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Great imitator — ocular syphilis

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ABSTRACT

We examined a 44-year-old woman without any known systemic disease with a decreased vision of a month duration in her left eye. She had undergone left uneventful cataract surgery with intraocular lens implantation elsewhere 14 years ago. On examination, her Snellen visual acuity was 1.0 (with the correction of –0.50–0.25 × 120) in the right eye and 0.4 (with the correction of +0.75–2.25 × 115) in the left. There was left hypochromic heterochromia. While the right anterior segment was unremarkable, there were small to medium-sized keratic precipitates, 3+ anterior chamber cells, a posterior chamber intraocular lens with an intact posterior capsule, and a few vitreous cells in the left eye. Fundus autofluorescence imaging revealed peripapillary hyperautofluorescence in the left eye, and fluorescein angiography revealed a marked optic nerve head and perivascular leakage at the posterior pole. The findings were not compatible with Fuchs uveitis, so the full infectious panel was worked out. Serologic investigations yielded the presence of syphilis, and the patient was treated successfully with systemic antibiotics. Our case demonstrates the mimicking nature of ocular syphilis and the importance of high clinical suspicion when reaching the correct diagnosis.

KEY WORDS: Fuchs' heterochromic iridocyclitis; Fuchs uveitis; Fuchs uveitis syndrome; ocular syphilis

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INTRODUCTION

Fuchs uveitis, also known as Fuchs uveitis syndrome or Fuchs' heterochromic iridocyclitis, was first described in 1906 [1]. Fuchs uveitis is generally a unilateral, chronic recurrent non-granulomatous uveitis that accounts for 2-11% of all uveitis cases [2]. Diffusely distributed stellate keratic precipitates, low-grade intraocular inflammation, iris heterochromia and/or atrophy, absence of posterior synechiae, and development of early cataract formation and glaucoma are among the characteristic features of the disease [1].

Various clinical entities such as toxoplasmosis [3], toxocariasis [4], rubella vaccination [5], cytomegalovirus [6], herpes simplex virus [7], Chikungunya virus infections [8], retinitis pigmentosa [9], sarcoidosis [10], Usher's syndrome [11] and Horner's syndrome [12] have been previously reported to have an association with Fuchs uveitis.

We present a case with Fuchs uveitis, who also developed unilateral panuveitis as a consequence of ocular syphilis.

CASE PRESENTATION

We examined a 44-year-old otherwise healthy woman with a left visual deterioration lasting a month. Her past and family histories were unremarkable. However, she had heterochromia since early childhood and underwent uneventful left cataract surgery with intraocular lens implantation elsewhere 14 years ago. There was left hypochromic heterochromia (Fig. 1). Pupillary light reflexes were normal, with no relative afferent pupillary defect.

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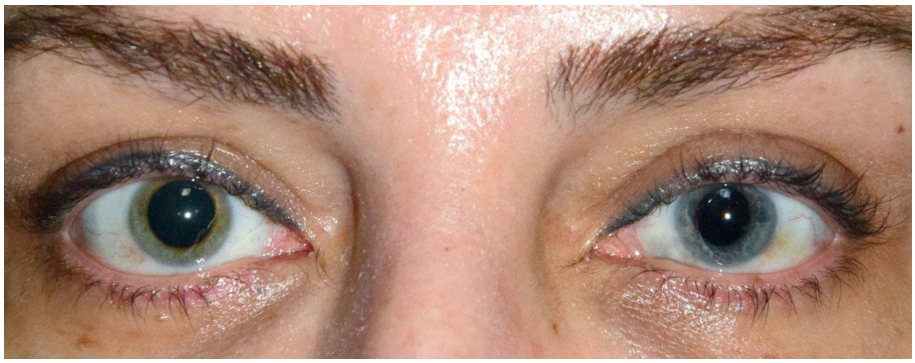

Figure 1. At admission. A facial picture depicting only the eyes revealed a left hypochromic heterochromia

