Case Report

An Elderly Man with Atypical Multiple Evanescent White Dot Syndrome

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Keywords
Multiple evanescent white dot syndrome · Fluorescein angiography · Fundus autofluorescence · Indocyanine green angiography · Optical coherence tomography

Abstract
A rare occurrence of an atypical case of multiple evanescent white dot syndrome (MEWDS) in a 75-year-old man without viral prodrome or white dots on fundus that presented with acute, severe left eye visual loss, which returned to baseline without treatment in several weeks. Multimodal imaging, including fluorescein angiography (FA), fundus autofluorescence (FAF), indocyanine green angiography (ICG), and optical coherence tomography (OCT) demonstrated classical presentation of MEWDS with wreath-like lesions and inflammatory foci in the retinal pigment epithelium that correlated among modalities. Possible underlying systemic disorders were ruled out through extended work up. To the best of our knowledge, this is the first report to show atypical MEWDS in an elderly man with classic changes on FA, FAF, ICG, and OCT.

Introduction
Multiple evanescent white dot syndrome (MEWDS) is a rare, idiopathic posterior uveitis predominantly presenting in young women [1, 2]. The disease is associated with acute vision loss, which spontaneously recovers to baseline vision without treatment. MEWDS is generally preceded by a viral prodrome and characterized by scattered white dots in the posterior pole.
of the fundus. Multimodal imaging such as fluorescein angiography (FA), autofluorescence imaging (FAF), indocyanine green angiography (ICG), and optical coherence tomography (OCT) demonstrated characteristic diagnostic findings, such as a “wreath-like” pattern of hyperfluorescence or hypofluorescent lesions [3]. Here we present an elderly man who developed severe, transient vision loss of the left eye without the typical white spots on fundus but demonstrating acute outer retinal lesions resembling MEWDS on multimodal imaging.

**Case Report**

A 75-year-old pseudophakic man with dyslipidemia and hypertension presented with subacute left eye vision loss for 2 weeks. He received his first dose of the COVID-19 vaccine 1 week before onset of symptoms. He did not have any signs of viral prodrome. He complained of seeing black spots in his temporal visual field with no headaches, dizziness, or other systemic complaints. The right eye visual acuity was 20/25 with an overall unremarkable exam. His left eye had a best corrected visual acuity of 20/200 (decreased from 20/32), Ishihara color test of 2/8, and a positive afferent pupillary defect. Slit-lamp exam showed trace cells in anterior chamber (AC), mild haze in the vitreous with an otherwise unremarkable fundus exam (Fig. 1a). Visual field testing demonstrated an enlarged blind spot and nonspecific temporal defects (mean defect = −6.55 dB).

His multimodal imaging on the same visit showed that the right eye was unremarkable except for drusen with pigmentation at the macular area and a small retinal pigment epithelial (RPE) detachment on OCT. There were a number of findings from his left eye. On FA, multiple

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**Fig. 1.** Multimodal imaging of an elderly man with atypical MEWDS in the left eye on initial visit. **a** Fundus imaging showing mild vitreous haze and no white lesions. **b** FA showing early wreath-like pattern of hyperfluorescent lesions. **c** FAF showing hyperfluorescent lesions. **d** ICG showing hypofluorescent lesions in the posterior pole. **e** OCT showing hyperreflective foci in the outer retinal layer. RPE layer had early cessation and hyperreflective dome-shaped lesions that extend into or through the ellipsoid zone (yellow arrow). Small hyperreflective foci can also be seen in the vitreous, lower macula, and inner retina. OCT raised suspicion of a scleral protrusion into the eye (red arrow), which was ruled out by ultrasound.
hyperfluorescent patches in a wreath-like pattern can be seen in the posterior pole and periphery with no leakage (Fig. 1b) with matching hyperfluorescent patches on FAF (Fig. 1c). Hypofluorescent dotted lesions of different sizes were seen on the entire length of the posterior pole on ICG, which correlate with the FA and FAF lesions (Fig. 1d). OCT studies demonstrated hyperreflective foci in the outer retinal layers and atrophy of the photoreceptor layer without the presence of fluid. The RPE layer had early cessation and hyperreflective dome-shaped lesions that extend into or through the ellipsoid zone. Small hyperreflective foci can also be seen in the vitreous, lower macula, and inner retina. OCT findings raised suspicion for a scleral protrusion into the eye that was not present in ultrasound imaging (Fig. 1e).

Additional work up was completed to rule out systemic disorders such as malignant, infectious, or autoimmune disorders. His CT orbits with and without contrast were within normal ranges. His magnetic resonance imaging orbit with and without contrast demonstrated a slight asymmetry between the optic nerves without any compressive lesions. Intraocular lymphoma was considered but ruled out because of negative malignant cells from his AC tap. His AC tap revealed an IL-10 of 0 pg/mL and IL-6 of 170 pg/mL, making lymphoma unlikely; an IL-10/IL-6 ratio of greater than one or an absolute IL-10 level of ≥50 pg/mL indicates lymphoma [4, 5]. Therefore, his AC tap was more in line with uveitis. His extended lab work was all within normal limits including complete blood count, comprehensive metabolic panels, syphilis-related tests (venereal disease research laboratory, Treponema pallidum hemagglutination), sarcoidosis (lactate dehydrogenase and C-reactive protein), and rheumatic markers (antinuclear antibodies, antineutrophil cytoplasmic autoantibody-associated diseases, C3, and C4). Low-dose prednisone (0.5 mg/kg) was prescribed in consideration of possible inflammatory causes after ruling out infectious disorders. The patient decided to not start prednisone.

