Case Report

Choroidal neovascularization as the initial manifestation of multiple evanescent white dot syndrome

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Abstract: To report the case of a patient who presented with idiopathic choroidal neovascularization (CNV) as the first sign of multiple evanescent white dot syndrome (MEWDS). A 25-year-old woman presented with recent onset of decreased vision and metamorphopsia in the right eye. The results of fundoscopic examination, fluorescein angiography, and optical coherence tomography (OCT) were compatible with a diagnosis of idiopathic CNV, which was treated with one intravitreal injection of bevacizumab. Five years later, the patient returned complaining of photopsia and decreased vision in the same eye. The fundoscopic examination showed typical signs of MEWDS. After 3 months, recurrence of CNV was observed in the same eye. In conclusion, idiopathic CNV might be the only manifestation of a subclinical occurrence of MEWDS. In this case, it was followed by a recurrence of MEWDS and subsequent reactivation of CNV.

Keywords: Idiopathic; choroidal neovascularization (CNV); multiple evanescent white dot syndrome (MEWDS); tomography; optical coherence; fluorescein angiography

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Introduction

Choroidal neovascularization (CNV) is an abnormal ingrowth of the choroidal vasculature that penetrates through Bruch's membrane into the subretinal space. When CNV occurs in patients aged up to 50 years, etiologies such as high myopia, ocular histoplasmosis, trauma, and inflammatory conditions could be responsible. However, in younger patients, idiopathic CNV remains the second most common etiology (1).

Multiple evanescent white dot syndrome (MEWDS) is an acute onset inflammatory disease predominantly affecting the outer retina (2). The disease typically affects young to middle-aged myopic women in the second to fourth decades of life. Patients complain of acute, mostly unilateral, painless loss of vision accompanied by photopsia and central or paracentral scotoma (3). Fundoscopy reveals multiple gray to white dots located at the posterior pole that are later replaced by granular macular pigmentary changes. The disease is self-limiting and patients typically regain their previous visual acuity. Rarely, persistent blind spot enlargement is observed (3). It has been suggested that some cases of idiopathic CNV are secondary to a previous occurrence of MEWDS (4-6).

The present report describes a 25-year-old woman initially diagnosed with idiopathic CNV who developed clinical signs of MEWDS 5 years later. It is believed that CNV was the first observed clinical sign of MEWDS.

Case presentation

A 25-year-old woman complaining of recent-onset decreased vision and metamorphopsia in her right eye was referred to a retina clinic in April 2009. The patient stated no pertinent medical or ocular history and no significant family history. The best corrected visual acuity (BCVA) was 20/30 and 20/20 in the right and left eye, respectively. No relative afferent pupillary defect was detected. Slit lamp

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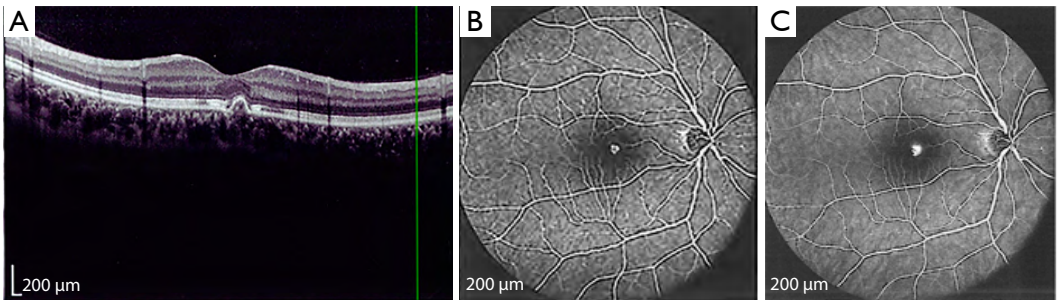


Figure 1 CNV manifestation in the right eye of a 25-year-old woman. (A) OCT of the right eye shows a small CNV lesion manifesting as subretinal fluid and focal irregularity of the central macula; (B) fluorescein angiography of the right eye demonstrate a hyperfluorescent lesion with a lacy pattern in the early phase; (C) late leakage of the juxtafoveal CNV lesion. CNV, choroidal neovascularization; OCT, optical coherence tomography.

