색소상피박리가 동반된 소실성다발흰점증후군 환자에서 발생한 맥락막신생혈관

Choroidal Neovascularization after Multiple Evanescent White Dot Syndrome in a Patient with Pre-existing Pigment Epithelial Detachment

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Purpose: To report a case of choroidal neovascularization (CNV) occurring after resolution of multiple evanescent white dot syndrome (MEWDS) in a patient with pre-existing pigment epithelial detachment.

Case summary: A 31-year-old woman with decreased vision in the right eye as her main complaint visited our clinic. Initial examination showed mild inflammation of the anterior chamber and disc swelling with multiple white dots on the fundus. Hyper-fluorescent spots on fluorescein angiography (FA) and hypo-fluorescent spots on indocyanine green angiography (ICGA) were observed. Optical coherence tomography (OCT) showed small pigment epithelial detachment (PED) at the macula, but there was no angiographic evidence of active CNV. She was diagnosed with MEWDS. After 1 month of treatment with an oral corticosteroid, the white dots completely disappeared. Two weeks later, she complained of a central scotoma in the same eye. OCT showed intraretinal and subretinal hemorrhages in the macula and subretinal fluid (SRF), with FA and ICGA showing a hyper-fluorescent spot on the fovea with leakage, findings compatible with active CNV. SRF did not resolve after six serial intravitreal injections of bevacizumab, half-fluence photodynamic therapy, and or serial intravitreal injections of aflibercept, but resolved completely after three serial intravitreal injections of ranibizumab.

Conclusions: Active CNV can occur after MEWDS resolution in patients with predisposing conditions. Close follow-up after MEWDS resolution may be needed to detect CNV, particularly in patients with macular abnormalities such as PED.

Keywords: Choroidal neovascularization; Multiple evanescent white dot syndrome (MEWDS)

Introduction

Multiple evanescent white dot syndrome (MEWDS) is an inflammatory chorioretinopathy characterized by the presence of unilateral, multiple, faint white dots in the mid-peripheral fundus [1,2]. Despite MEWDS usually being a self-limiting

disease with good prognosis, choroidal neovascularization (CNV) secondary to MEWDS may occur [3-6]. This report describes a patient with pre-existing pigment epithelial detachment (PED) who experienced CNV 2 weeks after the resolution of MEWDS.

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Received: 2016. 10. 20

Revised: 2016. 12. 28

Accepted: 2017. 1. 9

Case Report

A 31-year-old woman who experienced decreased vision in her right eye 2 days earlier (best corrected visual acuity [BCVA] was 20/200) visited our clinic. She had also experienced flu-like symptoms a few days earlier. Initial slit-lamp examination of her right eye showed mild inflammation in the anterior chamber and disc swelling with multiple white dots. Optical coherence tomography (OCT) showed PED, fluorescein angiography (FA) revealed hyper-fluorescent spots, and indocyanine green angiography (ICGA) showed hypo-fluorescent spots corresponding to multiple white dots on the fundus (Fig. 1A-C). There was no angiographic evidence of active CNV, but ICGA showed small, multiple

Figure 1. Findings in the right eye of this patient during the initial examination and after 1 month of treatment with oral steroid. Fundus photograph showing multiple white dots and mild disc swelling (A). Optical coherence tomography showing pigment epithelial detachment (B). Fluorescein angiography showing multiple hyper-fluorescent spots (left) and indocyanine green angiography (ICGA) showing hypo-fluorescent spots (right), corresponding to the white dots on the fundus. There was no evidence of active choroidal neovascularization, but small, multiple pinpoint-like hyper-fluorescent spots were found along the margin of fovea on ICGA (right) (C). Complete resolution of multiple white dots at the posterior pole and disc swelling after 1 month oral steroids therapy. Mild foveal hemorrhage was observed (D).

pinpoint-like hyper-fluorescent spots along the margin of the fovea (Fig. 1C). She was diagnosed with MEWDS superimposed on previously existing PED. After 1 month of treatment with oral prednisolone and steroid eye-drops, the white dots completely disappeared and her visual acuity improved to 20/60. However, mild inflammation was still observed in the anterior chamber and there was evidence of mild foveal hemorrhage (Fig. 1D).

Two weeks after the complete disappearance of the white dots, the BCVA in the right eye remained stable at 20/60, but she complained of a small central scotoma in that eye. OCT showed intraretinal hemorrhage with serous detachment of the macula and subretinal fluid (SRF). FA and ICGA revealed hyper-fluorescent spots on the fovea with progressive leakage, findings consistent with active CNV (Fig. 2). She was diagnosed with active CNV after MEDWS and treated with a series of intravitreal injections of bevacizumab, an antibody to vascular endothelial growth factor (VEGF).

