CASE REPORT
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Multiple Evanescent White Dot Syndrome: A Case Report and Experience with Corticosteroid Therapy

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ABSTRACT

Multiple evanescent white dot syndrome (MEWDS) is an inflammatory eye disease of the outer retina, retinal pigmented epithelium, choroid presenting with photopsia, loss of vision, and temporal scotoma. The patient was a 31-year-old female with a history of vision loss since 11 days ago (left eye). At presentation, best-corrected Snellen visual acuity was 20/140 in the Snellen chart. We decided to treat her with short time corticosteroid therapy (0.75 mg/kg/day prednisolone which was tapered in 3 weeks) for any possible rapid recovery of vision. The visual acuity of the involved eye was improved to 20/25 and 20/20, one week and three weeks after starting treatment respectively. Thus, it seems that short-term oral steroids might be an alternative method of management for patients with MEWDS.

Keywords: Inflammation; Prednisolone; White dot syndromes

INTRODUCTION

White dot syndromes comprise of diverse groups of disorders with inflammatory white dots or patches in the outer retina, retinal pigmented epithelium, or choroid. Multiple evanescent white dot syndrome (MEWDS) usually presents with unilateral numerous nummular grayish-white dots at fundus that typically affects healthy women age 20–50 years. Pathophysiology of this disease is still unclear but there are some cases reported after a viral infection[1] or vaccination.[2,3] High levels of IgG and IgM in patient's serum and flu-like symptoms have been considered as a kind of proof for the hypothesized infection-related etiology.[1,4] The most common clinical presentation is an acute painless visual disturbance. Other symptoms described by the patients are photopsia, dyschromatopsia, and para-central or temporal scotoma.[5] Spontaneous vision recovery usually occurs in 7-10 weeks with uncommon recurrences. [4] Fovea in MEWDS has a pathognomonic yellow to orange granular appearance, which was seen at the time of

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presentation or even after inflammation has taken over. Hyperfluorescence of the optic nerve in fluorescein angiography lesions could be observed.[6] Characteristic imaging findings are discontinuation and attenuation of inner segment-outer segment (ellipsoid) zone in optical coherence tomography (OCT), early wreath-like hyperfluorescence spots with late staining in fluorescein angiography and hypocyanescent spots in indocyanine green angiography.[5,7]

CASE PRESENTATION

A 31-year-old healthy Caucasian female was referred to our affiliated hospital (Farabi Eye Hospital, Tehran University of Medical Sciences, Tehran, Iran) with 11-day history of unilateral painless marked visual acuity loss in her left eye (OS). She also reported photopsia but scotoma was not presented. The past medical, drug and family histories were unremarkable. In the physical exam, the visual acuity (VA) was 20/20 in the right eye (OD) and 20/140 in OS (evaluated by Snellen chart). The anterior segment examination was normal for both eyes. A 2+ vitritis was detected in OS. In fundoscopy, several small white dots were seen in the deep retina. Optical Coherence Tomography showed disruption in the ellipsoid zone. Multiple hyper autofluorescence spots (100-200 µm) have been seen in fundus autofluorescence (FAF). Fluorescein angiography revealed both early and late hyperfluorescence dots with a wreath-like appearance which was more frequent in comparison to FAF lesions (suggestive MEWDS diagnosis) plus disc leakage. Indocyanine green angiography showed multiple hypofluorescence spots in the early and late phases (Figure 1). Based on all the findings, the patient was diagnosed with MEWDS.

Because of the intensive need for rapid vision recovery (due to the patient's insist and her issues), 0.75 mg/Kg/Day of oral prednisolone was started for the first week. She has been monitored weekly using fundus photography, OCT, and FAF. VA was improved to 20/25 the week after starting the treatment. This dosage was tapered off, with half of the dose every week, until it was discontented after 3 weeks of starting the treatment which at this point VA rose to 20/20. methods used for follow up. After 3 weeks, OCT showed restoring of the ellipsoid zone and fundus autofluorescence showed a diminishing of hyperautofluorescence spots (Figure 2). No recurrence was observed in the next 9 months of follow up.