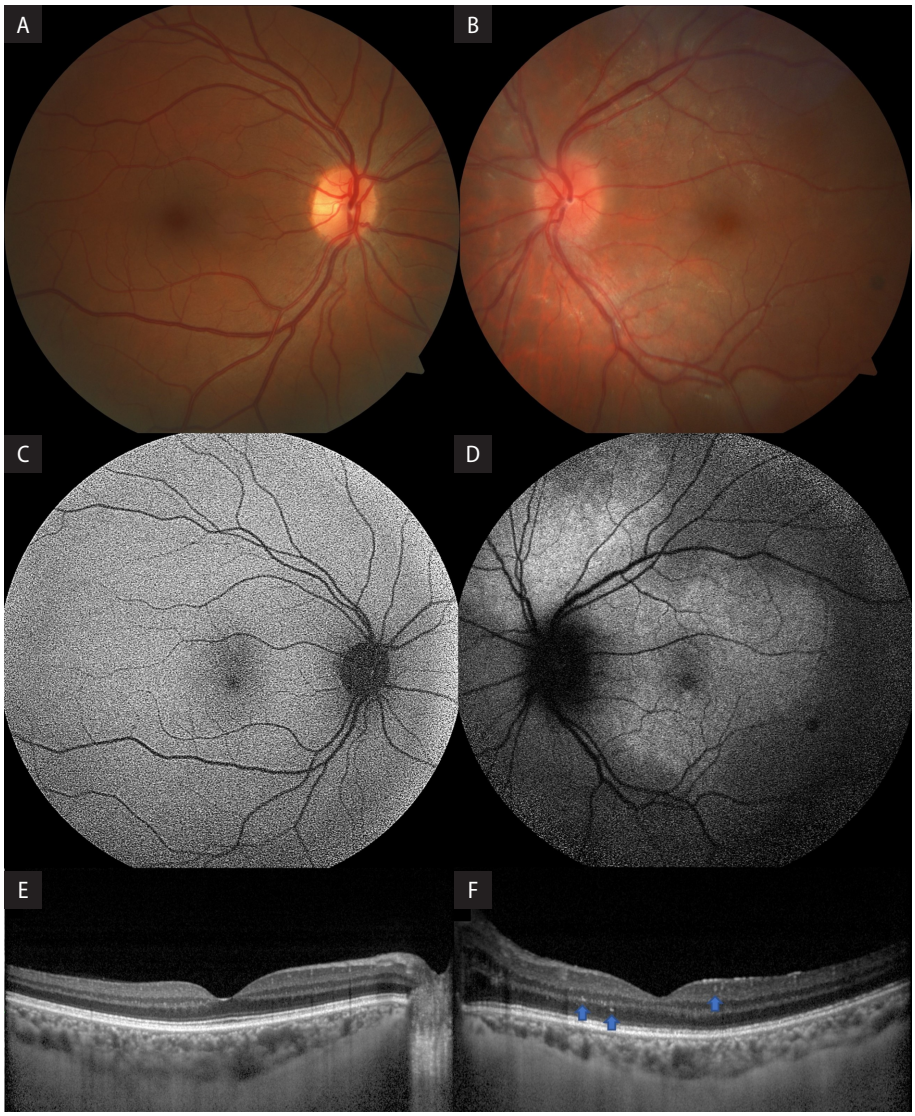

Figure 2. At admission. **AB.** Color fundus images: right eye (**A**) and left eye (**B**). While the right fundus was normal, there was a hyperemic optic disc with blurry margins and a normal-looking macula in the left eye; **CD.** Fundus autofluorescence images: right eye (**C**) and left eye (**D**): right autofluorescence picture was unremarkable, and there was marked peripapillary and macular hyperautofluorescence in the left eye. **EF.** Enhanced depth imaging optical coherence tomographic sections: right eye (**E**) and left eye (**F**): foveal contour was normal in the right eye; there were multiple intraretinal hyperreflective dots (blue arrows) in the left eye

Her best-corrected Snellen visual acuity was 1.0 with the correction of –0.50–0.25 × 120 in the right eye and 0.4 with the correction of +0.75–2.25 × 115 in the left eye. Color vision was 21/21 bilaterally with the Ishihara pseudoisochromatic plates. While slit-lamp examination of the right eye was unremarkable, there was a posterior intraocular lens with an intact posterior capsule, small to medium-sized scattered keratic precipitates, 3+ anterior chamber cells, and occasional vitreal cells in the left eye. Intraocular pressure was 16 mm Hg bilaterally with Goldmann applanation tonometry. While the right fundus was normal (Fig. 2A), the left optic disc margins were somewhat blurry and hyperemic with a normal-looking macula (Fig. 2B). Right fundus autofluorescence (Heidelberg Spectralis, Heidelberg Engineering, Heidelberg, Germany) was normal (Fig. 2C), but there was marked peripapillary and macular hyperautofluorescence in the left eye (Fig. 2D). Enhanced depth imaging optical coherence tomographic (Heidelberg Spectralis, Heidelberg Engineering, Heidelberg, Germany) sections depicted normal foveal contour in the right eye (Fig. 2E). Still, some intraretinal hyperreflective dots were in the left eye (Fig. 2F). Fluorescein angiogram (Heidelberg Spectralis, Heidelberg Engineering, Heidelberg, Germany) showed no abnormality in the right eye, but there was disc hyperfluorescence and slight hyperfluorescent perifoveal leakage in the left eye (Fig. 3A–D). Optical coherence tomography angiography (Triton, Topcon Inc., Oakland, New Jersey, United States) findings were almost unremarkable in both eyes.

