Case Report

Optical coherence tomography in resolution of photoreceptor damage in multiple evanescent white dot syndrome

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Abstract

Multiple evanescent white dot syndrome (MEWDS) is an acute-onset chorioretinal inflammatory disease. This disorder is characterized by unilateral multiple gray-white dots of the posterior pole in young healthy women. Symptoms include blurred vision, photopsia, and visual field (VF) defects. Although the etiology of MEWDS is still unknown, most patients have spontaneous improvement in vision and fundus appearance within a period of weeks. Herein, we report a typical case of MEWDS, and describe the spontaneous resolution of photoreceptor damage during the entire course, demonstrated by serial optical coherence tomography (OCT). The OCT is a sensitive and noninvasive tool for the diagnosis and serial microstructure evaluation of patients with MEWDS.

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1. Introduction

Multiple evanescent white dot syndrome (MEWDS) is an acute-onset chorioretinal inflammation which is characterized by unilateral multiple gray-white dots on the posterior pole in young healthy women. This disease entity was first reported by Jampol et al in 1984.1 Symptoms include blurred vision, photopsia, and visual field (VF) defects. Fluorescein angiography (FA) shows multiple hyperfluorescent dots with late staining.2 Enlarged physiologic blind spot may be seen in VF testing. Electroretinography (ERG) tests reveal photoreceptor dysfunction with decreased a-wave amplitude.3 Most patients have spontaneous improvement in vision and fundus appearance within a period of weeks. Although the clinical feature of MEWDS is well described, its etiology is still unknown. Recently, optical coherence tomography (OCT) has provided a facilitative method for in vivo cross-sectional retinal microstructure assessment.4 Stratus OCT image, with a 10-µm axial resolution, shows a highly reflective line on the border between sensory retina and retinal pigmented epithelium (RPE) layer. Results of ultrahigh-resolution OCT confirms that this reflective line is the junction of the photoreceptor’s inner and outer segments (IS/OS).5,6 Herein, we report a typical case of MEWDS, and describe the spontaneous resolution of photoreceptor damage during the entire course, demonstrated by serial OCT imaging.

2. Case report

A 21-year-old myopic female presented to our clinic with a complaint of sudden blurred vision with scotoma and flashes in her left eye for 2 days. She denied any systemic diseases or preceding upper respiratory tract infection (URI)-like illness. The initial best corrected visual acuity (BCVA) was 6/6, with −4.75
diopters (D) in the right eye and 6/60 with −4.5 D in the left eye. Intraocular pressure was 19 mmHg in the right eye and 20 mmHg in the left eye. Movement of both eyes was full and free. The relative afferent pupillary defect was negative in the right eye and positive in the left eye. Color test was 15/15 in the right eye and 1/15 in the left eye.

When examined using a slit lamp, her cornea was clear and there was no cell or flare in the anterior chamber or vitreous cavity. Fundus examination revealed multiple gray-white spots deep in the retina located at the macula and peripapillary area in the left eye (Fig. 1A). The macula had tiny yellow dots with foveal granularity appearance in the lesion eye. FA showed early hyperfluorescence, and late staining of these white dots in the left eye (Fig. 1B and C). The late dye leakage and staining from retinal vessels and optic disc were secondary to the acute chorioretinal inflammation. Results of VF testing (Humphrey 30-2) revealed enlarged blind spot and a central scotoma in the central 20° field with a 13.28-dB decrease in mean deviation in the left eye (Fig. 2A). Magnetic resonance imaging showed no significant signal change in bilateral optic nerves. Stratus OCT (Carl Zeiss Meditec, Inc., Dublin, CA) showed a focal disruption of the photoreceptor IS/OS junction line with a 5-mm horizontal line scan crossing the fovea in the left eye (Fig. 3A), and no abnormality in the right eye (data not shown). Scotopic and photopic full-field flash ERGs were performed. The amplitude of maximal rod response in the affected eye was significantly smaller compared with the nonaffected eye (Table 1). The ERG a-wave amplitude of photoreceptor cone response in the affected eye was significantly decreased compared with the nonaffected eye (Table 1). Results of her electrooculogram study showed slightly smaller Arden ratio in the lesion eye (reference data: 180%) (Table 1). Visual-evoked potentials revealed slightly prolonged implicit time of P wave in the lesion eye compared with the nonaffected eye (Table 1).

During follow-up examination, the white dots on the macula quickly moved to the midperipheral retina, and their intensity faded after 1 week; the vision improved to 6/20. The diagnosis of MEWDS was impressive. After 2 weeks, the BCVA became 6/15, and the fundus lesions were completely resolved. After 4 weeks, the BCVA improved to 6/12 and the patient’s symptoms had resolved. Serial ophthalmologic examinations were done during the follow-up period. Results of VF testing on recovery phase revealed improved and slightly enlarged blind spot with a 3.97-dB decrease in mean deviation in the left eye (Fig. 2B). A serial OCT follow-up showed that the initial disruption of the photoreceptor IS/OS junction line being gradually resolved. The disruption of the photoreceptor IS/OS line was replaced by a focal irregularity of the IS/OS line with a dome-shaped highly reflective material at the foveal area after 3 days (Fig. 3B). The IS/OS line was subsequently restored but remained indistinct at 1 month, the reflective material became smaller and less reflective (Fig. 3C), and the continuous IS/OS line was completely resolved at 3 months after presentation (Fig. 3D). During the follow-up period, the initially decreased ERG amplitude of maximal rod response and cone a-wave response gradually recovered in the affected eye to the same level as that of the nonaffected eye from 1 to 3 months (Table 1). At 3 months, the BCVA had recovered to 6/6, and the fundus examination showed completely normal retina without any visible lesions. One year after presentation, the retina remained normal without any sign of recurrence (Fig. 1D).

