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*Case Report*

# THE ASSOCIATION OF FUCHS' UVEITIS SYNDROME WITH BEHÇET'S DISEASE

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***Abstract***

*The characteristic clinical features of Fuchs' uveitis syndrome are: diffusely distributed stellate keratic precipitates, atrophy of the anterior layer of iris, especially in the peripupillar region (heterochromia), the absence of posterior synechiae, development of posterior subcapsular cataract, Amsler sign. In relevant publications the syndrome was described in association with ocular toxoplasmosis, sarcoidosis, retinitis pigmentosa. The cases of recurrent subconjunctival hemorrhages and iris crystals (Russell bodies, i.e., immunoglobulin accumulation within plasma cells) in patients with Fuchs' uveitis syndrome were also reported.*

*Among 44 patients with Fuchs' uveitis syndrome examined at Uveitis Department of the Ophthalmological Center after S.V. Malayan, one patient had atypical clinical presentation, which was associated with Behçet's disease. This association, to our knowledge, has never been described.*

*In cases of association with ocular toxoplasmosis, according to H. Saraux and associates (1985), Fuchs' uveitis syndrome represents an autoimmune reaction targeted towards the retina affected by Toxoplasma gondii. We assume that the reason of Fuchs' uveitis syndrome association with Behçet's disease relies on a certain type of immune responses. Further investigations, including genetic studies, are needed to explain this association in view of the high prevalence of Fuchs' uveitis syndrome in Armenians.*

***Keywords:*** *Fuchs' uveitis syndrome, heterochromia, synechia, cataract, Behçet's disease.*

In 1906 Ernest Fuchs, from the University of Vienna, produced a comprehensive summary of 38 patients with features of the condition that now bears his name. In several reported series the prevalence of Fuchs' uveitis syndrome varied from 1.2% to 4.5% of patients with uveitis [*Bloch-Michel E., 1981; Chung Y. et al., 1988*]. The disease has no racial or sexual predilection [*Liesegang T., 1982*].

As a rule, Fuchs' uveitis syndrome is a unilateral disease, though about 7.8% to 10% of patients have bilateral impairment [*Jones N., 1991*]. The characteristic clinical features of the disease are: diffusely distributed stellate keratic precipitates, atrophy of the anterior layer of iris, especially in the peripupillar region (heterochromia), absence of posterior *synechiae*, development of posterior subcapsular cataract, Amsler sign (filiform hemorrhage in the anterior chamber angle after paracentesis or in association with cataract surgery, glaucoma surgery, gonioscopy, applanation tonometry) [*Jones N., 1991*]. Glaucoma has been described in up to 59% of patients and is the most common cause of permanent vision loss in patients with occluded vessels [*Liesegang T., 1982*].

Small gelatinous nodules on the iris surface (*Busacca nodulae*) or at the pupillary margin (*Koeppe nodulae*) are observed in a variable number of patients with Fuchs' uveitis syndrome.

Among 44 patients with Fuchs' uveitis syndrome examined at Uveitis Department of the Ophthalmological Center after S.V. Malayan, one patient had atypical clinical presentation: association with Behçet's disease. To our knowledge, this association was not described in available publications. Behçet's disease is a multisystem inflammatory illness

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characterized by intraocular inflammation (ocular Behçet's disease), oral and genital ulcerations, skin lesions: erythema nodosum-like lesions, acneiform lesions or folliculitis, thrombophlebitis, cutaneous hypersensitivity and variety of other disorders involving multiple organ systems in the body, such as epididymis, joints, intenstine, vascular system (vasculo-Behçet's disease) and the central nervous system (neuro-Behçet's diease). The disease is commonly found in Mediterranean countries. Amazingly, Fuchs' uveitis syndrome is also common in Armenians; it accounts for about 12% among all uveitis cases in Armenia.

***Report of a Case***

A 40-year-old man, M.H., was accepted to the Uveitis Department of the Ophthalmological Center after S.V. Malayan with complaints of low excentric vision in his right eye. The patient presented with clinical features, which were characteristic for Behçet's disease: oral and genital ulceration, cutaneous hypersensitivity, left knee joint pain and hypopion. The complaints were present for 3 years. The patient was prescribed 150 *mg/day* Imuran ("Moskhimfarm", Russia).

At objective examination the following was recorded.

*OD*: At slit lamp examination the patient had moderately expressed inflammatory reaction (2+ cell) in the anterior chamber aqueous humor of the right eye. Fundus examination revealed that the optic nerve head was pale with distinct borders. There was atrophic old scar in macular region, occluded vessels in midperiphery (Figures 1-2). The patient had history of cataract extraction and intraocular lens implantation in the past.

*OS*: The left eye had diffusely distributed stellate keratic precipitates, rare cells (mild inflammatory reaction) in the anterior chamber aqueous humor, peripupillary iris atrophy (Figure 3) and mild posterior subcapsular cataract. Moderate inflammatory reaction (2+ cell) was observed in the vitreous body. Fundus examination revealed cystoid macular edema. There were no signs of vasculitis at the first presentation.