We put the patient on prednisolone acetate drops every two hours and cyclopentolate drop t.i.d. and performed a complete systemic workup as the clinical presentation was not entirely compatible with Fuchs uveitis. Cytomegalovirus and rubella virus immunoglobulin G tests were positive. To our surprise, non-specific anti-treponemal tests of the venereal disease research laboratory and rapid plasma reagin were positive. The tests of other sexually transmissible diseases, such as human immunodeficiency virus (HIV) and hepatitis, were negative.

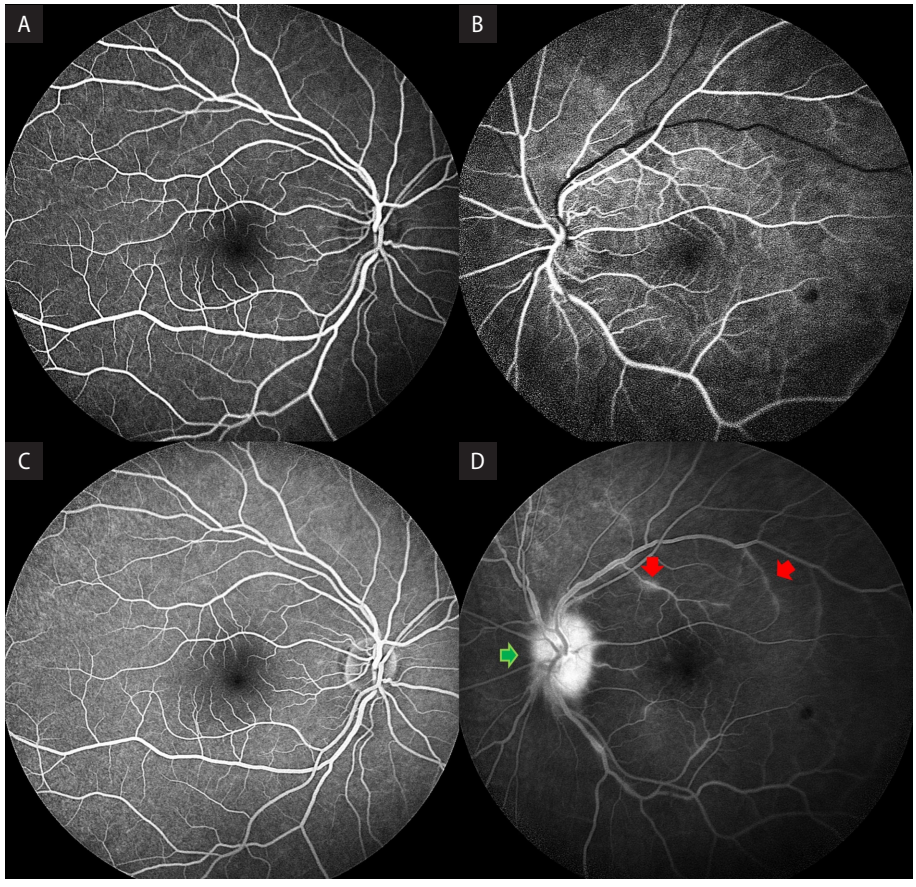

Figure 3. At admission. Early and late phases of fluorescein angiogram of right eye (**A**, **C**) and left eye (**B**, **D**): while angiographic findings were unremarkable in the right eye disc staining (green arrow) and mild perifoveal leakage (red arrows) were visualized in the left eye

