Persistent photopsia: multiple evanescent white dot syndrome in a sexagenarian

Christine Bourke, Kirk A J Stephenson, Yvonne Delaney, James Morgan

Ophthalmology Department, Mater Misericordiae University Hospital, Dublin, Ireland

Correspondence to
Dr Kirk A J Stephenson;
kirkstephenson@hotmail.com

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DESCRIPTION

A 67-year-old emmetropic woman presented with a 1-month history of coin-shaped flashes in her right temporal visual field with best corrected visual acuity of 6/6 in each eye. She had no associated floaters, headache, viral prodrome or significant medical history.

Examination showed negative relative afferent pupillary defect, full Ishihara pseudoisochromatic colour plates, quiet anterior chambers, intraocular pressures of 15 mm Hg, no vitritis and no vitreoretinal traction, retinal tears or detachment. There were scattered yellow-orange outer retinal deposits predominantly at the right posterior pole with a distorted foveal reflex (figure 1A,B). Autofluorescence imaging (Optos, UK) showed hyperautofluorescence in a wreath-like configuration extending beyond the posterior pole (figure 1C). Fundus fluorescein angiography (Optos plc, UK) demonstrated staining in the same distribution without leakage or vasculitis (figure 1D) and indocyanine green angiography (ICGA) showed no choroidal lesions. Visual fields (Octopus, Haag-Streit, Switzerland) only showed enlargement of the right eye blind spot. Macular optical coherence tomography (OCT, Cirrus 5000, Carl Zeiss Meditec, USA) showed normal retinal lamination on B-scan; however, *en face* analysis revealed hyper-reflective dots at the photoreceptor inner segment/outer segment junction (figure 2). *Borrelia*, tuberculosis, *Toxoplasma* and *Treponema* serology were negative and a working diagnosis of multiple evanescent white dot syndrome (MEWDS) was made. As vision was excellent, no treatment was recommended and the patient was observed closely. Symptoms had resolved at the 1-month visit including return of the blind spot to normal. Interestingly in this case, non-invasive imaging modalities (ie, autofluorescence and *en face* infrared reflectance imaging/OCT) were more helpful than traditional angiography in diagnosis and monitoring.

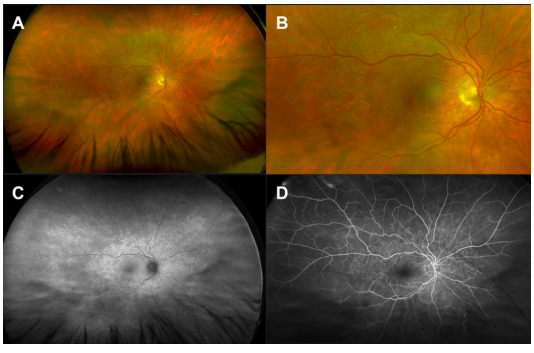

Figure 1 (A) Widefield colour photograph of the right fundus of this patient showing multiple white-yellow subretinal dots. (B) Higher magnification colour photograph showing white-yellow dots concentrated in the macula. (C) Widefield fundus autofluorescence demonstrating wreath-like hyperautofluorescence surrounding the macula and vascular arcades. (D) Fundus fluorescein angiography of the right eye showing hyperfluorescent staining of the white dots matching the distribution seen in the autofluorescence imaging. All images were taken with the Optos 'California' camera, Optos plc, UK.

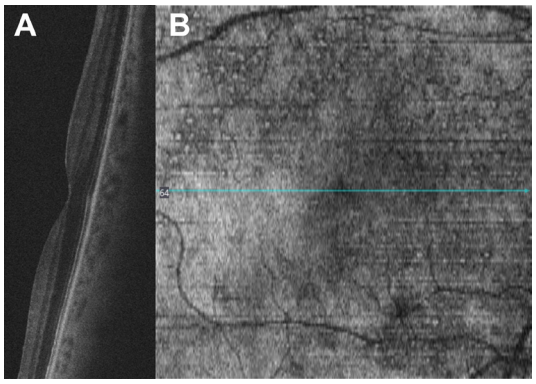

Figure 2 (A) Horizontal OCT B-scan through the fovea showing normal retinal lamination and no oedema. (B) *En face* infrared reflectance OCT image delineating hyper-reflective foci corresponding to outer retinal white dots in figure 1A,B. OCT, optical coherence tomography.

