https://doi.org/10.4038/jcosl.v28i2.66

Case report

Advances and challenges of using multimodal imaging for the diagnosis of Multiple Evanescent White Dot Syndrome

S. C. De Silva[1], K. Niruththan[2], D. C. Elvitigala[3], D. H. H. Wariyapola[4]

The Journal of the College of Ophthalmologists of Sri Lanka 2022; 28: 88-90

Introduction

From the early days of ophthalmology, viewing and imaging of the retina has been a challenge. However, during the past two decades, there has been a rapid development of novel imaging techniques which have provided invaluable insight into diagnosing and treating retinal diseases. The term "multimodal imaging" is now used increasingly to describe the usage of older and newer imaging techniques in tandem or in sequence to diagnose a single retinal disease. This approach exemplifies the utilization of certain information obtainable only by one or few imaging modalities and combines the information in a complementary manner based on clinical judgement to arrive at a diagnosis. The modalities commonly utilized include widefield color fundus photography, fundus autofluorescence (FAF), high resolution spectral domain optical coherence tomography (SD-OCT), fundus fluorescein angiography (FFA), indocyanine green angiography (ICGA), and OCT angiography[1].

The usage of multimodal imaging has become indispensable in diagnosing medical retinal disease with fleeting or subtle clinical findings. The group of inflammatory chorioretinopathies of unknown aetiology described as "White Dot Syndromes" is characteristically difficult to diagnose because of its nature of transient clinical signs. Multiple Evanescent White Dot Syndrome (MEWDS) is an uncommon inflammatory chorioretinopathy that affects otherwise healthy young females grouped under the white dot syndromes. Herein we demonstrate how multimodal retinal imaging was aptly utilized to diagnose a case of MEWDS were no single serological or radiological investigation is available for diagnostic certainty.

Case report

A 49-year-old female presented to the outpatient eye clinic at Sri Jayawardenapura General Hospital, Kotte, Sri Lanka with nonspecific visual blurring and photopsias of 6 days duration in her right eye. She was otherwise healthy with no significant past medical history. Her ocular examination showed mild hyperopia. The right eye had a grade 1 RAPD. Her best corrected distant visual acuity was 6/9 OD, 6/6 OS. Her anterior segment examination was normal in both eyes. The posterior segment of her right eye showed a normal looking optic nerve head and multiple discrete white spots spread perifoveally which further extended into the far periphery of the retina (Figure 1).

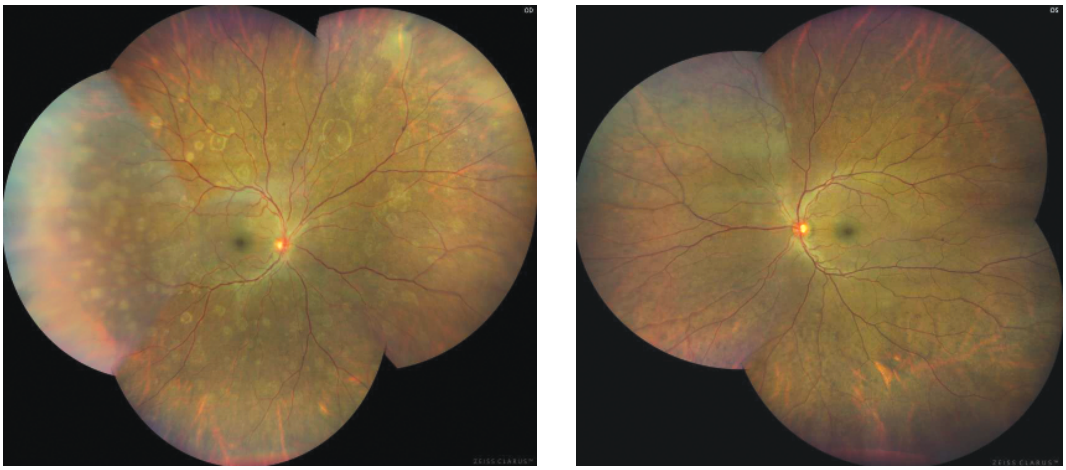

Figure 1. *Color fundus photograph of the right eye (left image) showing MEWDS lesions. The uninvolved left eye (right image) shows a normal looking fundus.*

[1]*Registrar,* [2]*Senior Registrar,* [3]*Registrar,* [4]*Consultant Eye Surgeon, Sri Jayawardenapura General Hospital, Kotte, Sri Lanka.*

There were a few posterior vitreous cells as well. Her posterior segment of the left eye was normal. Fundus autofluorescence images (FAF) of her right eye showed hyperautofluorescent spots corresponding to the fundus lesions. Also visualized on the FAF image were hypoautofluorescent spots which were not visible on clinical examination or color fundus photography (Figure 2). Fundus fluorescein angiography showed characteristic late-staining hyperfluorescence of these lesions (Figure 3). SD-OCT images of her right eye showed outer retinal ellipsoid zone changes where the lesions were present (Figure 4). Imaging of her left eye was normal.

