Fuchs' Uveitis

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Introduction

Fuchs' uveitis syndrome (FUS) or Fuchs' heterochromic iridocyclitis or Fuchs' uveitis is a chronic nongranulomatous uveitis which was described by Ernst Fuchs (1851–1930), an Austrian Ophthalmologist. He also described Fuchs' endothelial dystrophy and Fuchs' spot in myopia. The patients are usually young to middle aged and present with complaints of floaters or visual decline. Some patients may be detected incidentally also. Severe pain and redness are typically absent. Usually, one eye is involved, though bilateral involvement has been reported in approximately 10% cases. There is usually no or minimal circumcorneal congestion. The cornea shows stellate diffuse keratic precipitates (KPs) extending over the whole cornea (including superior cornea) in comparison to the other uveitides where a triangular area of the inferior cornea (Arlt's triangle) is usually involved. The KPs are small, white, nonpigmented, and connected with each other by fine fibrils. Only minimal anterior chamber reaction/flare is seen and posterior synechiae are absent, though synechiae may form after anterior segment surgery. There is a loss of definition of the surface of the iris (washed out/moth eaten appearance). The sphincter around the pupil and normal radial iris vessels may be visible due to loss of iris tissue. Surface rugae and crypts are blunted. Collarette may be lost. Stromal atrophy may lead to transillumination defects, though sectoral iris atrophy typical of herpetic uveitis is not seen. The pupillary ruff may have gaps in some areas. The atrophy of sphincter and dilator pupillae may lead to anisocoria. FUS

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usually causes hypochromic iris, rarely hyperchromic iris can be noted in severe stromal atrophy and visibility of pigmented posterior epithelium of iris. The heterochromia is usually not very prominent in dark eyes, but slit lamp biomicroscopy usually picks up the subtle difference between the eyes. There may be nodules at the pupillary region of iris (Koeppe's nodules) or rarely in the stroma of iris (Busacca nodules). Shiny dot-like lesions may be seen over the iris and represent Russel bodies (plasma cells filled with immunoglobulins). Visual loss is usually due to cataract, glaucoma, or rarely extensive vitreous debris. Unilateral cataract is usually seen which starts as central posterior subcapsular cataract and may progress to total cataract. Gonioscopy may show fine radial vessels which may bleed during limbal incisions of cataract surgery resulting in hyphema (Amsler-Verrey sign).

The retrolental space may show cells. There may be vitreous opacities in the anterior vitreous cavity. However, cystoid macular edema is typically not seen in contrast to intermediate uveitis. Peripheral chorioretinal scars have been reported in up to 65% patient with FUS, leading to the proposal of association with toxoplasmosis or histoplasmosis. Also, rubella and cytomegalovirus have been implicated in the etiopathogenesis of FUS. Glaucoma is associated in up to 59% of cases and may cause severe vision loss. Usually, open angle glaucoma is seen. Other causes of glaucoma in FUS include corticosteroid-induced glaucoma, peripheral anterior synechia, angle neovascularization, trabeculitis, and phacolytic glaucoma. In comparison to Posner Schlossmann syndrome, the intraocular pressure does not usually respond to topical steroids. The anterior chamber reaction is usually very mild and does not need topical steroids. Steroids also have the risk of causing steroid induced glaucoma. FUS is not associated with posterior synechiae and anterior chamber inflammation is minimal, so it does not need treatment with cycloplegic agents also. Glaucoma may be difficult to treat and may not get controlled with topical or oral antiglaucoma medications. Filtration surgeries with antimetabolites (mitomycin-c or 5-fluorouracil) and glaucoma drainage devices show promise in such cases. The cataract surgery is usually uneventful as the pupil dilates fully and the cataract is usually soft. However, intraoperative hyphema and postoperative posterior synechia can occur. Preoperative and postoperative topical steroids are essential to avoid/control inflammation. Extensive vitreous membranes or debris causing visual loss may need to be removed with pars plana vitrectomy.

