

A Case of Multiple Evanescent White Dot Syndrome Treated by Steroid Pulse Therapy

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A Case of Multiple Evanescent White Dot Syndrome Treated by Steroid Pulse Therapy

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Abstract

Multiple evanescent white dot syndrome (MEWDS) is characterized by acute visual loss which is usually spontaneously restored after a few months. While occasional treatments with local or oral steroids have been reported, these are hardly effective. A 25-year-old man, a Sumo wrestler, was diagnosed with MEWDS, and as he wanted a quick recovery to compete in an upcoming tournament treatment with steroid pulse therapy was initiated 8 days after the symptoms appeared. Before treatment, visual acuity in his left eye was 20/400. Multiple white dots were seen at the level of the deep retina or retinal pigment epithelium, and the Mariotte's blind spot extended to the center of the visual field. Immediately after the end of steroid pulse therapy, left visual acuity increased to 20/25, and left visual field recovered remarkably. No white dots were seen funduscopically. Steroid pulse therapy might provide early improvement of visual functions, and we believe it could be a treatment option for initiating an early recovery from MEWDS. However, steroid pulse therapy may also result in lethal damage including disorders of the circulatory organs. Thus, it should only be applied in limited situations in which patients had rapid decline in visual function and needs for social return at an early stage like that of our patient.

Key Words: Immune response; Multiple evanescent white dot syndrome; Steroid pulse therapy

Introduction

Multiple evanescent white dot syndrome (MEWDS) is characterized by acute visual loss that is usually spontaneously restored after several weeks or more¹⁾. However, protracted visual loss, recurrent and intractable cases have been reported²⁾, and choroidal neovascularization, while rare, might occur in such cases³⁾. Local or oral steroids have sometimes been used²⁻⁵⁾. However, it took several weeks to improve visual functions and fundus conditions in some such cases²⁻⁴⁾.

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Thus, both treatments were considered to be hardly effective in comparison with the natural course. In addition, treatments with steroid pulse therapy have not been previously documented. Since inflammation may occur as indicated by occasional intravitreal inflammatory cells¹⁾ and dye staining of the optic disc in fluorescein angiograms⁵⁾, intense steroid treatment could promote rapid recovery from the disease.

Here, we report a case of MEWDS with acute and severe visual loss, where the patient achieved an early and remarkable recovery of visual functions following steroid pulse therapy.

Case Report

A 25-year-old man had been experiencing acute visual loss and temporal scotoma in his left eye. There was no viral prodrome. Before treatment, best-corrected visual acuity was 20/20 OD, and 20/400 OS. Left fundus examination revealed multiple white dots located at the level of the deep retina or retinal pigment epithelium (Fig. 1A). The optic disc was normal. Fluorescein angiography revealed early-phase punctate hyperfluorescence corresponding to the white dots, and late-phase staining of the same spots and the optic disc. Early-phase indocyanine green angiography showed no remarkable changes. In the late phase, punctate hypofluorescence reflected both clinically apparent white dots, and clinically undetectable dots. Electroretinography findings were normal. Goldmann visual field examination revealed enlargement of the left Mariotte's blind spot, reaching the center of the field (Fig. 1B).

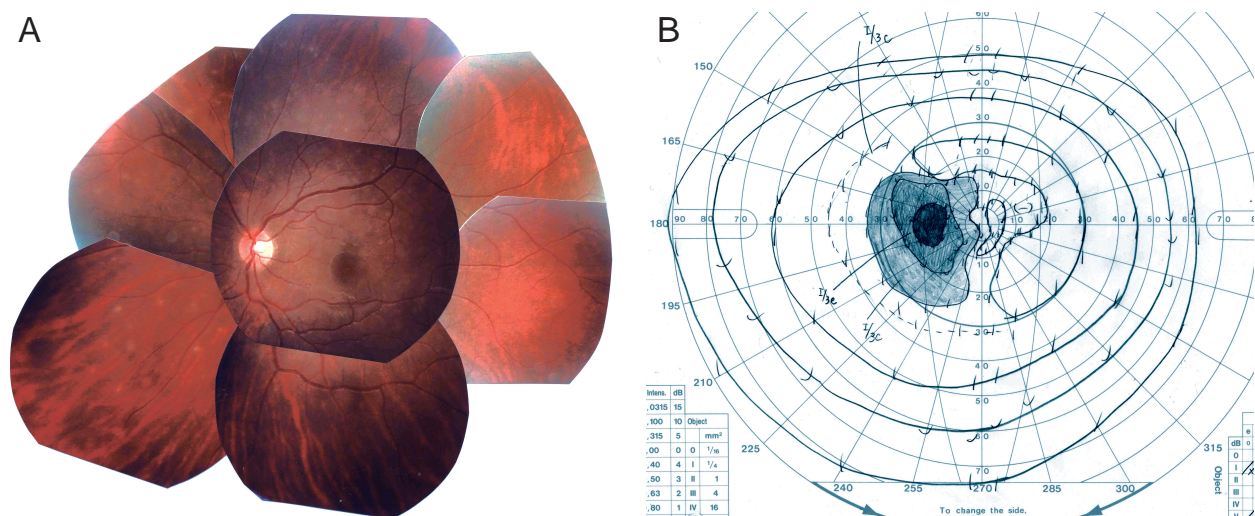


Figure 1. Initial examinations. A, Fundus examination of the left eye reveals multiple, slightly indistinct white dots, each with a diameter 1/7-1/3 of that of the optic disc, and located at the level of the deep retina or retinal pigment epithelium. B, Goldmann visual field examination of the left eye reveals enlargement of the Mariotte's blind spot reaching the center of the field.