At the 2-week follow-up, he presented with spontaneously improved vision in his left eye. The visual acuity had improved dramatically from 20/200 to 20/40. Compared to his previous tests, the hypoautofluorescent lesions have reduced on FAF, and the hyperreflective foci were flatter on OCT (Fig. 2). Due to the spontaneous improvement and strong indication of MEWDS, no further treatment was provided. His vision continued to improve spontaneously. At the 3-month follow-up, he reported not seeing any black dots in his temporal visual field of his left eye. The best corrected visual acuity of his left eye was 20/25 (improved from 20/40). On visual field testing, the temporal defects have regressed (mean defect = −3.86 dB) in comparison to the initial visit (mean defect = −6.55 dB). On imaging, the hypoautofluorescent lesions have almost completely regressed on FAF, and the hyperreflective foci had flattened and mostly disappeared on OCT (Fig. 2).

Discussion

MEWDS is a rare posterior uveitis with an annual incidence of 0.22 per 100,000, mostly seen in young woman and presents with characteristic white spots in the fundus with other commonly documented findings in multimodal imaging [2]. The disease can present with a viral prodrome followed by acute visual loss and recovery to baseline visual acuity without treatment [1–3]. One case of MEWDS was reported in 1999 in a 60-year-old patient presenting with transient, acute visual acuity decline, peripheral visual field loss, and the classic white dots on the fundus that regressed without treatment [6].

The elderly male patient in the present report presented with MEWDS without a viral prodrome or white spots in the fundus but with an otherwise typical clinical presentation (acute transient vision loss, temporal visual field loss, blind spot enlargement, and vitreous haze) and classic features of MEWDS on imaging including wreath-like pattern of early hyperfluorescence in FA, hyperautofluorescent lesions on FAF, hypofluorescent lesions on ICG, and
ellipsoid zone disruption and hyperreflective lesions in the RPE on OCT that correlate to each other in pattern and location among the modalities [1–3]. Atypical presentations of MEWDS have also been documented. A case series of young patients demonstrated classical clinical presentations of MEWDS but without the classical white dots on the fundus and no lesions on select imaging like ICG and FAF [7, 8]. For our patient, the lack of white dots on the fundus, classical presentation on all imaging modalities, transient vision loss with spontaneous recovery, and negative systemic work up highly indicate that our patient had MEWDS with atypical presentation.

The cause of MEWDS is unknown. One possible etiology indicates that some people are more genetically predisposed to an immune-mediated response to a viral-like infection, which can potentially lead to inflammation in the peripapillary circulation. While our patient did not have a viral infection, he did receive a COVID-19 vaccine 1 week prior to presentation of his symptoms. It is known that COVID-19 vaccine could trigger ocular inflammation [9]. Direct retinal inflammation may be the cause of his vision loss. Moreover, inflammation from the peripapillary circulation can spread through the ciliary arteries to the retinal circulation causing inflammatory material to spread throughout the retinal layers and accumulate in the RPE [1]. Alternatively, the disease mechanism can involve inflammatory nonperfusion of the choriocapillaris with vaso-occlusive issues at the inner choroid, leading to ischemia of the outer retina and photoreceptor outer segments [3]. Once the acute inflammatory phase weans, patients recover their original vision since all retinal changes including those in the RPE and ellipsoid zone fully recovers with no signs of photoreceptor cell damage, which was demonstrated in our patient [2]. While no treatment is required for MEWDS, it is still essential to rule out potential underlying sources of inflammation and other diagnoses with similar features like lymphoma, syphilis, and sarcoidosis that require urgent treatment.

In conclusion, MEWDS is a rare disease of young women, but atypical cases could be seen in older male patients. To our knowledge, this is the first case to document atypical MEWDS in an elderly man using a full range of ophthalmic imaging modalities including FA, FAF, ICG, and OCT.
Statement of Ethics

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. Ethical approval is not required for this study in accordance with local or national guidelines.

Conflict of Interest Statement

The authors have no disclosures.

Funding Sources

No funding received.

Author Contributions

Liang Wang, Polina Lankry, David Rabinovitch, Ryan Gallo, Rita Laiginhas, Prashanth Iyer, Shiri Shulman, and Omer Trivizki contributed to the design and implementation of this report, to the analysis of the results, and to the writing of the manuscript. Liang Wang, Polina Lankry, David Rabinovitch, Ryan Gallo, Rita Laiginhas, Prashanth Iyer, Shiri Shulman, and Omer Trivizki attest that they meet the current ICMJE criteria for authorship.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

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