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SHORT COMMUNICATION

Case report

Treatment of multiple evanescent white dot syndrome with cyclosporine

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PURPOSE. *To report the seven-year follow-up of a patient with multiple evanescent white-dot syndrome (MEWDS).*
METHODS. *Case report: A 46-year-old woman presented recurrent episodes of bilateral MEWDS.*
RESULTS. *During the seven-year follow-up there were nine episodes of MEWDS. After four bouts in the first two, cyclosporine therapy was started. During two years of treatment there were no recurrences except when the dose was reduced or discontinued.*
CONCLUSIONS. *The etiology of MEWDS is still unknown but the absence of new episodes during cyclosporine treatment and the recurrence immediately after decreasing or discontinuing the drug suggests an autoimmune origin, with the involvement of cellular immunity in the pathogenic process. (Eur J Ophthalmol 2001; 11: 86-8)*

KEY WORDS. *Cyclosporine, Juxtapapillary choroidal neovascularization, MEWDS*

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INTRODUCTION

Multiple evanescent white-dot syndrome (MEWDS) is an inflammatory disorder of the outer retina and RPE. It is seen in young people, predominantly females, and is usually unilateral, rather than bilateral. We report the seven-year follow-up of a 46-year-old woman who showed multiple, bilateral episodes of MEWDS.

Case report

In the first two years, the patient had three episodes of MEWDS, two of them bilateral. At each recurrence the typical multiple deep 200-micron retinal white lesions were observed (Fig. 1, left), with disc edema and vitreous cells. Fluorescein angiography showed initial hypofluorescence and late hyperfluorescence of the lesions (Fig. 1, right). The patient also had clinically visible cysts and angiographic signs of cystoid macular edema at each recurrence (Fig. 2). The retinal lesions completely disappeared after each episode, with total recovery of visual acuity and fundus appearance. In the second episode a bilateral juxtapapillary choroidal neovascularization was also seen and treated with argon laser photocoagulation.

Seven months later a new episode was diagnosed in the left eye. Anterior chamber and vitreous flare and cells were found in both eyes, and the left fundus had a white lesion which was larger and deeper than those previously described, below the superotemporal arcade (Fig. 3, left). Fluorescein angiography of the left eye showed lesion hypofluorescence and recurrence of choroidal neovascularization (Fig. 3, right).

After these two years of follow-up with several episodes of MEWDS with cystoid macular edema, disc edema and an occasional anterior tyndall, we decided to undertake therapy with cyclosporine. We began with a dose of 5 mg/kg/24h that was gradually decreased to 3 mg/kg/24h. Ten months after the start of cyclosporine treatment, arterial hypertension appeared and the dosage was further cut to 2 mg/kg/day. Five days later a severe anterior chamber and vitreous tyndall was disclosed and typical MEWDS foci were detected. All

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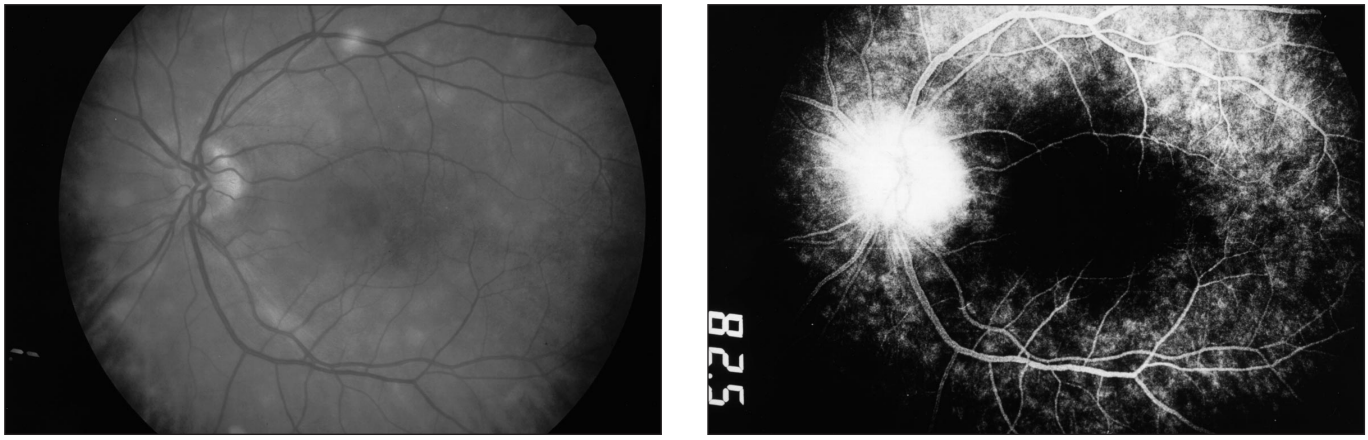

Fig. 1 - ***Left:*** *Small white retinal lesions typical of MEWDS.* ***Right:*** *Fluorescein angiography showing multiple punctate hyperfluorescent dots.*

lesions disappeared when the cyclosporine dosage was raised to 5 mg/kg/day. Once the new episode was controlled, the dosage was gradually tapered down to a maintenance level of 3 mg/kg/day. After a year of 3 mg/kg/24h of cyclosporine and no new episodes, we decided to discontinue treatment in October 1995. Between October 1995 and October 1998 four new episodes of MEWDS occurred.