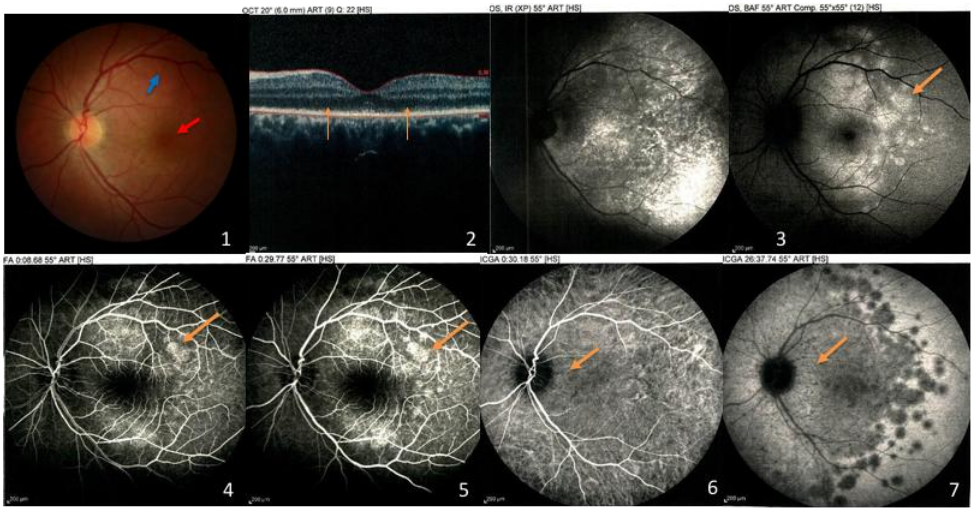

Figure 1. Color fundus photograph (1) representing multifocal deep, flat, and gray-white lesions in the retina (blue arrow) and yellow-orange granularity in the fovea (red arrow). Optical coherence tomography (OCT) shows abnormal ellipsoid zone marked by arrows (2). Infra-red and Fundus Autofluorescence images demonstrate hyperreflective and Hyperautofluorescent lesions which may not be visible in clinical examination (3, left and right, respectively). Fluorescein angiography shows early hyperfluorescence spots with the wreath-like pattern shown by arrow (4) and late staining of the lesions shown by arrow (5). Indocyanine green cholangiography (ICG) indicates multiple hypofluorescence areas in the early and late phases (arrow) especially around the optic disk (5 and 6).

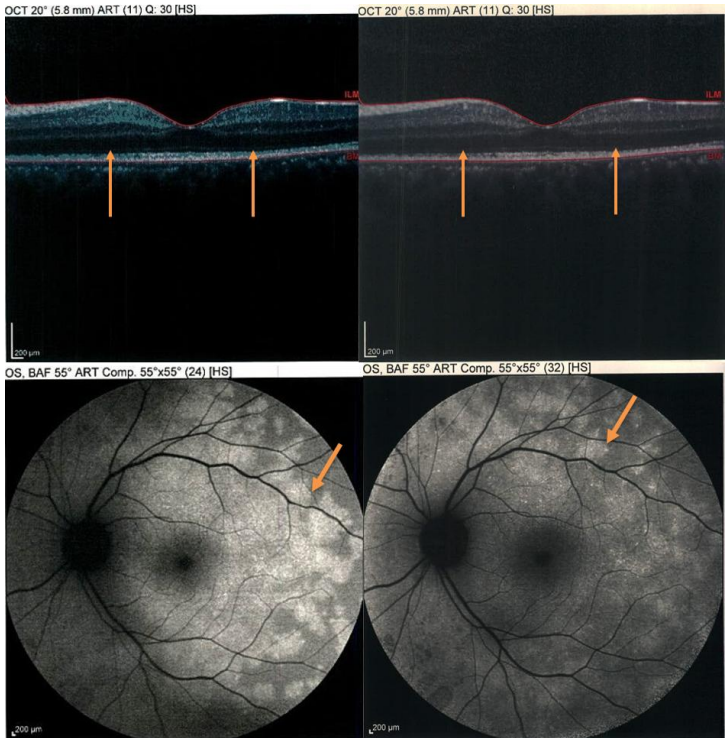

Figure 2. 1: Fundus autofluorescence and 2: Optical coherence tomography (OCT) of the patient in week 2; hyperautofluorescence is still seen in fundus autofluorescence image (arrow), the impaired ellipsoid band is seen in OCT (arrows). 3 and 4: Fundus autofluorescence and Optical coherence tomography (OCT) of the patient in week 3; resolution of hyperautofluorescence foci are seen in fundus autofluorescence image (arrow) and the ellipsoid band is resolved in OCT (arrows).

DISCUSSION

Considering the possible inflammatory basis of MEWDS and its self-limiting nature, observation is the most favorite method of management among most ophthalmologists. Although the lesion could be healed in a week to a month, in some cases, the disease can lead to a drastically decreased VA even to 20/400 requiring medical intervention to improve patient's quality of life through this episode.[8, 9] Takahashi et al., reported that pulse steroid treatment in a patient with MEWDS improved vision and funduscopic abnormalities very quickly. Their patient was a 25-year-old male with the unilateral MEWDS in the left eye associated with a 20/400 VA.[10] In another report, corticosteroid prescription in MEWDS after vaccination for human papillomavirus has been also noted.[11] Although spontaneous visual and anatomical recovery is a usual finding in the vast majority of patients, the development of chorioretinal scars, peripapillary atrophy, and choroidal neovascularization[12] have been reported in some cases. The amount of steroid prescribed for our patient was able to improve VA in a week and full recovery was attained in 3 weeks (approved with OCT changes).

The rapid and complete recovery of discontinued and interrupted ellipsoid zone in our patients could be encouraging. Although more controlled studies are needed, it seems that treatment with a tapering corticosteroid could be a choice for rapid recovery of vision in eligible MEWDS cases. The eligible cases are those who faster vision recovery is very important in their personal/carrier and are proper candidates for the treatment (considering steroid contraindication)

As it was mentioned, the spontaneous vision recovery is common in the patients diagnosed with MEWDS a period mostly 7-10 weeks. However, in some cases with serious personal/carrier problems, like our case, patients may ask their clinicians to consider the treatment to decrease the mentioned period. Herein, it was shown that oral steroid consumption was able to reduce the duration of the disease and to accelerate vision recovery.

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