POSSIBLE RECURRENT MULTIPLE EVANESCENT WHITE DOT SYNDROME AND CHOROIDAL NEOVASCULARIZATION

POSOBLE SÍNDROME DE MÚLTIPLES MANCHAS BLANCAS EVANESCENTES RECURRENTE Y NEOVASCULARIZACIÓN COROIDEA

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ABSTRACT

Case report: We present an unusual case of recurrent and bilateral multiple evanescent white dot syndrome (MEWDS), who had three bilateral episodes in a period of nine years. Thirteen years after the first episode, she was diagnosed with subretinal neovascularization (SRNV) which showed a satisfactory response to photodynamic therapy, with a final visual acuity of 20/40.

Discussion: MEWDS is considered classically to be an inflammatory disease with a good visual prognosis. Although the development of SRNV is unusual, we recommend long term follow-up of these patients since the early diagnosis and treatment of SRNV are both relevant for the retention of visual acuity (Arch Soc Esp Oftalmol 2007; 82: 587-590).

Key words: Multiple evanescent white dot syndrome, recurrent, bilateral, choroidal neovascularization, treatment.

RESUMEN

Caso clínico: Se presenta un caso de presunto síndrome de múltiples manchas blancas evanescentes (MEWDS) recurrente y bilateral (tres episodios bilaterales en nueve años). Trece años después del episodio inicial, presentó una membrana neovascular subretiniana (MNVSR) que respondió favorablemente a terapia fotodinámica, con una agudeza visual final de 20/40.

Discusión: El MEWDS se considera una enfermedad inflamatoria con buen pronóstico visual. Sin embargo, existe la posibilidad de desarrollar a largo plazo complicaciones como la MNVSR. Por ello, recomendamos el seguimiento periódico de estos pacientes por la importancia del diagnóstico y tratamiento precoz de las MNVSR en la agudeza visual final.

Palabras clave: Síndrome de múltiples manchas blancas evanescentes, recurrente, bilateral, neovascularización coroidea, tratamiento.

Received: 3/8/06. Accepted: 25/7/07.
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INTRODUCTION

The multiple evanescent white dot syndrome (MEWDS) is defined as an entity whose clinical progression is self-limited, benign, of unknown etiology and rarely recurrent. Nevertheless, the severe loss of vision secondary to a choroidal neovascularization has been described in the literature (1,2). The present case presumably involves a recurrent MEWDS that developed a subretinal neovascular membrane (SRNV) thirteen years after the initial episode.

CASE REPORT

In January 1992, a 36-year-old emetropic female arrived in our emergency unit reporting blurry vision without any previous flu episode. Exploration revealed 20/30 visual acuity (VA) in the right eye (RE) and 20/60 in the left eye (LE), 1+ cells in the anterior chamber for both eyes (BE) and 100-200 micra multiple white dots, more abundant in the macula’s temporal area in BE. The fluorescein angiography (FAG) showed multiple hyperfluorescent dots (figs. 1 and 2). Treatment began with 40 mg per day of IM methylprednisolone (Urbason® soluble 40 mg, Aventis Pharma S.A., Madrid), 10 mg/ml of topical atropine sulphate (1% Atropina® Alcon Cusí S.A., Barcelona) every 12 hours and 1 mg/ml topical dexamethasone (Maxide x® Alcon Cusí S.A., Barcelona) every 6 hours. After administering treatment for two weeks, VA reached 20/20 in BE. The requested systemic tests (basic blood testing, chest x-rays, luetic serology, angiotensin-converting enzyme and Mantoux) were negative.

Four years later, the patient had a new relapse, more marked in the RE, with loss of vision of 20/60 in the RE and 20/30 in the LE, yet showing no visual field alterations. The same systemic pattern seen in the previous episode emerged, with full visual recovery after one month of treatment, although some white dots persisted together with the upper temporal peripapillary pigment in the RE.

In September 2001, a new outbreak occurred, although slighter in nature. VA stood at 20/25 in the RE and 20/20 in the LE, while funduscopic examination revealed white-yellowish dots spread across the posterior pole and alteration of the macular retinal pigment epithelium (RPE) in BE. As in the first episode, treatment included topical and systemic corticoids. Three weeks after the onset of the new outbreak, VA decreased to 20/40 in the RE and 20/60 in the LE, which led to adding oral metotrexate (Metotre xato Lederle®, Whyeth, Madrid) at low doses. The requested HLA A29 study turned out negative.

