• Letter to the Editor •

Spectral-domain optical coherence tomography dynamic changes and steroid response in multiple evanescent white dot syndrome

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Dear Editor,

Multiple evanescent white dot syndrome (MEWDS) was first described in 1984 as a rare, acute, unilateral, multifocal retinochoroidal disorder, typically affecting young myopic women[1]. Previous studies with fluorescein angiography (FA) and electrophysiology suggested that MEWDS to be a disease in the retinal pigment epithelium (RPE) or outer retina[2], while recent studies with spectral-domain optical coherence tomography (SD-OCT) suggested it may be an outer retinal disease due to observation of hyperreflective material in outer retina and subtle disruptions of the ellipsoid zone without RPE disruption[3]. However, some studies with indocyanine green angiography (ICGA) and choroidal thickness measurement suggested that it may be a choroidal vascular or inflammation disease[4]. It has not been clearly demonstrated in the literatures that 1) what is the sequence of recovery for the hyperreflectant materials and the disrupted ellipsoid zone; 2) whether sub-lesion choroidal thickness changes sensitively during the process of the disease; 3) what is the role of short term steroid in accelerating the recovery. Here we report a MEWDS case with optical coherence tomography (OCT) findings of an initial resolution of the hyperreflectant materials preceding ellipsoid zone, with dynamic changes of sub-lesion choroidal thickness, and a fast recovery after short-term median dose steroid therapy.

A 27-year-old health man presented with blurry vision and photopsia in his left eye for two days. He has some flu-like symptoms 2wk earlier. His ocular history was significant for amplyopia in his right eye. He wore contact lenses measuring -11 DS OD and -9.5 DS OS. We observed the recovery process in this case with ophthalmoscopy, SD-OCT, FA, ICGA, and Humphrey perimetry examinations.

At the first visiting, best-corrected visual acuities (BCVA) were 20/200 OD and 20/60 OS. Slit-lamp examination was unremarkable. Fundus examination showed multiple gray-white, punctate dots located from posterior pole to the midperipheral region, and several Fuchs spots in nasal fundus and peripapillary atrophy OS (Figure 1B). There was an irregular optic nerve with peripapillary atrophy, some lacquercracks and Fuchs spots at the posterior pole OD (Figure 1A). SD-OCT showed some disruptions of the ellipsoid zone with hyperreflective material resting on the RPE and extending toward the inner retina through interdigitation zone, ellipsoid zone and outer nuclear layer which correlated to the white dot lesions observed by ophthalmoscopy. Humphrey 30-2 visual field test revealed a marked enlarged blind spot with central scotoma (Figure 2A). Because FA cannot be performed at his first visit, the patient was scheduled this test on his next visit.

Based on the clinical findings and ancillary testing results, the patient was diagnosed MEWDS. Due to the low vision in his right eye, the patient wanted a quick recovery for the left eye to resume normal work and life. The patient, therefore, was prescribed methylprednisolone 40 mg once daily for 7d.

At his 1-week follow-up visit, the patient reported that the left vision improved remarkably and photopsias had resolved

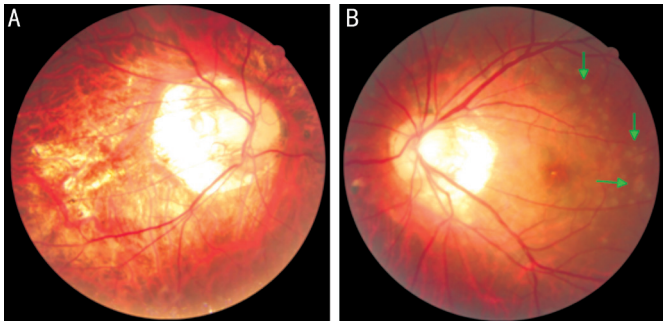

Figure 1 Color photography in baseline A: Irregular optic nerve with peripapillary atrophy, some lacquercracks and Fuchs spots at the posterior pole OD; B: Multiple gray-white, punctate dots located lesions (green arrows), several Fuchs spots in nasal fundus and peripapillary atrophy OS.

