Persistent photopsia: multiple evanescent white dot syndrome in a sexagenarian

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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. 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.5 There are no systemic manifestations.2

The aetiology of MEWDS is debated, possibly a postviral inflammatory insult to the retinal pigment epithelium/photoreceptors.3 The
diagnosis is clinical, with laboratory, electrophysiological and angiographic investigations excluding other aetiologies. 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 retinal 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. MEWDS is considered within the MEWDS spectrum. 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 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 it 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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