Multiple evanescent white dot syndrome: Bilateral disease may be silent and asymmetric

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ABSTRACT

Purpose: We report a patient with unilateral symptoms presenting with bilateral multiple evanescent white dot syndrome (MEWDS) in order to highlight the utility of multimodal imaging in revealing asymptomatic lesions in the fellow eye and underscore the importance of looking for silent bilateral disease.

Observations: A 39-year-old man presented with blurry vision and photopsias in the left eye (OS). Funduscopic examination revealed characteristic granular white dots in the posterior pole OS. Multimodal imaging included fundus autofluorescence, which revealed numerous hyperautofluorescent lesions in both eyes, more than appreciated on clinical examination alone and corresponding ellipsoid disruption on OCT. Seven bilateral cases have been previously reported, all of which are asymmetric, similar to the case reported and are summarized here.

Conclusions and Importance: While MEWDS is most often thought of as a unilateral disease, it may rarely present bilaterally as in the case presented here. Multimodal imaging is especially useful in diagnosis and follow-up. Fundus autofluorescence may be the most sensitive and practical test for detecting MEWDS, revealing lesions in the absence of white dots on clinical exam.

1. Introduction

Multiple evanescent white dot syndrome (MEWDS) is a rare, inflammatory disease most commonly affecting young people between the ages of 15 and 50 years with a 4:1 female to male predilection.1 Patients often report blurry vision, temporal or paracentral scotoma, dyschromatopsia, and photopsia on initial presentation. Funduscopyc examination reveals a quiet anterior chamber, mild vitritis, and multiple granular appearing grayish white lesions measuring 100–200 μm throughout the posterior pole that may leave mild foveal granularity upon resolution.2 Optic disc edema may be present and visual field testing often reveals an enlarged blind spot, temporal, or paracentral scotoma. The clinical course for MEWDS is short and self-limited, with most patients experiencing resolution of symptoms and excellent visual recovery within a few months.

MEWDS is typically a unilateral entity, with bilaterality and recurrences being unusual for this disease. We report a rare bilateral presentation of MEWDS and highlight the utility of multimodal imaging in diagnosing the disease and revealing lesions that may not be seen on clinical examination alone. Additionally, this report summarizes previously rarely reported cases of bilateral MEWDS in the literature.

2. Case report

A healthy 39-year-old man reported blurry vision and photopsias in the left eye (OS) ongoing for one week. He denied any eye redness, pain, or photophobia. His best corrected visual acuity was 20/20 in both eyes (OU). Pupils, intraocular pressure, and confrontational visual fields were normal in both eyes. A slit lamp examination showed a normal anterior segment with a quiet anterior chamber OU. A dilated fundus examination revealed 1+ vitreous cell in the right eye (OD), trace vitreous cell OS, and small yellowish-white spots throughout the posterior pole OU, more prominent OS than OD (Fig. 1A and B). On careful review of systems, the patient reported a recent flu-like illness, but no other systemic symptoms nor exposures. Fluorescein angiography revealed early and late hyperfluorescence of the lesions (Fig. 1C and D) and fundus autofluorescence highlighted the multiple hyperautofluorescent lesions through the posterior pole, more numerous than appreciated on examination alone (Fig. 2A and B). Optical coherence tomography through a lesion OS showed disruption of the ellipsoid zone in this area.
Fig. 1. Multiple granular white spots are seen in the posterior pole OU (A and B). Fluorescein angiography shows late staining of these lesions (C and D).

Fig. 2. Fundus autofluorescence reveals multiple hyperautofluorescent lesions throughout the posterior pole OU more numerous than appreciated on fundus photography or clinical exam (A and B). OCT though one lesion shows disruption of the ellipsoid zone as delineated by the yellow arrows (C). The hyper-autofluorescent lesions resolve on follow-up (D and E) and there is restoration of the ellipsoid zone on OCT (F). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)
ophthalmic findings. prodrome without any other systemic manifestations may precede the based on clinical exam alone, no laboratory testing is required. A viral (MEWDS) with bilateral presentation. As the diagnosis of MEWDS is resolution of FAF and OCT changes (Fig. 2D and E,F).

3. Discussion

Given the clinical history, examination, and imaging findings, the patient was diagnosed with multiple evanescent white dot syndrome (MEWDS) with bilateral presentation. As the diagnosis of MEWDS is based on clinical exam alone, no laboratory testing is required. A viral prodrome without any other systemic manifestations may precede the evaluation. However, multimodal imaging is helpful in diagnosing the disease and differentiating it from other inflammatory and infectious etiologies. MEWDS predominantly affects the outer retina and ellipsoid zone. Spectral-domain optical coherence tomography (SD-OCT) correspondingly demonstrates disruption of the ellipsoid zone in the acute phase of the disease, though the outer nuclear layer and interdigitation zone may also be involved. Fluorescein angiography reveals “wreath-like” early hyperfluorescent spots with late staining and indocyanine green angiography (ICGA) demonstrates early and late hypocyanescence. As demonstrated by this case, fundus autofluorescence (FAF) imaging can nicely highlight the lesions, which appear hyperautofluorescent, and is a useful imaging modality to follow resolution. Reports suggest FAF may be the most sensitive and practical test for detecting MEWDS, revealing lesions in the absence of white dots on clinical exam.

With clinical resolution of the disease, the imaging findings typically resolve with restoration of the photoreceptor outer segments, though there may be persistent granular pigmentary changes on fundoscopy and continued hypocyanescence on ICGA in areas of the previously seen lesions. Corticosteroids and antibiotic treatments are not indicated as MEWDS resolves spontaneously and affected patients are observed for resolution of symptoms and fundoscopic findings. While rare, 10% of patients may experience recurrence and require treatment with immunosuppression.

While this entity is typically unilateral, bilateral cases such as that in our patient have been infrequently reported. It is notable that in all of these previously reported bilateral cases the disease was asymmetric and more subtle or asymptomatic in the less affected eye (these cases are summarized in Table 1). Six of these previously reported cases occurred in women and 2 in men. All of these bilateral cases were self-limited. Two of the previously reported cases of bilateral disease were separated in presentation by several years. Furthermore, in patients with only unilateral symptoms and clinical disease, there is evidence to suggest that photoreceptor dysfunction and visual field deficits may be found in both eyes despite lack of symptoms. Similar to prior reports of bilateral MEWDS, our patient was only symptomatic in his left eye, but clinical exam and imaging revealed the presence of silent bilateral disease. Given the current case and prior reports, it is plausible bilateral disease is more common on presentation than is recognized, and multimodal imaging may help reveal potentially silent and asymmetric bilateral disease.

4. Conclusion

While MEWDS is most often thought of as a unilateral disease, it may rarely present bilaterally as in the case presented here. Fundus autofluorescence is a non-invasive and practical imaging modality that is particularly useful in highlighting lesions that may have otherwise been missed on fundoscopic examination alone. This report highlights the importance of a thorough bilateral exam in patients whom MEWDS is suspected and the clinical utility of multimodal fundus imaging in the diagnosis and monitoring of the disease. We suggest fundus autofluorescence is used in all cases of suspected MEWDS and believe this disease entity may more often be bilateral than previously recognized using clinical examination alone.

Informed consent

The patient consented to publication of the case both orally and in writing. Additionally, this report does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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Declaration of competing interest

The authors report no conflicts of interest.

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