One month after the first bevacizumab injection, her visual acuity improved to 20/20 and SRF on OCT decreased. However, despite six serial intravitreal injections of bevacizumab, SRF did not completely resolve (Fig. 3A, B). She was therefore started on half-fluence photodynamic therapy

Figure 2. Findings in the right eye of this patient at presentation of a central scotoma. Fundus photograph, showing intraretinal and subretinal hemorrhages and serous detachment on the macula (A). Optical coherence tomography, showing subretinal fluid, intraretinal fluid, and intraretinal hyper-/hypo-reflective lesions (B). Fluorescein angiography (left) and indocyanine green angiography (right), showing a hyper-fluorescent spot on the fovea (C).

Figure 3. Findings in the right eye of this patient after six serial intravitreal bevacizumab injections, half-fluence photodynamic therapy (PDT) and two serial intravitreal aflibercept injections. Fundus photograph showing that the subretinal and intraretinal hemorrhages had resolved and the serous detachment on the fovea had decreased but remained inferiorly displaced (A). Optical coherence tomography (OCT) showing subretinal fluid (SRF) and choroidal neovascularization (CNV) (B). Fundus photograph and OCT after half-fluence PDT, showing no change in SRF and CNV (C, D). Fundus photograph and OCT after two serial aflibercept injections, showing that SRF did not resolve completely (E, F).

(PDT). One month later, her visual acuity improved to 20/40 and SRF decreased slightly. Two months after PDT, her visual acuity again decreased to 20/60, and SRF increased (Fig. 3C, D). She was therefore treated with intravitreal injections of another anti-VEGF agent, aflibercept. However, after two injections, her visual acuity and SRF did not improve (Fig. 3E, F). She was therefore started on intravitreal injections of a third anti-VEGF agent, ranibizumab. After three serial intravitreal injections of ranibizumab, her visual acuity improved to 20/40 and SRF was completely resorbed (Fig. 4).

Discussion

MEWDS is an inflammatory chorioretinopathy characterized by the presence of unilateral, multiple, faint white dots

Figure 4. Fundus examination findings after three serial ranibizumab injections. The subretinal fluid (SRF) was completely resorbed and the macula showed no evidence of SRF accumulation in fundus photograph (A) and optical coherence tomography (B).

in the mid-peripheral fundus. MEWDS is usually self-limiting, with good visual outcomes [1,2].

CNV can occur secondary to inflammatory chorioretinopathies, such as MEWDS [3-6], multifocal choroiditis [7] and punctate inner choroidopathy [8]. CNV secondary to MEWDS, however, is rare [5,6] because the duration of MEWDS and the extent of inflammation are relatively limited [9], but this condition can cause a severe loss of visual acuity in such patients.

Previous reports describing patients with pre-existing CNV before MEWDS have suggested that baseline choroidopathy inflammatory conditions are related to the occurrence of CNV and MEWDS [10,11]. In some patients, inflammation at the level of the choroid was found to result in ischemia of the choriocapillaris-retinal pigment epithelium complex, with this ischemia triggering choroidal neovascularization [10,12,13].

Several characteristics of the present patient suggested that she might have already had vascularized PED in the macula at the time of her MEWDS diagnosis, rather than de novo CNV secondary to MEWDS. First, OCT showed that PED had multiple, not just round curvatures, and that the mild hyper-reflective lesion inside the PED was compatible with vascularized PED. Second, ICGA at the time of MEWDS diagnosis showed small, multiple pinpoint-like hyper-fluorescent spots along the margin of the fovea, with these marginal hyper-fluorescent spots being the same as the areas of PED on OCT. Third, the sites of suspicious foveal hemorrhages 1 month after oral corticosteroid therapy were the same as those of the intraretinal hemorrhages that occurred 2 weeks later. Fourth, the CNV site on ICGA was identical to the site of the previous PED.

CNV in our patient was resistant to intravitreal injection

tions of bevacizumab and aflibercept, as well as PDT, while being responsive to intravitreal ranibizumab. CNV before MEWDS has been treated successfully with intravitreal bevacizumab [10] and CNV secondary to MEWDS has been found to be responsive to a single intravitreal injection of ranibizumab [4] or single PDT [3]. To our knowledge, there have been no previous reports on activated CNV after MEWDS in patients with pre-existing PED, precluding a comparison of our results with those of previous patients. The resistance to bevacizumab and PDT in our patient may have been due to the occurrence of an inflammatory condition during activation of CNV after MEWDS.

In summary, inflammatory conditions developing after MEWDS may be related to the activation of CNV. Following complete resolution of MEWDS, patients, especially those with macular abnormalities, should be closely followed-up to detect the onset of CNV.

Conflicts of interest

The authors have no proprietary or commercial interest in any material discussed in this article.

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