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

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 choriotinal 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 choriotinal 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 choriotinal 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 choriotinal 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,
Table 1 Results of investigation of intraocular production of antibodies against various micro-organisms

<table>
<thead>
<tr>
<th>Toxoplasma gondii</th>
<th>HSV CMV EBV VZV</th>
</tr>
</thead>
<tbody>
<tr>
<td>Serum titre</td>
<td>1:16</td>
</tr>
<tr>
<td>Antibody titre</td>
<td>1:4</td>
</tr>
<tr>
<td>Coefficient</td>
<td>22</td>
</tr>
<tr>
<td>Serum titre</td>
<td>&lt;1:16</td>
</tr>
<tr>
<td>Antibody titre</td>
<td>0:0</td>
</tr>
<tr>
<td>Coefficient</td>
<td>&lt;1:2</td>
</tr>
<tr>
<td>Serum titre</td>
<td>1:256</td>
</tr>
<tr>
<td>Antibody titre</td>
<td>1:2</td>
</tr>
<tr>
<td>Coefficient</td>
<td>1:4</td>
</tr>
</tbody>
</table>

HSV = herpes simplex virus, CMV = cytomegalovirus, EBV = Epstein-Barr virus, VZV = varicella zoster virus

1+ cells in the aqueous, and many vitreous opacities. No iris abnormalities were seen. The fundus was difficult to examine owing to many vitreous opacities; disc oedema, periphlebitis, and an active focal retinoc choroidal lesion could be discerned. A diagnostic anterior chamber paracentesis was performed. Using an immunofluorescence test, local intraocular antibody production against various micro-organisms was investigated, as described earlier. A positive Goldmann-Witmer coefficient of 22 (≥3 is considered positive) was found for Toxoplasma gondii, indicating an active intraocular production of Toxoplasma antibodies (Table 1). No intraocular production of antibodies against herpes simplex virus (HSV), cytomegalovirus (CMV), Epstein-Barr virus (EBV), or varicella zoster virus (VZV) could be detected. Based on the results of this aqueous humour analysis, a diagnosis of active ocular toxoplasmosis was made and therapy with clindamycin and sulphadiazine was started. Within several weeks the vitreous cleared; a retinoc choroidal lesion typical of toxoplasmosis appeared in satellite formation next to old pigmented scars (Figure 1).

Comment

Until now, only one study reported two patients with Fuchs' heterochromic cyclitis and active Toxoplasma retinoc choriditis, in whom the clinical diagnosis of toxoplasmosis was confirmed by aqueous humour analysis. We recently described a patient with a definite congenital bilateral toxoplasmosis who developed a unilateral Fuchs' heterochromic cyclitis. The patient described in the current report had no clinical characteristics of toxoplasmosis in his eye with Fuchs' heterochromic cyclitis, but developed an active toxoplasmosis in the opposite eye, which could be proved by aqueous humour analysis. These few cases support the hypothesis that Fuchs' heterochromic cyclitis may be secondary to congenital toxoplasmosis.

Fuchs' heterochromic cyclitis, although considered as a separate nosological entity, has also been described in association with other (ocular) diseases; retinitis pigmentosa, ocular trauma, the subclavian steal syndrome, hemifacial atrophy, and Horner's syndrome. In a recent study on the association between Fuchs' heterochromic cyclitis and toxoplasmosis several patients lacked the keratic precipitates typical of Fuchs' heterochromic cyclitis. On the other hand, patients with ocular toxoplasmosis sometimes have keratic precipitates characteristic of Fuchs' heterochromic cyclitis. Occasionally also pigmented keratic precipitates are observed in patients with a typical clinical picture of Fuchs' heterochromic cyclitis. It is often difficult to make the diagnosis Fuchs' heterochromic cyclitis, since no minimal diagnostic criteria have been established internationally: in fact, a spectrum of signs may be seen in patients with this disorder. It is therefore conceivable that Fuchs' heterochromic cyclitis may be a secondary phenomenon with a spectrum of clinical features and different causes, including congenital toxoplasmosis.

Figure 1 Fundus photograph of the right eye 3 weeks after the therapy was started. A fresh retinoc choroidal lesion typical of toxoplasmosis appeared in satellite formation next to old pigmented scars.