MEWDS is a rare (0.22 per 100 000), unilateral, self-limiting disease characterised by multiple discrete small white dots at the level of the outer retina, typically concentrated at the macula.[1 2] Lesions typically resolve over 2–3 months.[1] There is a young female predilection (mean 28 years, range 14–47 years) and a viral prodrome is reported in approximately 50%.[3 4] Presenting symptoms include sudden decrease in vision, paracentral scotomata and/or temporal photopsia.[1] *En face* infrared reflectance shows hyper-reflective outer retinal lesions corresponding to the white dots.[5 6] ICGA may reveal peripapillary hypofluorescence and more numerous hypofluorescent lesions than expected clinically.[7] Full-field electroretinogram (ERG) shows reversible photopic/scotopic a-wave attenuation, while multifocal ERG may delineate areas of normal function.[8] There are no systemic manifestations.[2]

The aetiology of MEWDS is debated, possibly a postviral inflammatory insult to the retinal pigment epithelium/photoreceptors.[9] The

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diagnosis is clinical, with laboratory, electrophysiological and angiographic investigations excluding other aetiologies.[2] The differential diagnosis of photopsias includes posterior vitreous detachment, optic neuritis, acute macular neuroretinopathy (AMN), acute idiopathic blind spot enlargement (AIEBS), acute zonal occult outer retinopathy and posterior uveitis (eg, sarcoid), among others. AMN (perifoveal outer nuclear layer hyper-reflectivity) and AIEBS (no discernible lesion) are rare conditions, also primarily affecting young women, resulting in central/paracentral scotomata and are considered within the MEWDS spectrum.[3 10] MEWDS is classically self-limiting (<3 months) with an excellent visual prognosis without treatment, though recurrence may occur in 10%.

Learning points

- Multiple evanescent white dot syndrome (MEWDS) is a rare unilateral self-limiting outer retinal inflammatory disease with a young (mean 28 years) female preponderance. It is typically self-limiting with excellent visual prognosis, though can recur in 10%.
- Inflammatory outer retinopathies (including MEWDS) should be included in the differential diagnosis of photopsia in the absence of vitreoretinal traction or rhegmatogenous retinal detachment.
- Diagnosis of MEWDS is clinical; however, multimodal imaging (eg, optical coherence tomography, autofluorescence, fundus fluorescein angiography, indocyanine green angiography) can be a useful diagnostic adjunct.

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ORCID iD

Kirk A J Stephenson http://orcid.org/0000-0002-7462-7725

REFERENCES

1 Abu-Yaghi NE, Hartono SP, Hodge DO, *et al*. White dot syndromes: a 20-year study of incidence, clinical features, and outcomes. *Ocul Immunol Inflamm* 2011;19:426–30.
2 Jampol LM, Sieving PA, Pugh D, *et al*. Multiple evanescent white dot syndrome. I. clinical findings. *Arch Ophthalmol* 1984;102:671–4.
3 Marsiglia M, Gallego-Pinazo R, Cunha de Souza E, *et al*. Expanded clinical spectrum of multiple evanescent white dot syndrome with multimodal imaging. *Retina* 2016;36:64–74.
4 Quillen D, Davis J, *et al*. Perspective: the white dot syndromes. *Am J Ophthalmol* 2004. doi:10.1016/j.ajo.2004.01.053
5 Nguyen MHT, Witkin AJ, Reichel E, Reichel E, *et al*. Microstructural abnormalities in MEWDS demonstrated by ultrahigh resolution optical coherence tomography. *Retina* 2007;27:414–8.
6 Pichi F, Srvivastava SK, Chexal S, *et al*. En face optical coherence tomography and optical coherence tomography angiography of multiple evanescent white dot syndrome: new insights into pathogenesis. *Retina* 2016;36 Suppl 1:S178–88.
7 Gross NE, Yannuzzi LA, Freund KB, *et al*. Multiple evanescent white dot syndrome. *Arch Ophthalmol* 2006;124:493–500.
8 Cheng JYC, Luu CD, Yeo IYS, *et al*. The outer and inner retinal function in patients with multiple evanescent white dot syndrome. *Clin Exp Ophthalmol* 2009;37:478–84.
9 dell'Omo R, Pavesio CE. Multiple evanescent white dot syndrome (MEWDS). *Int Ophthalmol Clin* 2012;52:221–8.
10 Bryan RG, Freund KB, Yannuzzi LA, *et al*. Multiple evanescent white dot syndrome in patients with multifocal choroiditis. *Retina* 2002;22:317–22.