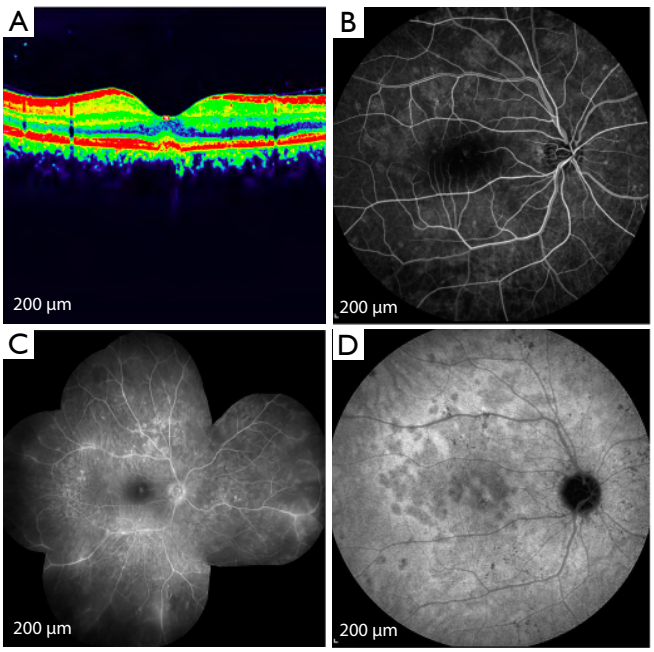


Figure 2 Typical presentation of MEWDS in the right eye of a 25-year-old woman. (A) OCT of the right eye shows focal retinal pigment epithelial irregularities in the central macula as a typical sign of MEWDS 5 years after the original treatment; (B) fluorescein angiography of the right eye illustrate multiple hypofluorescent spots in the early phase; (C) late staining as a sign of the MEWDS recurrence; (D) numerous hypofluorescent spots in the macula and the peripapillary region in the ICG angiography image at the time of the recurrent attack of MEWDS. CNV, choroidal neovascularization; MEWDS, multiple evanescent white dot syndrome; OCT, optical coherence tomography; ICG, indocyanine green.

biomicroscopic examination of the right eye revealed no signs of anterior segment pathology, vitreous inflammation, or ocular media opacity.

Fundoscopy revealed a small, round, grayish, elevated lesion with indistinct borders located in the juxtafoveal area of the right eye. There were no additional abnormalities affecting the optic disc, retinal vessels, or retinal periphery. Optical coherence tomography (OCT) showed a small amount of subretinal fluid and a focal irregularity of the retinal pigment epithelium layer of the central macula (Figure 1A). Fluorescein angiography demonstrated an early lacy pattern hyperfluorescent spot with late leakage compatible with a juxtafoveal CNV lesion (Figure 1B,C). Results of the ocular examination were unremarkable in the left eye. After a diagnosis of idiopathic CNV was made, the right eye underwent an intravitreal injection of 1.25 mg/0.05 mL bevacizumab (Genentech Inc., San Francisco, CA, USA). At 4 weeks post-injection, the BCVA improved to 20/20 and the metamorphopsia resolved.

Five years later, the patient returned complaining of deterioration in visual acuity and photopsia in the right eye. The BCVA was 20/25. Slit lamp examination was unremarkable. Fundoscopy showed granular lesions within the macular area with pigmentary changes. OCT showed only small focal retinal pigment epithelial irregularities (Figure 2A). Fluorescein angiography revealed multiple hypofluorescent spots in the early phase with late staining in the macular region (Figure 2B,C). Indocyanine green (ICG) angiography disclosed numerous hypofluorescent spots in the macula and peripapillary region compatible with choriocapillaritis patches (Figure 2D). Automated

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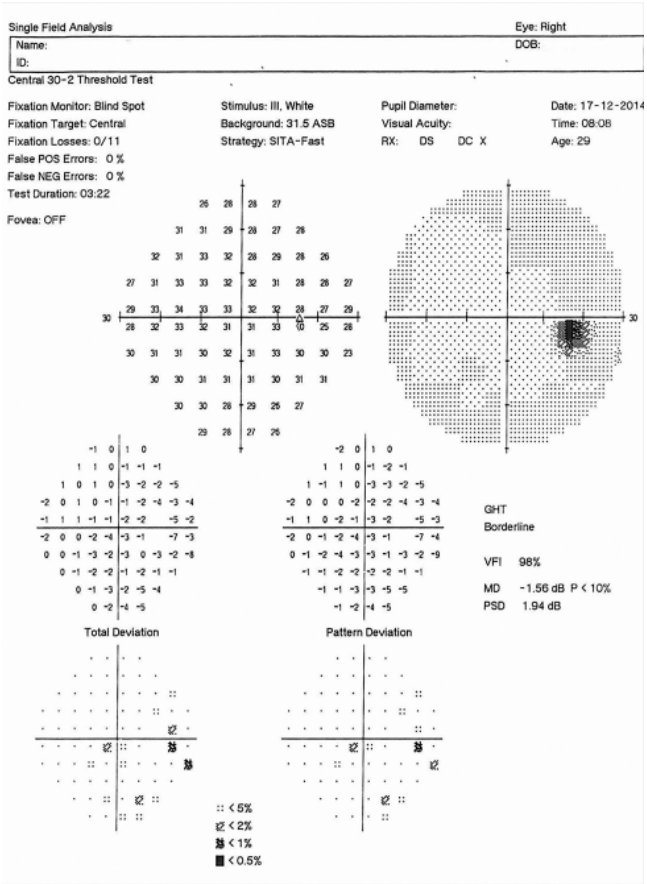


