Focal electroretinogram and visual field defect in multiple evanescent white dot syndrome

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Multiple evanescent white dot syndrome (MEWDS) is characterised by decreased visual acuity, white dots at the level of retinal pigment epithelium (RPE), scotoma, including enlargement of the blind spot, and visual return without treatment. Although electrophysiological data, such as reduced a-wave in electroretinogram (ERG) and reduced early receptor potential, indicate that the disease primarily affects the RPE and the outer segments of photoreceptors, optic nerve involvement has been reported to be a possible cause of the scotoma. On the other hand, several reports suggest that a disease known as 'acute idiopathic blind spot enlargement (AIBSE)', in which the enlargement is caused by retinal dysfunction, and MEWDS are the same clinical entities. We recorded focal electroretinographies in two patients with MEWDS to study the nature of the scotoma.

Case reports

CASE 1
A 32-year-old woman noticed a central scotoma in the left eye, and was examined 6 days after the onset. Corrected visual acuity was 1/2 right eye and 0-2 left eye. Fundus examination of the left eye revealed multiple white lesions deep in the retina and fluorescein angiography demonstrated...
Figure 1. The Octopus M1 program in case 1. (A) A normal left eye. (B) Right eye at initial visit. The threshold was remarkably elevated. (C) Left eye 1 month after initial visit. The threshold in the central portion was slightly elevated as compared with normal left eye. (D) Left eye 12 months after initial visit. The central threshold was still slightly elevated.

Figure 2. The full field ERG in case 1. (A) A normal left eye. (B) Right eye at initial visit. All components showed a slight reduction in the amplitude. (C) Right eye 1 month after initial visit. The ERG was normal except for a slightly reduced scotopic b-wave.

Figure 3. Focal macular ERG (spot size: 15') in case 1. (A) A normal left eye. (B) Right eye at initial visit. The focal ERG was deteriorated. (C) Left eye 1 month after initial visit. The amplitudes of a- and b-waves and oscillatory potentials were slightly reduced. (D) Right eye 12 months after initial visit. The amplitudes of all components were still slightly reduced.

patchy hyperfluorescence in the posterior pole. Psychophysical threshold was elevated in the central field of the left eye (Octopus Program M1) (Fig 1A (left eye), 1B (right eye)). While full field ERG, recorded using the standard technique reported previously, showed a slight reduction in amplitudes (Fig 2A (left eye), 2B (right eye)), focal macular ERG (spot size 15') was non-recordable in the left eye (Fig 3A (left eye), Fig 3B (right eye)). The details of our method for recording focal ERG have been described elsewhere. 

One month later, visual acuity of the left eye recovered to 1.2, and the psychophysical threshold in the macula was only slightly elevated (Fig 1C). Fundus abnormalities had disappeared, and full field ERG was normal except for a very slight reduction of the scotopic component (Fig 2C). However, the amplitudes of a- and b-waves and oscillatory potentials (OPs) in focal macular ERG remained reduced (Fig 3C).

Twelve months later the central threshold was still slightly elevated (Fig 1D), and the amplitudes of all components in focal macular ERG remained abnormal (Fig 3D).

CASE 2

A 24-year-old man reported a 5-day history of visual disturbance. Corrected visual acuity was 1.5 right eye and 0.4 left eye. Fundus examination of the left eye revealed multiple white dots deep in the retina and fluorescein angiography showed early hyperfluorescence in the macula and dye leakage from the disc. Goldmann perimetry revealed an enlarged blind spot (Fig 4A (right eye), 4B (left eye)) in the left eye. Focal ERG recorded beside the optic disc (spot size 10') had deteriorated (Fig 5A (right eye), 5B (left eye)), while the amplitudes of focal macular ERG (spot size 10') were slightly reduced (Fig 6A (right eye), 6B (left eye)).
Two months later, the corrected visual acuity in the left eye returned to 1.0, and the enlargement of the blind spot had minimised (Fig 4C). Fundus examination and fluorescein angiography revealed no abnormality. The amplitudes of all components in focal ERG beside the optic disc were slightly reduced (Fig 5C), whereas in the macula the amplitudes of a- and b-waves were normal but those of OPs were reduced (Fig 6C).

