (400 mg/mL), que é administrado por via intramuscular. O medicamento deve ser administrado de acordo com as indicações do tratamento e a presença de complicações graves, a avaliação dos benefícios e riscos do tratamento. O paciente deve estar consciente dos efeitos colaterais possíveis e deve ser instruído sobre a importância do uso de medicamentos. O paciente deve ser orientado sobre a necessidade de usar medicamentos adequadamente e de relatar qualquer efeito colateral ou reação adversa.
3. Instruções de uso: Após a aplicação do medicamento, o paciente deve ficar com o local aplicado limpo e não deve se expor a radiação ou radiação de alta intensidade. O medicamento deve ser administrado da forma indicada e a cada três meses. O paciente deve ser orientado sobre a importância de usar medicamentos adequadamente e de relatar qualquer efeito colateral ou reação adversa.

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Acetonido de Fluocinolina
190 microgramas
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Referências:
1. ILUVEN. Resultados dos estudos clínicos 1-2, 2015. Composição do medicamento 1,2.