Published in final edited form as:

JAMA Ophthalmol. 2014 August ; 132(8): 1015–1016. doi:10.1001/jamaophthalmol.2014.1444.

Shimmering Lights

Mehnaz Khan¹, P. Kumar Rao², and Rajesh C. Rao^{1,2,3}

¹Department of Ophthalmology and Visual Sciences, W.K. Kellogg Eye Center, University of Michigan Medical School, Ann Arbor

²Department of Ophthalmology and Visual Sciences, Washington University School of Medicine, St. Louis, MO

³Department of Pathology, University of Michigan Medical School, Ann Arbor

A healthy 37 year old male complains of “shimmering” lights” and blurry vision in his right eye. He denies any other ocular discomfort. His best-corrected visual acuity is 20/30 OD and 20/20 OS. His pupils, intraocular pressure, motility and confrontational visual fields are normal OU. External examination and slit-lamp examination of the anterior segment of both eyes are unremarkable. Dilated fundus exam shows clear vitreous OU, and small yellow-white retinal lesions OD. The patient denies history of recent illnesses.

[Image: Fundus photo, FA, 4 panels]

What would you do next?

- A. Observe
- B. Treat with oral corticosteroids
- C. Treat with topical corticosteroids
- D. Treat with intravitreal antibiotics

[word count of case presentation, question, 4 answers: 108]

Diagnosis

Multiple Evanescent White Dot Syndrome (MEWDS)

What to do next

- A. Observe

The deep yellow-white unilateral fundus lesions with corresponding “wreath-like” hyperfluorescence in the fluorescein angiogram (FA) are characteristic of MEWDS. Disease course is generally benign, with spontaneous improvement of vision and resolution of fundus lesions within several weeks. The disease is largely unilateral and monophasic (2).

Corresponding Author: Rajesh C. Rao, MD, W.K. Kellogg Eye Center, University of Michigan, 1000 Wall St, Brehm 8326, Ann Arbor, MI 48105, rajeshr@med.umich.edu.

The authors have no conflicts of interests to declare.

Image: FAF, OCT (6 panels)

Comment

MEWDS is a rare outer retinal and choroidal inflammatory condition first described by Jampol et al. in 1984 (2). Clinical symptoms include decreased visual acuity, scotomata, enlarged blind-spot, and photopsia. Deep yellow-white fundus lesions, foveal granularity, and optic nerve inflammation can be present. MEWDS often occurs following viral upper respiratory infection and may be related to an immune-mediated mechanism in a genetically susceptible individual (3). MEWDS has a predilection for women in the 2nd-5th decades (2).

Standard imaging modalities used for diagnosis of MEWDS include fundus biomicroscopy and FA. On fundoscopy, MEWDS lesions are scattered white-dots varying in size from 50-1000 microns, located in the outer retina/retinal pigment epithelium (RPE) and inner choroid (4). On FA, “wreath-like” clusters of hyperfluorescence and late staining in the acute phase of the disease are seen (4, 5). A case series has described the evolution of the dots on FA – most dots appear during the choroidal-filling of FA (believed to localize to RPE) while some dots fluoresce during the arterial phase (originating within retinal circulation) (6). Indocyanine green angiography (ICGA), unremarkable during the early and mid-phases of the study, shows late-phase hypofluorescent lesions in the same areas of hyperfluorescence on FA. Gross et al. showed more lesions on ICGA than on clinical examination/FA, highlighting a choroidal inflammatory component of MEWDS.