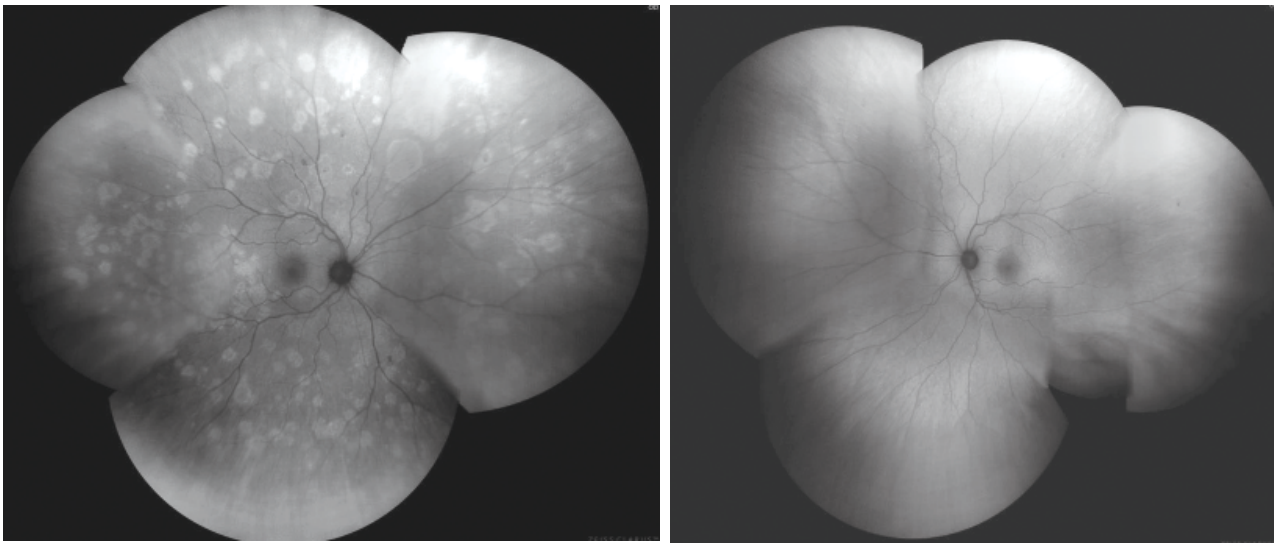

Figure 2. *Fundus autofluorescence (FAF) image of the patient's right eye (left image) shows hyperautofluorescent MEWDS lesions corresponding to the lesions on color fundal photography. The right image shows her normal left eye.*

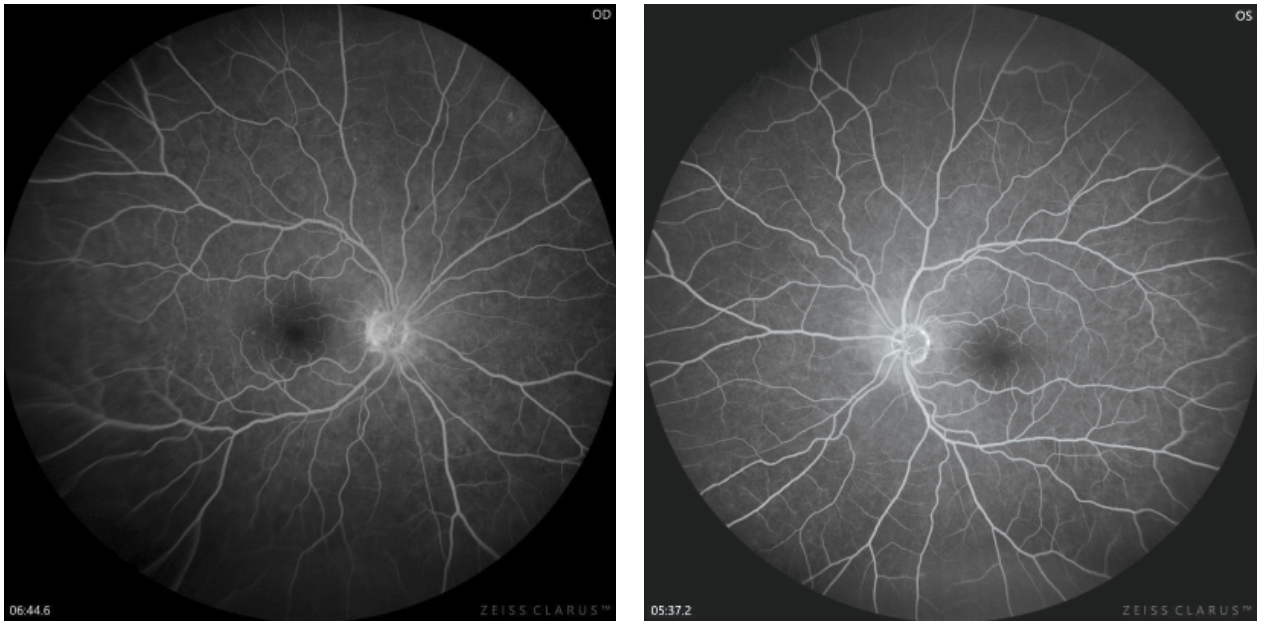

Figure 3. *Right eye (left image) shows late-staining of the lesions which is faintly discernable compared to the patient's left eye (right image).*