Hence, the mentioned patient was diagnosed with ocular Behçet's disease in one eye and – Fuchs-like picture present in the other eye – with Fuchs' uveitis syndrome as well. The patient was diagnosed with Behçet's disease according to systemic findings. It is possible that patient will later develop signs of ocular Behçet's disease in the left eye as well (hypopyon, occlusive vasculitis).

Fuchs' uveitis syndrome may have atypical clinical presentation. It has been described in association with ocular toxoplasmosis (toxoplasmic chorioretinitis), sarcoidosis (sarcoid chorioretinitis), and *reti-*

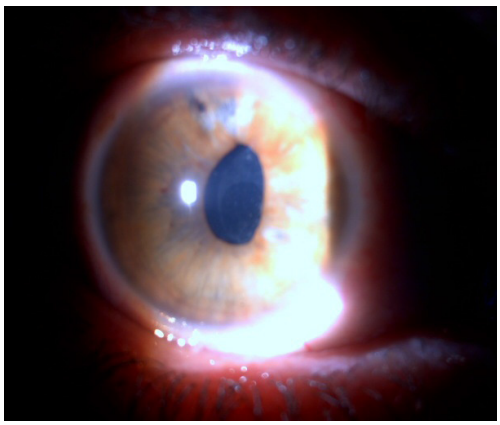

**Figure 1.** *Patient M.H. Photograph of the right eye. Behçet's disease.*

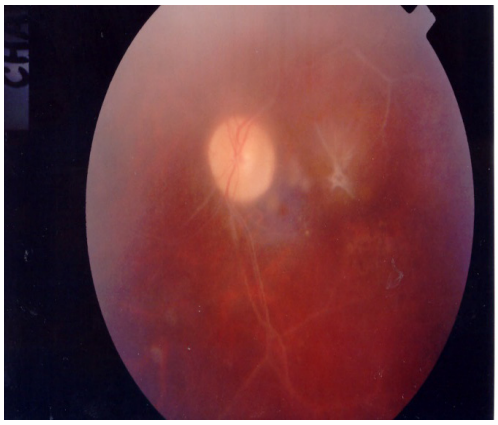

**Figure 2.** *Patient M.H. Photograph of the right eye. Occluded vessels, macular scar, pale optic nerve.*

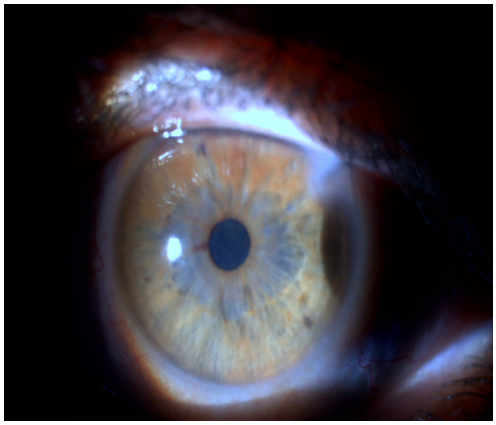

**Figure 3.** *Patient M.H. Photograph of the left eye. Peripupillary iris atrophy.*

*nitis pigmentosa* [*Schwab I., 1991; Goble R., Murray P., 1995; Chowers I. et al., 2000*]. The cases of recurrent subconjunctival hemorrhages and iris crystals (Russell bodies, i.e., immunoglobulin accumulation within plasma cells) were also reported in patients with Fuchs' uveitis syndrome [*Noda S., Hayasaka S., 1995; Cohen E., 1998*]. The significant point of the described clinical case was association of Fuchs' uveitis syndrome with ocular Behçet's disease. It should be emphasized that such an association of 2 pathologies was not described in the available publications.

In cases of association with ocular toxoplasmosis H. Saraux and associates have suggested that *Toxoplazma gondii* infection of the retina permits sensitization to a retinal antigen that may elicit Fuchs' uveitis syndrome [*Saraux H. et al., 1985*]. Recently a longitudinal study of patients with Fuchs' heterochromic uveitis was carried out. A sample of 54 patients with monocular and three with binocular Fuchs' heterochromic uveitis was followed for periods ranging from 8 months to 15 years. Chorioretinal scars were found in 6 patients, *Toxoplasma* serology was positive in 18(35%) patients, the authors concluded that connection with toxoplasmosis in individual cases cannot be excluded [*Norrsel K., Sjodell L., 2007*]. Within another study the local production of antibodies to *Rubella* were found in the anterior chamber aqueous humor of all patients with Fuchs' uveitis syndrome [*Mohamed Q., Zamir E., 2000*]. There is no explanation regarding the association of Fuchs' uveitis syndrome with sarcoidosis and *retinitis pigmentosa* [*Goble R., Murray P., 1995; Chowers I. et al., 2000*]. The etiology of Fuchs' uveitis syndrome is unclear and widely debated, and, probably, it is merely a secondary phenomenon or a clinical state of a number of possible infective and/or immunologic triggers [*Goble R., Murray P., 1995*]. We assume that the reason of such association with Behçet's disease relies on some type of immune responses. Further investigations are needed to explain this association, including the genetic studies, to explain such a high prevalence of Fuchs' uveitis syndrome in Armenia.

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