Imaging modalities in the diagnosis and evolution of MEWDS have been expanded to include fundus autofluorescence (FAF) and spectral-domain optical coherence tomography (SD-OCT). Early MEWDS is characterized by multifocal choroidal inflammation, resulting in phagocytosis of photoreceptor outer segments and increased lipofuscin production; this manifests as increased FAF. As the inflammatory response alleviates, the photoreceptor outer segments are restored, leading to decreased production of lipofuscin and normalization of FAF signal (7). In our patient, Fig 2 a-c showed increased FAF at the onset of symptoms with near resolution noted at 6 weeks follow-up without any treatment. SD-OCT also confirmed ellipsoid (inner segment/outer segment [IS/OS] photoreceptor) layer disturbance in the acute phase of the disease with structural restoration of the ellipsoid (IS/OS) layer during recovery. Reports of atypical cases of MEWDS without presence of white dots in the posterior pole and no findings on FA, ICG or FAF exist (8). However, even in these atypical cases, foveal granularity and disruption of ellipsoid (IS/OS) layer were noted. Therefore foveal granularity, in which outer retinal and RPE changes can be discerned with FAF, with accompanying ellipsoid (IS/OS) layer loss noted on OCT, appears to be fairly sensitive for MEWDS and can be used to diagnose the disease in the absence of other typical findings. Also there may be persistent hypofluorescent abnormalities in the ICGA may persistent after clinical signs resolve (9). Overall, given the widely varying presentation of MEWDS, the diagnosis can be reached based on history, clinical examination, course and multimodal retinal imaging.

MEWDS tends to resolve spontaneously, and affected patients can typically be observed. Corticosteroid use has been reported to resolve symptoms and fundoscopic findings

following three days of treatment (1). However, systemic complications of oral corticosteroid therapy, depending on dose, may outweigh the benefits of therapy. Antibiotic treatment is not indicated for MEWDS. During followup over 6 weeks, our patient did not experience a recurrent episode, visual acuity returned to 20/20, however subtle foveal granularity persisted.

[Word count for diagnosis, what to do next, comment: 600 words]

Acknowledgments

R.C.R. is supported by NIH/NEI K12EY022299. This sponsor had no role in the design and conduct of the study; collection, management, analysis, and interpretation of the data; and preparation, review, or approval of the manuscript.

References

1. Takahashi Y, Ataka S, Wada S, Kohno T, Nomura Y, Shiraki K. A case of multiple evanescent white dot syndrome treated by steroid pulse therapy. *Osaka City Med J*. 2006 Dec; 52(2):83–6. [PubMed: 17330396]
2. Jampol LM, Sieving PA, Pugh D, Fishman GA, Gilbert H. Multiple evanescent white dot syndrome. I. Clinical findings. *Arch Ophthalmol*. 1984 May; 102(5):671–4. [PubMed: 6721749]
3. Jampol LM, Becker KG. White dot syndromes of the retina: a hypothesis based on the common genetic hypothesis of autoimmune/inflammatory disease. *Am J Ophthalmol*. 2003 Mar; 135(3):376–9. [PubMed: 12614757]
4. Mamalis N, Daily MJ. Multiple evanescent white dot syndrome. A report of eight cases. *Ophthalmology*. 1987 Oct; 94(10):1209–12. [PubMed: 3684198]
5. Slusher MM, Weaver RG. Multiple evanescent white dot syndrome. *Retina*. 1988; 8:132–5. [PubMed: 3420313]
6. Gross NE, Yannuzzi LA, Freund KB, Spaide RF, Amato GP, Sigal R. Multiple evanescent white dot syndrome. *Arch Ophthalmol*. 2006 Apr; 124(4):493–500. [PubMed: 16606874]
7. Hua R, Chen K, Liu LM, Liu NN, Chen L, Teng WP. Multi-modality imaging on multiple evanescent white dot syndrome-A Spectralis Study. *Int J Ophthalmol*. 2012; 5(5):644–7. [PubMed: 23166880]
8. Shelsta HN, Rao RR, Bhatt HK, Jampol LM. Atypical presentations of multiple evanescent white dot syndrome without white dots: a case series. *Retina*. 2011 May; 31(5):973–6. [PubMed: 21358458]
9. Dell'omo R, Wong R, Marino M, Konstantopoulou K, Pavesio C. Relationship between different fluorescein and indocyanine green angiography features in multiple evanescent white dot syndrome. *Br J Ophthalmol*. 2010 Jan; 94(1):59–63. [PubMed: 19692364]

Figure 1.
(a) Color photography shows subtle yellow-white punctate lesions deep to retina. (b-d) FA demonstrates hyperfluorescent lesions in early and late phases, and late optic nerve staining (b-arterial, c-venous laminar, d-late).

Figure 2. (a-c) FAF shows early hyperfluorescent lesions in a wreath-like pattern, with near resolution over 6 weeks. (d-f) OCT highlights early photoreceptor IS-OS junction disturbance followed by structural restoration (arrow heads).