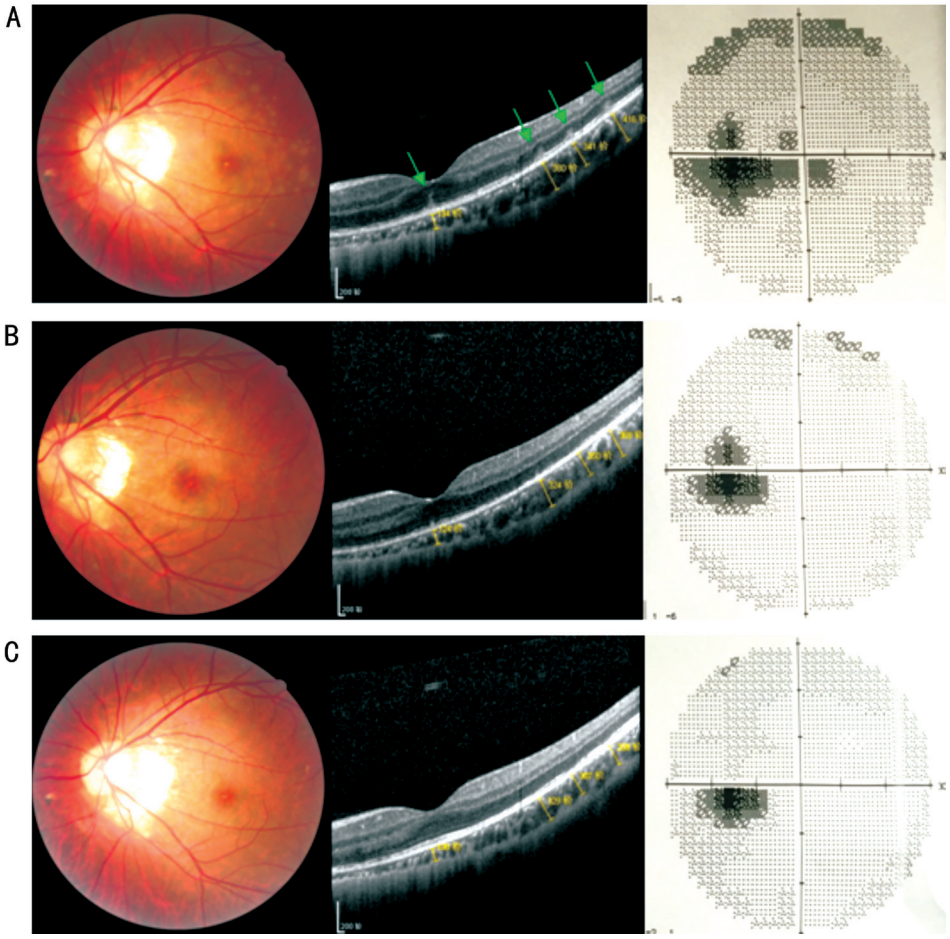

Figure 2 Changes in fundus, OCT and visual field A: Baseline: color photography showed multiple gray-white, punctate dots located lesions; OCT showed disruption of the ellipsoid zone and some hyperreflective lesions (green arrows); visual field test revealed a marked enlarged blind spot with central scotoma; B: One-week follow-up: color photography showed white dot lesions gradually resolved; OCT showed hyperreflective lesions disappeared but disruption of ellipsoid zone still exist; visual field defects gradually resolved; C: One-month follow-up: color photography, OCT and visual field are basically normal.

completely. The BCVA increased to 20/25 OS. Dilated fundus examination showed resolution of most white dot lesions. The visual field recovered remarkably (Figure 2B). FA demonstrated punctate hyperfluorescent in early stage and minimally staining in late stage (Figure 3A, 3B). ICGA showed some hypofluorescent spots in late phase, which were corresponded to the white dot lesions (Figure 3D). SD-OCT showed all the hyperreflective material disappeared but disruption of ellipsoid zone in the corresponding parts still exist (Figure 2B).

At his 1-month follow-up visit, the patient reported that his left vision acuity resolved completely, with BCVA of 20/20 OS. SD-OCT showed interdigitation zone, ellipsoid zone and outer nuclear layer completely recovered. Repeated visual field showed a mildly enlarged blind spot (Figure 2C).

Ten lesions on OCT were selected for choroidal thickness measurement on each visit. We performed a comparison of the choroidal thickness of the same ten lesions among three visit times (follow up scan mode) (Figure 2). Statistical analyses were performed with PASW 18.0 software. The mean choroidal thicknesses were 261.5 (±97.5) microns at the