In April 2005, the patient remained asymptomatic, her VA standing at 20/30 in the RE and 20/25 in the LE. The eye fundus revealed an elevation of the nasal juxtafoveal neuroepithelium in the RE; the optic coherence tomography confirmed the suspected diagnosis of SRNV (figs. 3 and 4). The patient rejected the treatment, although five months later, with VA at 20/60 in the RE, treatment began to address the SRNV (fig. 5) with photodynamic therapy (Visudy ne® Novartis Farmacéutica S.A., Barcelona) and 4 mg of intravitreous triamcinolone acetonide in .1 ml (Trigón depot® 40 mg/ml, Bristol-Myers Squibb, Madrid). VA in the RE improved to 20/40’.

Figs. 1 y 2: Fluorescein angiography in later times during the first bilateral MEWDS episode revealed multiple hyperfluorescent dots, more abundant in the temporal region, with a wreath-like pattern characteristic of MEWDS; the papilla shows as well the late hyperfluorescence with colorant leaking.

Fig. 3: Retinography of the RE revealing a bleeding subretinal neovascular membrane.
Currently, the patient’s VA is 20/40 in the RE and 20/25 in the LE, with faded white-yellowish dots in the anterior pole of both eyes, lesions related to a peripapillary atrophy in BE, involving the papillary-macular bean in the RE and several whitish, inactive lesions below the macula in the LE (fig. 6).

DISCUSSION

MEWDS is a rare ocular inflammatory disease of unknown etiology that tends to affect young women and is usually characterized by a self-limited progression, spontaneous recovery of visual acuity in 3 to 10 weeks and minimal sequelae in the RPE (1-5).

In the present case, three episodes were reported during 9 years, all of them bilateral. This clinical pattern of MEWDS presentation (bilateral and recurrent) is extremely unusual, not knowing the reason why some patients progress differently than others (2,3).

We found at least five cases described in the literature concerning SRNV in patients suffering from MEWDS (table I). In the present case, SRNV developed 13 years after the first MEWDS outbreak, with a final VA of 20/40.

During the progression of a recurrent MEWDS, the confluence of inflammatory areas and the persistence of inflammation may lead to chorioretinal scars (4). Our patient developed areas of peripapillary atrophy and several white-yellowish, atrophic lesions in areas of previous inflammatory activity. These alterations at the RPE level may predispose to the development of SRNV.

In the present case, the recurrent episodes of this disease where brought under control with low doses of metotrexate. Cyclosporine was also successfully used to achieve remission of outbreaks (3).

Our patient did not present vitritis, cystic macular edema, papillary edema, or the peripheral chorioretinal lesions that characterize multifocal choroiditis (MFC). Furthermore, she was emetropic, thus discarding a punctate inner choroidopathy. On the other hand, Kozielec et al (4) describe in patients suffering from recurrent and usually bilateral MEWDS focal lesions of chorioretinal atrophy.

Fig. 4: Optic coherence tomography of the RE, horizontal cut (black arrow in figure 3). Subretinal neovascular membrane 200 µm thick (white arrow).

Fig. 5: Retinography of the RE after treatment including photodynamic therapy. There is an atrophy area in the papillary-macular and peripapillary beam.

Fig. 6: Retinography of the LE revealing some faded white dots in the posterior pole. The black arrows point at two whitish lesions looking atrophic and inactive.
located near the papilla, in the posterior pole and mid-periphery resembling MFC lesions and presumed ocular histoplasmosis, although the scarring pattern is different. All of the above leads one to think of a probably atypical MEWDS. Bryan et al (5) describe a series of cases with previous MEWDS or after MFC where there is likely a susceptibility, etiology and/or pathogenesis common to these two clinical entities, even though this fact remains unknown.

Despite the good visual prognosis for patients suffering from MEWDS, we believe these patients should be assessed in the long term due to the risk of developing SRNV.

**REFERENCES**