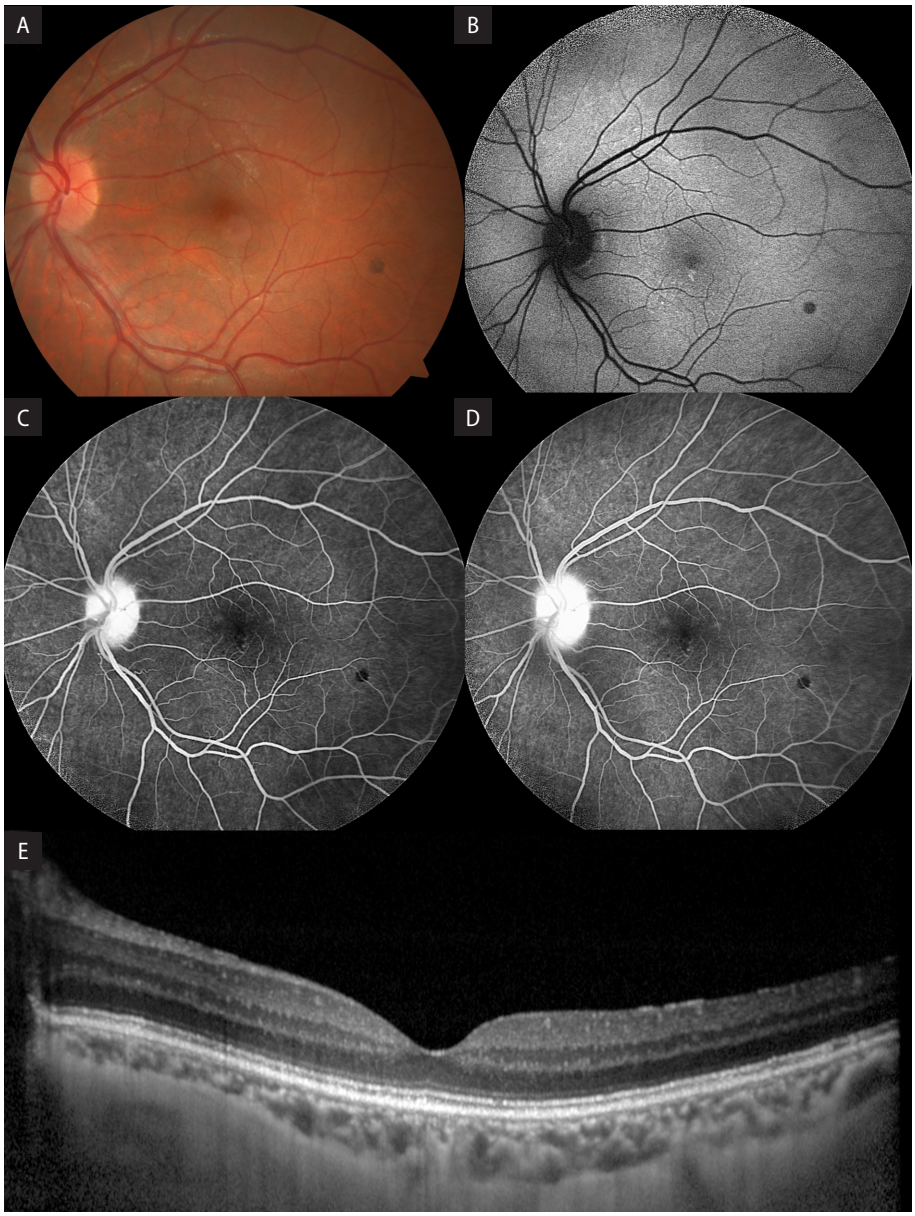

Figure 4. Ten weeks after the diagnosis. Left eye. **A.** Color fundus image: almost normal-looking optic disc and macula; **B.** Normal-looking autofluorescent image; **CD.** Fluorescein angiographic pictures revealed only slight disc staining and no macular capillary leakage. **E.** On enhanced depth imaging, optical coherence tomography, normal foveal contour with very few hyperreflective dots.

The results of cranial and orbital magnetic resonance imaging were normal. Upon neurology and infectious disease consultations, a regimen of a 20-day course of daily intramuscular 1 g ceftriaxone was prescribed as the patient denied intravenous treatment.

Ten weeks after the diagnosis, left best-corrected Snellen visual acuity improved to 0.7. The anterior chamber was clear, and left optic disc hyperemia diminished markedly (Fig. 4A). On autofluorescence imaging, left peripapillary and macular hyperautofluorescence was not detectable anymore (Fig. 4B). On fluorescein angiography, disc staining looked less, and perifoveal leakage vanished (Fig. 4C–D). On enhanced depth imaging optical coherence tomography, the foveal contour was normal except for a few hyperreflective dots (Fig. 4E).