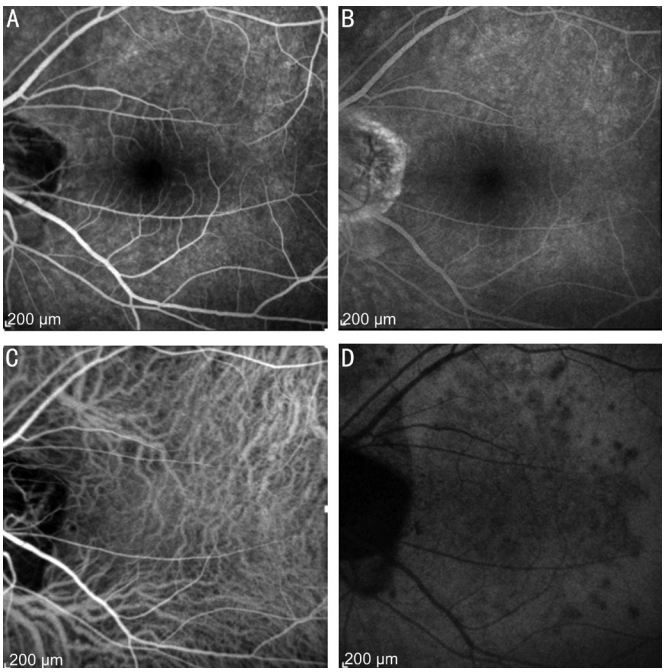

Figure 3 FA and ICGA A: Punctate hyperfluorescent in early phase of FA; B: Minimally staining in late phase of FA; C: No distinct abnormal finding in early phase of ICGA; D: Some hypofluorescent lesions at the posterior pole in late phase of ICGA.

baseline, 228.2 (±90.1) microns at the 1-week visit and 210.2 (±85.9) microns at the 4-week visit. The choroidal thickness of the ten lesions was thicker at onset than that at the 1-week visit (P=0.012) or that at the 4-week visit (P=0.001). However, the difference between 1 and 4wk visit was not statistically significant (P=0.052).

With the widespread clinical use of OCT, retinal changes of MEWDS in OCT have been reported in recent years. Diffuse disruptions of ellipsoid zone without RPE disruption were reported in previous studies[3,5]. In addition to ellipsoid zone disruptions, Marsiglia *et al*[3] found some protrusions of the hyperreflectant material from the ellipsoid layer toward the outer nuclear layer correspond to the location of dots seen with photography, ICGA, and FA in MEWDS. All the retinal changes in OCT in the present case at the onset were consistent with this literature reports. At the same time, we found resolution of hyperreflective material occurred earlier than recovery of ellipsoid zone, which has not been reported in previous literature. The resolution of hyperreflective materials were corresponded to the regression of white dot lesions and with the clinical improvement in visual acuity and visual field.

Regard to choroidal thickness change of MEWDS in OCT, it just has been reported in a few studies[3-5]. In those studies, choroidal thickness was thicker in the acute phase and decreased slightly in the convalescent phase. However, one report found the difference between acute phase and convalescent phase was not statistically significant[3]. Furthermore, in all these studies, only subfoveal choroidal thickness has been measured and compared[3-5]. In present case, ten lesions on OCT were selected for choroidal thickness measurement and the choroidal thickness of beneath ten lesions was thicker at onset than that in the convalescent phase (P=0.001). To the best of our knowledge, there are no reports discussing the choroidal thickness change in MEWDS with multiple lesions. We speculated that choroidal thickening beneath the lesion in acute phase may represent dilated choroicapillaries resulting from choroidal inflammation or ischemia, which corresponded to finding in ICGA. Measuring the choroidal thickness just beneath the lesions may be a more sensitive method to study choroicapillary changes in MEWDS.

Our patient was interesting in its faster recovery process. As MEWDS is a self-resolving condition, Marsiglia *et al*[3] observed 34 MEWDS patients without treatment and found the mean interval was 10wk for visual recovery and Lombardo[2] reported visual acuity restoration and clinical findings resolution in MEWDS without treatment are usually noted after 6 to 10wk. In an en-face OCT research in MEWDS, at 6-month follow-up, incomplete recovery of the ellipsoid zone were observed in all the 4 patients who have not been treated. Thus, the research suggested a possibility that a corticosteroid treatment could have allowed larger recovering in the photoreceptors integrity[6]. A case reported by Takahashi *et al*[7] seems to support this possibility; in that case, the visual acuity increased from 20/400 to 20/25 within 3d of the steroid pulse therapy (3000 mg for 3d). In the present patient, immediately after the median dose steroid therapy, the visual acuity of left eye increased from 20/60 to 20/25, the visual field recovered remarkably, white dot lesions disappeared completely and choroidal thickness decreased. This treatment result might suggest that steroid therapy may have a hope of promoting an early recovery from MEWDS. Although the etiology of MEWDS remains elusive, immunologic and infectious theories have been proposed[2]. According to a recent report, the presenting signs of MEWDS may link to activation of the microglia or dilation of the deep retinal capillary by the inflammation[3]. Due to the inflammatory process in MEWDS, steroid therapy may be an effective treatment to promote the regression of the lesion and shorten the course of MEWDS. Compared with pulse steroid therapy, medium-dose steroid was a relatively safe treatment with a low risk of side effect and complication. Thus, short term medium dose steroid treatment may be a choice to be applied in limited situation in which patients with rapid decline in vision acuity and need for a quick vision recovery to resume normal daily life and work like that of the present patient.

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