Macular edema in multiple evanescent white dot syndrome

Edema macular na síndrome dos múltiplos pontos brancos evanescentes

Raul N. G. Vianna1,2, André Soares Maia1, Leticia Rielo de Moura1, Ana Lúcia Peixoto1, Hugo Soares Maia1

ABSTRACT

Eletroretinographic findings show that the transient decreased vision seen in patients with the multiple evanescent white dot syndrome (MEWDS) is related to metabolic disturbances at the level of the retinal pigment epithelium and photoreceptor complex. In this paper, we present a patient with a typical picture of MEWDS associated to macular edema, which could also be a factor to reduce vision in this disorder. Case report of a 53-year-old woman complaining about unilateral decreased vision of 7 days’ duration was sent to our retina clinic. A complete ophthalmic examination was performed as well as fluorescein angiography, indocyanine green angiography, Goldmann visual fields and optical coherence tomography (OCT). Best-corrected visual acuity was 20/40 and 20/20, in the right and left eye respectively. Ophthalmoscopy of the affected eye revealed multiple focal outer retinal gray lesions in the perimacular and peripapillary region. There were several orange punctate lesions in the foveolar region. Fluorescein angiography disclosed faint multiple foci of staining in the perimacular and peripapillary area, and some staining of the optic disc. A discrete hyperfluorescence was also observed in the foveal region. OCT disclosed an increase in foveal thickness (231 µm), approximately 25% thicker than the opposite normal eye (186 µm). Within 3 months her visual acuity had returned to 20/20 and the foveal thickness returned to a normal value (189 µm). Although the mechanism of transitory blurred vision is not completely elucidated in cases of MEWDS, we suggest that macular edema may play a role.

Keywords: Electroretinography; Macular edema; Retinal diseases; Vision disorders; Syndrome; Case reports

RELATO DE CASO

Raul N. G. Vianna1, André Soares Maia1, Leticia Rielo de Moura1, Ana Lúcia Peixoto1, Hugo Soares Maia1

1Uveitis, Retina and Tumors Unit, Instituto Brasileiro de Oftalmologia – IBOL - Rio de Janeiro (RJ), Brazil;
2Department of Ophthalmology, Hospital Universitário Antonio Pedro, Universidade Federal Fluminense – UFF – Niterói (RJ), Brazil;

From the Uveitis, Retina and Tumors Unit, Instituto Brasileiro de Oftalmologia – IBOL - Rio de Janeiro (RJ), Brazil.

Recebido para publicação em: 8/12/2009 - Aceito para publicação em 10/2/2010

INTRODUCTION

Multiple evanescent white dot syndrome (MEWDS), initially described in 1984, is an acute, multifocal, usually unilateral retinopathy affecting young adults.

Eyes with MEWDS usually show several yellow-white dots at the level of the deep retina or retinal pigment epithelium (RPE). The visual acuity ranges from 20/20 to 20/40 and most patients complain of the presence of a scotoma and associated photopsias. Besides the typical retinal lesions, a characteristic granular appearance to the fovea is present acutely, and the fovea usually does not return to a normal appearance. The exact mechanisms of visual loss in MEWDS are not well understood but may represent photoreceptor, RPE, and optic nerve dysfunction.

In this paper we present the optic coherence tomography (OCT) findings of a patient with active MEWDS. Based on these findings we suggest that macular edema may play a role in the transient decreased visual acuity seen in patients with this disorder.

Case report

A 53-year-old woman noted photopsia and blurred vision in her right eye. Her visual acuity was 20/40 in the right eye (RE) and 20/20 in the left eye (LE). Biomicroscopic examination revealed a mild inflammatory reaction (+/4 cells) in the anterior chamber of the RE. The intraocular tension was inside normal limits in both eyes. The vitreous humor showed a few cells (trace). Ophthalmoscopy of the affected eye revealed multiple focal outer retinal gray lesions in the perimacular and peripapillary region (figure 1A). There were several orange punctate lesions in the foveolar region. Fluorescein angiography disclosed faint multiple foci of staining in the perimacular and peripapillary area, and some staining of the optic disc. A discrete hyperfluorescence was also observed in the foveal region (figure 1B). Indocyanine green angiography (ICG-A) showed multiple nonfluorescent lesions in the choroid and in the peripapillary region, in a pattern similar to the areas of staining in the fluorescein angiogram (figure 1C). Goldmann visual field of the affected eye revealed an enlarged blind spot (figure 1D) and the electroretinogram showed a decreased a-wave. These features supported the diagnosis of MEWDS.