DISCUSSION

The incidence of Fuchs uveitis among Turkish uveitic patients has been reported to be 6.3% [13]. Fuchs uveitis is characterized by diffusely scattered medium and/or stellate keratic precipitates,

and diffuse iris stromal atrophy, often with hypochromia and absence of posterior synechia before cataract surgery [14].

Bouchenaki and Herbort [15] retrospectively reviewed the data of 105 patients with Fuchs uveitis and found that the most frequent clinical sign was vitreous infiltration in 97.4 % of the eyes. The degree of vitreous involvement ranged from slight with a few cells (46.15%) to moderate (34.1%) and severe (14.5%) with dense vitreous strands and condensations. A fluorescein angiogram was performed in 39 patients. Disc hyperfluorescence was present in all but one patient (97.7%), and was not correlated with the degree of vitreous involvement.

Tugal-Tutkun et al. [16] reviewed the clinical charts of 181 eyes of 172 patients with Fuchs uveitis. There were vitreous cells and debris in 130 eyes (71.8%), and vitreous condensation was present in 88 eyes (48.6%). Fourteen eyes (7.7%) had chorioretinal scars.

Zarei et al. [17] evaluated the optic nerve head involvement in 43 eyes of Fuchs uveitis without clinical optic disc edema and obtained a fluorescein angiogram. 71% of those eyes showed optic nerve head hyperfluorescence.

Özdamar et al. [13] studied the 281 eyes of 258 patients with Fuchs uveitis. There was vitreous infiltration of varying grades in all eyes. Vitreous infiltration was found at the lowest grade, 0.5+ in 53 eyes (18.9%), 1+ in 96 eyes (34.2%), 2+ in 69 eyes (24.5%), and 3+ in 63 eyes (22.4%). Fundoscopy yielded the presence of chorioretinal scars in 28 eyes (10%) and mid-peripheral sectorial vascular sheathing in 25 eyes (8.8%). A fluorescein angiogram was performed in 40 eyes (14.2%). Nineteen of those eyes (47.5%) had optic disc hyperfluorescence; mid-peripheral vascular leakage was present in 13 eyes (32.5%). The authors concluded that posterior segment involvement was a feature of Fuchs uveitis that should not be ignored.

Ocular syphilis may present with a variety of manifestations. Ocular manifestations may involve the anterior segment of the eye, lens, uveal tract, retina, retinal vasculature, optic nerve, cranial nerves, and pupillomotor pathways. The most common ocular manifestation of syphilis is uveitis which may be granulomatous or non-granulomatous [18, 19]. Anterior segment involvement consists of keratitis, iris nodules, iridocyclitis, episcleritis, scleritis, and conjunctivitis. Posterior segment involvement in ocular syphilis is more common. Panuveitis, posterior uveitis, chorioretinitis, necrotizing retinitis, retinal vasculitis, central retinal artery/vein occlusion, vitritis, exudative retinal detachment, and neuroretinitis are among the clinical presentations of syphilis [19, 20]. Optic nerve involvement is rare and has been reported in approximately 20% of ocular syphilis cases. Optic nerve involvement at syphilis may be unilateral or bilateral and may present as papillitis, perineuritis, anterior optic neuritis, retrobulbar neuritis, papilloedema, gumma of the optic disc, and optic atrophy [19].

The treatment of ocular syphilis is the same as neurosyphilis, and the recommendation is intravenous crystalline penicillin G 18–24 mIU per day for 10–14 days. If the patient has a penicillin allergy, ceftriaxone 2 g daily, either intramuscularly or intravenously, for 10-14 days can be used [20].

A high index of suspicion enabled us to reach the correct diagnosis of syphilis despite the initial impression of Fuchs uveitis. Syphilis is 'the great imitator' and may mimic different ocular disorders. Thus, a delay in diagnosis may lead to unwanted clinical consequences.

CONCLUSIONS

Ocular syphilis clinically may present with a multitude of inflammatory appearances, and even in a patient with Fuchs-like unilateral uveitis, syphilis should be in the differential diagnosis as syphilis is the great mimicker.

Conflict of interests

The authors declare no potential conflicts of interest concerning this article's research, authorship, and/or publication.

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