Since the patient was a Sumo wrestler, and wanted a quick recovery to compete in an upcoming tournament. Steroid pulse therapy (1000 mg/day methylprednisolone for 3 days) was started 8 days after symptoms appeared. Oral steroid therapy as maintenance treatment was not performed after steroid pulse therapy. Immediately after termination of treatment, visual acuity increased to 20/25, retinal abnormalities disappeared funduscopically (Fig. 2A), and fluorescein was not present angiographically. The scotoma was remarkably reduced (Fig. 2B),

and there were no treatment side effects. Visual acuity was 20/16, and visual field findings were back to normal 20 days after the start of treatment. MEWDS did not recur in 1.5 years of follow-up.

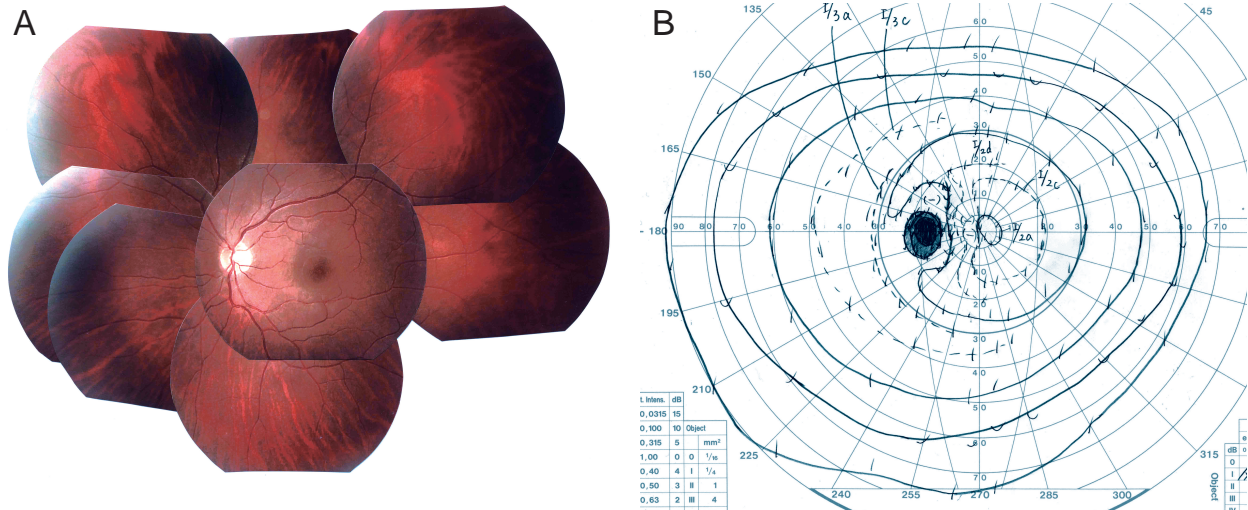


Figure 2. Examinations 4 days after initiation of treatment. A, White dots have disappeared funduscopically. B, Visual field recovers earlier, and remarkably.

Discussion

MEWDS affects the deep retina or the retinal pigment epithelium, and retinal inflammation may remain after the disappearance of the white dots⁶; thus, visual loss can recur in MEWDS². The lasting inflammation may also lead to irreversible changes including choroidal neovascularization³.

Immune responses may be closely related to retinal changes in MEWDS². Low-dose steroid therapy (<1.0 mg/kg/day) only results in anti-inflammatory effects, thus high-dose therapy is necessary to achieve both anti-inflammatory and immunosuppressive effects^{7,8}. Steroid pulse therapy enables the maintenance of 100 times the blood concentration produced in oral steroid therapy, and allows the saturation of 99.9% of glucocorticoid receptors for an extended time⁹. Since long-term oral administration of large amounts of steroids tends to lead to side effects, steroid pulse therapy may be preferable. However, steroid pulse therapy may also result in lethal damage including disorders of the circulatory organs. Moreover, the effects in patients with recurrent MEWDS and for the prevention of relapses or recurrences of MEWDS are uncertain. Thus, steroid pulse therapy should be performed carefully in limited situations in which patients had rapid decline in visual function⁴ and needs for social return at early stage like that of our patient.

In the present case study, through the use of steroid pulse therapy an early and remarkable improvement of the visual field was obtained in our patient, and visual acuity and fundus abnormalities improved. Recovery of visual field defects is generally slow in most MEWDS patients, requiring more than 10 weeks^{2,3}. However, visual acuity and abnormal findings of fundus, fluorescein angiography, and electroretinography usually spontaneously recover after less than 10 weeks^{1,10,11}.

The present study showed that steroid pulse therapy might provide for an early and remarkable improvement of visual function. We believe that steroid pulse therapy is a treatment option for early recovery from MEWDS, but its use must be limited to specific appropriate situations.

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