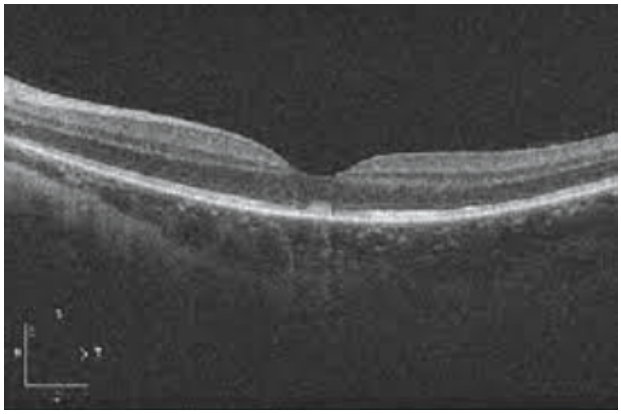

Figure 4. *SD-OCT showing ellipsoid zone changes in outer retina.*

Testing of her visual fields using standard automated static perimetry was normal OU. She was evaluated for posterior uveitis with extensive biochemical and radiological testing including a complete blood count, ESR, CRP, screening for syphilis and tuberculosis to exclude other infective and inflammatory aetiologies. These tests results were normal. The discrete white spots started fading after 2 weeks without treatment with complete resolution of the lesions on FAF images after 1 month. Her vision improved to 6/6 OU concurrent with the resolution of her symptoms. The clinical picture in combination with multimodal retinal imaging confirmed the diagnosis of MEWDS.

Discussion

MEWDS is an inflammatory chorioretinopathy of unknown aetiology typically affecting young moderately myopic females[2]. Frequently a viral prodrome is seen though an infective agent is yet to be identified[3]. The pathophysiology is thought to be due to a choriocapillaritis causing vaso-occlusive disease at the level of the inner choroid producing ischaemia in the outer retina with damage to the outer segments of the photoreceptors[4]. This process corresponds to the late staining on FFA and ellipsoid zone changes seen on SD-OCT.

Patients typically present with acute unilateral blurred vision and photopsias with a central or paracentral scotoma. Funduscopy during the acute phase will visualize the discrete white-to-orange spots of 100-200μm in diameter spread over the midperipheral retina. The presence of a granular macular pigmentary change is pathognomonic for the disease and is especially useful when these transitory lesions have faded. A variable degree of vitreous inflammation is also a common finding.

MEWDS is typically a monophasic disease with excellent visual prognosis giving weight to its moniker a "common cold of the retina". Visual recovery occurs within 2-10 weeks without any treatment. Residual symptoms such as photopsias and enlargement of the blind spot may persist for months. Uncommon recurrences which occur in 10-15% have similarly good prognosis[2].

The challenges underlying diagnosis of MEWDS is due to the transitory nature of the lesions. Several case series show that absence of lesions on color fundus photography can occur in upto 70% of patients. Coupled with good visual recovery and the monophasic nature of the disease, the diagnosis can frequently be missed or misdiagnosed. Since there is no serological, electrophysiological or single imaging study for confirmatory diagnosis of MEWDS, multimodal retinal imaging presents itself as an indispensable tool in the armamentarium of the medical retina specialist. However the non-availability, cost, and potential complications of certain imaging modalities are also problematic especially in resource poor countries.

References

1. DiCarlo E, Augustin AJ. The emergence of multimodal imaging in ophthalmology. Ophthalmology times Europe, accessed 28 April 2023.
2. Uveitis and ocular inflammation. Basic and Clinical Sciences Course 2021-2022, American Academy of Ophthalmology, San Francisco, CA, 2022.
3. Herbort CP Jr. Multiple Evanescent White Dot Syndrome (MEWDS) In: Zierhut M, Pavésio C, Ohno S, Oréfice F, Rao NA, editors. Intraocular Inflammation. Berlin Heidelberg, *Springer* 2016: 997-1005.
4. Papasavvas I, Mantovani A, Tugal-Tutkun I, Herbort CP Jr. Multiple evanescent white dot syndrome (MEWDS): update on practical appraisal, diagnosis and clinico-pathology; a review and an alternative comprehensive perspective. *J Ophthalmic Inflamm Infect*. 2021; **11**(1): 45. doi: 10.1186/s12348-021-00279-7. PMID: 34921620; PMCID: PMC8684571.