Fig. 1. (A) Fundus photography of the left eye showed foveal granularity and multiple gray-white spots at the macula and peripapillary area. (B,C) Results of fluorescein angiography showed early hyperfluorescence and late staining of these white dots as well as retinal vessels and optic disk. (D) One year after presentation, the retina remained normal, without any sign of recurrence.
3. Discussion

MEWDS was originally thought to be an inflammatory disease with foveal granularity and multiple white dots at the level of the deep retina or RPE layer.\(^1\) We have demonstrated that there was characteristic loss or irregularity of photoreceptor IS/OS junction line crossing the fovea in acute phase of MEWDS by OCT in the current case. This OCT feature was concomitant with the typical VF and ERG findings of MEWDS. The a-wave on ERG, which represents the photoreceptor function, was markedly reduced. Enlarged blind spot and central scotoma on VF was well explained by the fundus and FA appearance. Serial OCT findings in recovery phase show the temporary nature of the damage to the photoreceptor, which regains its normal contour within a matter of weeks. These evanescent OCT abnormalities were compatible with the clinical course of spontaneous resolution of white dots and visual function recovery of MEWDS in our case.

The diagnosis of MEWDS is based on the clinical history and ophthalmoscopic features. The differential diagnosis with other noninfectious white dot syndromes needs to include acute posterior multifocal placoid pigment epitheliopathy (APMPPE). APMPPE is a bilateral inflammatory disease with yellow, creamy, placoid lesions at the macula. There is no difference in occurrence between the sexes. FA discloses characteristic blockage of fluorescence in early phase with diffuse late staining.

The IS/OS line, the junction between the photoreceptor inner and outer segments, was initially demonstrated by stratus OCT.\(^5,6\) This line represents a sharply increased reflectivity of return beam on OCT imaging at the beginning of the photoreceptor outer segment. In human eyes, the length of the photoreceptor outer segment, or the length between the IS—OS

![Fig. 2](image1)

(A) Visual field (VF) (Humphrey 30-2) testing revealed enlarged blind spot and a central scotoma in central 20° field with a 13.28-dB decrease in mean deviation in the left eye. (B) Results of VF testing on recovery phase at 3 months revealed improved and slightly enlarged blind spot with a 3.97-dB decrease in mean deviation in the left eye.

![Fig. 3](image2)

(A) Results of optical coherence tomography showed a focal disruption of the photoreceptor inner/outer segment (IS/OS) junction line (long arrow) crossing the fovea in the left eye. (B) After 3 days, a focal irregularity of the IS/OS line was still noted, associated with a dome-shaped highly reflective material (short arrow) located at the outer nuclear layer and photoreceptor IS/OS junction. (C) At 1 month, the IS/OS line was subsequently restored but remained indistinct, and the reflective material had become smaller and less reflective. (D) The continuous IS/OS line was completely resolved at 3 months after presentation.

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<thead>
<tr>
<th>Table 1</th>
<th>The data of electrophysiologic study in the initial and recovery phases.</th>
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<tr>
<td></td>
<td>Initial</td>
</tr>
<tr>
<td>OD</td>
<td>OS</td>
</tr>
<tr>
<td>ERG rod maximal response (µV)</td>
<td>129.4</td>
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<tr>
<td>ERG cone A wave (µV)</td>
<td>165.04</td>
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<td></td>
<td>307.37</td>
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<tr>
<td>EEG Arden ratio (%)</td>
<td>182</td>
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<tr>
<td>VEP P wave (ms)</td>
<td>98.50</td>
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EOG = electrooculogram; ERG = electroretinography; OD = oculus dexter; OS = oculus sinister; VEP = visual-evoked potential.
line and the RPE, is 30 μm at the macula. Theoretically, the IS–OS line is detectable with stratus OCT if the photoreceptor outer segment is >10 μm. Thus, loss of the IS–OS line represents the loss of the photoreceptor outer segment or shortening <10 μm. Kanis and van Norren first reported that, using stratus OCT, there was loss of integrity of foveal cone cell in MEWDS that recovered after 21 weeks. Recently, ultrahigh-resolution OCT showed a disrupted IS/OS line in patients with MEWDS. Spaide et al also reported a case of MEWDS with acute widespread loss of visualization of the IS/OS boundary that recovered by 4 months. In this report, we have demonstrated the characteristic photoreceptor damage in acute MEWDS, which spontaneously and completely recovered 3 months after presentation.

The actual etiology of MEWDS is still unknown. In our patient, subtle disruption of the photoreceptor IO/OS junction line was the main pathological feature in acute MEWDS, which was evanescent and recovered to a continuous IO/OS line. These OCT findings may suggest that the disrupted outer segment but intact cell bodies of photoreceptor cells may contribute to the complete recovery of the outer segment and good visual outcome in MEWDS. The reduced a-wave amplitude of ERG in acute phase, which gradually recovered in association with resolved IO/OS line of OCT, further supports the main damage or inflammation of acute MEWDS being located at the photoreceptor cell outer segment but not the choriocapillaris or RPE. Moreover, in our case, OCT also showed a dome-shaped highly reflective material at the IO/OS junction crossing the foveal area, which was completely absorbed on recovery phase. Similar OCT findings in acute MEWDS have been reported by Amin. This material may be the aggregated waste debris of the damaged outer segment.

In conclusion, we have reported a case of MEWDS with complete spontaneous resolution of white dots, and evanescent microstructure abnormalities demonstrated by serial OCT. We have demonstrated that OCT provides a sensitive and non-invasive tool for the adjunct of diagnosis, understanding of pathogenesis, and for the serial anatomical evaluation of patients with MEWDS.

References