Fourteen months later, the enlargement of the blind spot had almost disappeared (Fig 4D). All components in focal ERG remained abnormal beside the disc (Fig 5D), while they returned completely to normal in the macula (Fig 6D).

Comment
Although Dodwell et al believed that optic nerve dysfunction produces scotoma in MEWDS, several authors suggested that AIBSE where multifocal study proved the retinal nature of scotoma is a subset of MEWDS. Sigh et al considered that NEWDS is a subset of AIBSE. The nature of scotoma in MEWDS needed to be studied before any conclusion can be drawn.

In case 1 with central scotoma, the amplitude was more reduced in macular ERG than in full field ERG, and its recovery was much slower in the former. In case 2 with blind spot enlargement, the amplitude of focal ERG was more reduced beside the optic disc than in the macula, and the recovery of the amplitude slower beside the optic disc. These data indicate that the scotoma was caused by retinal dysfunction in our patients. Furthermore, the results of case 2 support the hypothesis of clinical entities being similar between MEWDS and AIBSE. Finally, our ERG technique, totally different from the
multifocal ERG, recorded a- and b-waves and OPs, and demonstrated delayed recovery of OPs in case 2, which may imply inner retinal layer involvement in MEWDS. Similar findings have been reported in central serous chorioretinopathy.14


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Contralateral active ocular toxoplasmosis in Fuchs' heterochromic cyclitis

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Many authors have published papers on the assumed association between Fuchs' heterochromic cyclitis and ocular toxoplasmosis. Most studies reported on the presence of choroidal scars which were clinically consistent with a previous intraocular toxoplasmosis. In the majority of cases these toxoplasmosis-like lesions were present in the cyclitic eye. Only a few patients with Fuchs' heterochromic cyclitis and active Toxoplasma retinochoroiditis have been described. Until now, this clinical picture of toxoplasmosis was confirmed by aqueous humour analysis in two cases only. In a recent study we found a significantly higher incidence of toxoplasmosis-like choroidal scars in patients with Fuchs' heterochromic cyclitis than in a control group of patients with an HLA-B27-positive anterior uveitis. Although we found this positive clinical association between Fuchs' heterochromic cyclitis and toxoplasmosis-like scars, it could not be substantiated by serological tests for toxoplasmosis (immunofluorescence or ELISA) or by a test for cellular immunity to Toxoplasma antigen. Analysis of aqueous humour samples for Toxoplasma antibodies also yielded negative results. One must keep in mind, however, that no active choroidal lesions were present in the patients with Fuchs' heterochromic cyclitis at the time of blood sampling, nor at the time when the aqueous humour samples were obtained (during cataract surgery).

Here, we report on a patient with unilateral Fuchs' heterochromic cyclitis who developed an active toxoplasmosis of the contralateral eye, which could be proved by aqueous humour analysis.

Case report
A 28-year-old patient consulted our ophthalmology department with complaints of a diminished visual acuity of the left eye (6/6 on the right, hand movements (HM) on the left). Ophthalmic examination disclosed small white keratic precipitates scattered on the endothelium, 1+ flare in the aqueous, diffuse iris stromal atrophy, no posterior synechiae, and evident heterochromia. A dense subcapsular cataract was present. Tests to exclude other causes of uveitis were all within normal range. Based on the clinical picture our diagnosis was Fuchs' heterochromic cyclitis. The right eye showed no anterior segment abnormalities, but fundus examination disclosed small pigmented chorioroidal scars nasally. During the cataract extraction we obtained aqueous humour of the left eye after informed consent. No Toxoplasma antibodies could be detected, even in undiluted aqueous humour. The Toxoplasma antibody titre in a paired serum sample was 1:32. No fundus lesions were seen after removal of the cataract. Two years later, this patient returned with a diminished visual acuity of the right eye (6/12 on the right, 20/24 on the left). The left eye still showed the typical clinical features of Fuchs' heterochromic cyclitis, as described above. The right eye showed mutton fat keratic precipitates,