To evaluate the macular structure we performed OCT (Zeiss-Stratus 3), which disclosed an increase in foveal thickness (231 µm), approximately 25% thicker than the opposite normal eye (186 µm) (figure 2). The total macular volume was 7.73 mm³ in the affected eye and 6.86 mm³ in the normal eye. These features led to the diagnosis of macular edema in the RE. The patient was followed monthly and within 3 months, the edema resolved.
months her visual acuity had returned to 20/20. Foveal thickness and total macular volume also returned to normal values (189 µm and 6.96 mm³ respectively).

Comments

It has been suggested that the decreased vision observed in eyes with MEWDS may be related to transient metabolic disturbances at the level of the retinal pigment epithelium – photoreceptor complex. Indeed, during the acute phase of the disease, the ERG a-wave and early receptor potential amplitudes are profoundly decreased in most affected patients, which suggests a primary involvement of the outer segments of photoreceptors. In addition, Keunen and van Norren have used foveal densitometry and color matching to show that, even in those few patients with normal ERG findings, abnormalities exist during the active stage of MEWDS at the level of the cone photoreceptor outer segments. Nevertheless, focal ERG studies revealed delayed recovery of oscillatory potential, which also implies some inner retinal involvement.

Using Stratus-OCT III, Chan et al. estimated that the normal foveal thickness was 182 ± 23 µm. The OCT of our case disclosed an increase in foveal thickness (231 µm, approximately 25% thicker than the opposite normal eye, 186 µm), supporting the diagnosis of macular edema. It must be pointed out that large differences of the foveal thickness between eyes in patients with macular disease may represent a true derangement from normality. Indeed, it has been shown that there is striking symmetry of the foveal thickness between the eyes of each normal individual.

Moreover, varying degrees of intraocular inflammation have been reported in patients with MEWDS. Thus, there is a rationale for the development of inflammatory macular edema in these eyes. Although the mechanism of transitory blurred vision is not completely elucidated in cases of MEWDS, we suggest that macular edema may play a role.

RESUMO

Propósito: Achados eletroretinográficos revelam que a baixa visual transitória observada em pacientes com a síndrome dos múltiplos e evanescentes pontos brancos na retina (MEWDS) está relacionada a distúrbios metabólicos ao nível do epitélio pigmentado da retina e dos fotorreceptores. No presente artigo, nós apresentamos um paciente com um quadro típico de MEWDS associado a edema macular, o qual pode ser um fator que reduz a acuidade visual. Método: Using Stratus-OCT III, Chan et al. estimated that the normal foveal thickness was 182 ± 23 µm. The OCT of our case disclosed an increase in foveal thickness (231 µm, approximately 25% thicker than the opposite normal eye, 186 µm), supporting the diagnosis of macular edema. It must be pointed out that large differences of the foveal thickness between eyes in patients with macular disease may represent a true derangement from normality. Indeed, it has been shown that there is striking symmetry of the foveal thickness between the eyes of each normal individual.

Moreover, varying degrees of intraocular inflammation have been reported in patients with MEWDS. Thus, there is a rationale for the development of inflammatory macular edema in these eyes. Although the mechanism of transitory blurred vision is not completely elucidated in cases of MEWDS, we suggest that macular edema may play a role.

REVIEWS


Correspondence to:
Raul N. G. Vianna
Rua General Mariane, nº 88/802
CEP 22221-100 - Laranjeiras – RJ
Phone/Fax: 55 (21) 2610-1051
E-mail: raulngvianna@ig.com.br