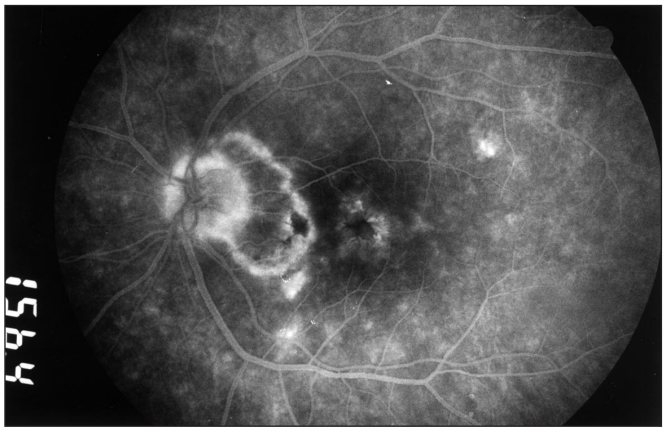

Fig. 2 - *Fluorescein angiogram showing the typical petaloid appearance of cystoid macular edema. Also evident is a recurrence of the peripapillary choroidal neovascular membrane adjacent to the inferior margin of the juxtapapillary laser scar.*

DISCUSSION

MEWDS was first described by Jampol in 1984 (1). Since the original description, there have been numerous reports of bilateral involvement as well as mul-

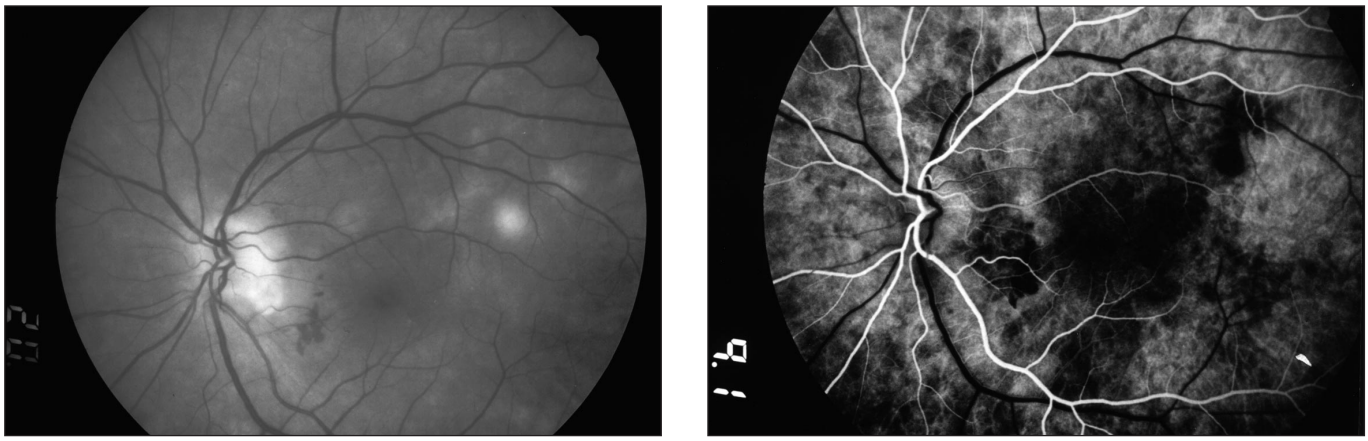

Fig. 3 - ***Left:*** *Fundus photograph showing a deep, round, white lesion below the superotemporal arcade. A subretinal hemorrhage in the inferior part of the papillomacular bundle due to the recurrence of a choroidal neovascular membrane is also evident.* ***Right:*** *Early frame of the fluorescein angiography shows the hypofluorescence of this focus.*

tiple recurrences (2-4). Our patient presented nine episodes of MEWDS in seven years, three of them bilateral. Contrary to the typical clinical features of MEWDS, our patient presented signs of cystoid macular edema and severe anterior inflammatory reaction at each recurrence. Although bilateral choroidal neovascularization has been associated with MEWDS (5, 6), it is very unusual. Another clinical feature of this patient was the appearance of a deep, larger lesion (Fig. 3), which may be a coalescence of various foci.

The frequent recurrences of MEWDS associated with disc edema, cystoid macular edema and panuveitis led us to start cyclosporine treatment. During the 22 months of treatment no new episodes of MEWDS were observed. However, only five days after tapering the dose of cyclosporine, a new episode of MEWDS with panuveitis was observed, disappearing when the previous dosage was re-established. Moreover, four more episodes of MEWDS occurred after the discontinuing cyclosporine.

The etiology of MEWDS is still unknown. A possible infectious origin has been claimed based on its association with flu-like symptoms (1), increased IgM levels (7) and its occurrence after varicella infection (6). If an infectious agent was the cause of this disease, the recurrences could be considered similar to those in patients with Herpes virus infections (4). An autoimmune etiology has also been suggested, based on the recurrent course that is sometimes seen in these diseases. In our patient, the absence of new episodes during cycloslporine treatment and its recurrence immediately after decreasing the dosage or discontinuing the drug, points to an autoimmune origin linked to cellular immunity.

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