Figure 3 Humphrey visual field test report of the right eye showing enlargement of the blind spot due to MEWDS attack. MEWDS, multiple evanescent white dot syndrome.

perimetry (Carl Zeiss Meditec Inc., Dublin, USA) revealed enlargement of the blind spot (Figure 3). There were no abnormal findings in the left eye. A diagnosis of MEWDS was recorded for the right eye.

Three months later, the patient returned with a complaint of metamorphopsia and vision deterioration in the right eye. Reactivation of the previous CNV lesion was detected without further remarkable change in the fundus. OCT revealed neurosensory detachment (Figure 4). Following injection of 1.25 mg/0.05 mL intravitreal bevacizumab, the CNV resolved and the BCVA returned to 20/20.

Discussion

In the current case study, we suggest that idiopathic CNV may occur following a subclinical episode of MEWDS. The presumed relationship was confirmed by the occurrence

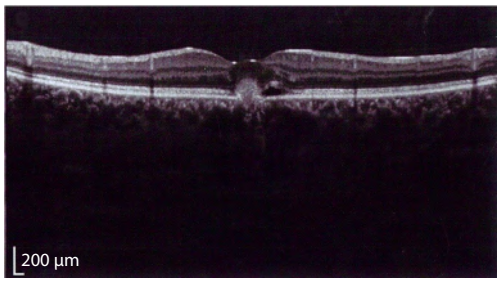


Figure 4 OCT of the right eye showing active CNV lesion with adjacent small neurosensory detachment of the macula 3 months after recurrence of MEWDS. CNV, choroidal neovascularization; MEWDS, multiple evanescent white dot syndrome; OCT, optical coherence tomography.

of a typical attack of MEWDS 5 years after a diagnosis of idiopathic CNV was made in the same eye. MEWDS is characterized as a self-limiting monophasic disease for which recurrence is unusual (2). However, in our patient, recurrence of MEWDS was observed.

Atypical recurrent forms of MEWDS have been reported in several case studies (6-10). It is interesting to note that the number, laterality, and clinical presentation of the recurrent attacks varied widely among the previous reports. The time between the first episode and its recurrence ranged from 4 months to 9 years. In addition, maintenance therapy with immunosuppressive agents was needed to successfully maintain remission in two cases with multiple uncontrolled outbreaks (5,7). It remains unknown as to why so few cases of MEWDS tend to recur and why different clinical presentations have been described for recurrences of the disease. It is noteworthy that the actual rate of recurrence might be underestimated and that subclinical or atypical attacks could be missed.

The number of MEWDS cases that are complicated by CNV is unknown and their description is confined to a few reports (2,4-12). In the present study, it is believed that idiopathic CNV occurred following an unrecognized subclinical episode of MEWDS, which had resolved by the time of CNV presentation. Therefore, it is advisable to consider MEWDS, particularly in young myopic patients, before making a diagnosis of idiopathic CNV. Machida et al. and Papadia et al. observed comparable cases of presumed idiopathic CNV in which clinical signs of MEWDS manifested only during follow-up examinations (4,5). This finding emphasizes the importance of following patients with idiopathic CNV in order to determine the underlying

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etiology of the disease.

CNV was reactivated following the recurrence of MEWDS. Treatment of CNV involved one injection of intravitreal bevacizumab for each occurrence. Rouvas et al. showed that the use of intravitreal ranibizumab (Lucentis; Genentech; USA) resulted in the regression of CNV associated with MEWDS (11).

In conclusion, it is suggested that a subclinical episode of MEWDS could be, in part, responsible for clinically diagnosed idiopathic CNV. This conclusion was drawn from the 5-year follow-up of a patient with presumed idiopathic CNV in which recurrence of MEWDS was observed. Long-term follow-up is recommended to clarify the etiology of some cases of presumed idiopathic CNV.

Acknowledgements

None.

Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

Informed Consent: Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

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