Case 1: A Young Female with Unilateral Visual Decline and Floaters

A 28-year-old Indian female presented with chief complaints of painless progressive diminution of vision in the right eye for 8 months associated with floaters for 2 months in the same eye. The dimness of vision was insidious in onset and increased in severity for the past 2 months. There was no redness, ocular pain, or photophobia. The patient did not give any history of ocular disease and trauma/ surgery. She was asymptomatic in the fellow eye. She was being treated outside with cyclopentolate and betamethasone eye drops for the past 4 weeks without resolution of symptoms and was referred to us for further management. There was no history of lower back pain, joint pain, tuberculosis, and oral or genital ulcers. Investigations done elsewhere showed ESR of 18 mm/hr. Chest X-ray was unremarkable. Mantoux test showed an induration of 8 × 8 mm. The patient had a best corrected visual acuity (BCVA) of 6/9 in the right eye and 6/6 in the left eye. Intraocular pressure was 18 mm Hg in the right eye and 16 mm Hg in the left eye by applanation tonometry. Pupils were brisk bilaterally, and there was no relative afferent papillary defect (RAPD) in the right eye. The right eye showed discrete small to medium size, white, nongranulomatous keratic precipitates (KPs) over the corneal endothelium (Fig. 1). The KPs were mostly distributed in the inferior half of the cornea (Arlt's triangle), though KPs were noted over the superior corneal endothelium also. The KPs were mostly round; however, some KPs had a stellate appearance. There were 1+ cells and 1+ flare in the right eye. The iris was uniformly depigmented

(Fig. 2a, hypochromic heterochromia) compared to the fellow eye (Fig. 2b). There was no obvious loss of crypts and no abnormal vessels or neovascularization was noted. Multiple (15–20), medium size, discrete, elevated, creamy iris nodules, located mostly around the pupillary ruff and peripupillary zone were noted (Fig. 3). Posterior synechiae were absent and the pupil dilated to 8 mm. Posterior subcapsular cataract was noted. There was no pigment on the anterior or posterior lens surface. Vitreous cells were 1+ and vitreous did not reveal any vitreous membranes. Fundus examination was unremarkable and no chorioretinal scar, snowball, snow banking, or cystoid macular edema was noted. A diagnosis of Fuchs' uveitis syndrome was made. Intermediate uveitis was excluded by the absence of snowballs and snowbanks, absence of cystoid macular edema, and absence of peripheral retinal vascular involvement. Posner Schlossmann syndrome was ruled out as the intraocular pressure was normal, there was no history of ocular redness and pain, there was no corneal edema, and too many KPs were present. Considering the typical presentation of FUS, no further investigations were ordered. The patient was counseled about the need for cataract surgery when routine activities become difficult. Topical steroid and cycloplegic were stopped. Also, she was informed of the possibility of development of glaucoma and the need of regular follow-up.

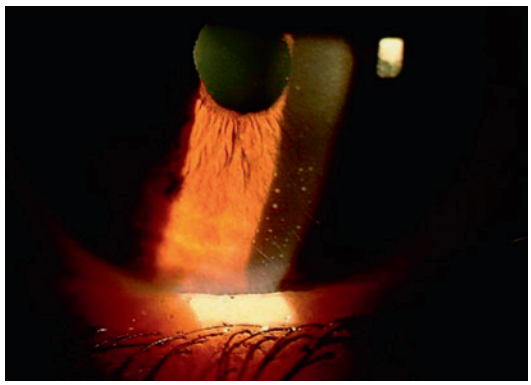

Fig. 1 Slit lamp photograph of the right eye of case 1 showed small white keratic precipitates

Case 2: A Young Female with Cataract in the Right Eye and Pseudophakia in the Left Eye

A 30-year-old Indian female presented with complaints of decreased vision in the right eye for 1 year associated with floaters noted for 4 months. The visual loss was insidious onset, increasing in severity for the past 4 months. There was no

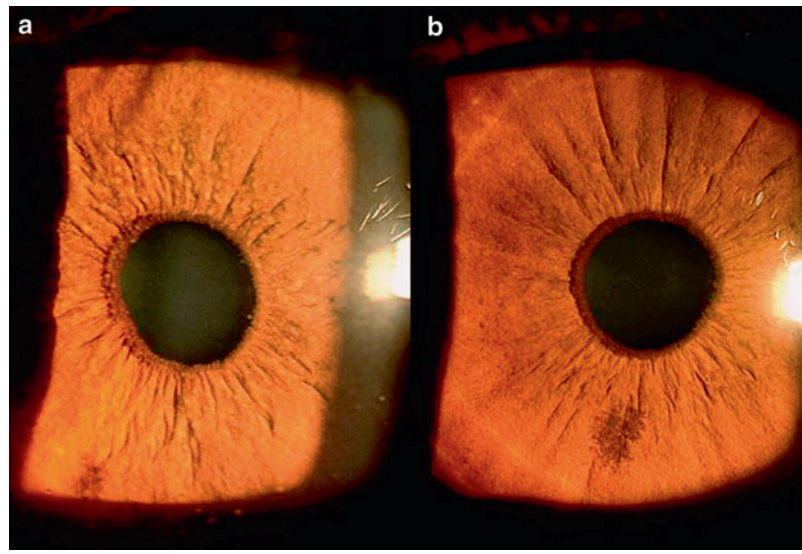

Fig. 2 Comparison between the right (**a**) and the left iris (**b**) of case 1 revealed subtle hypochromia in the right eye

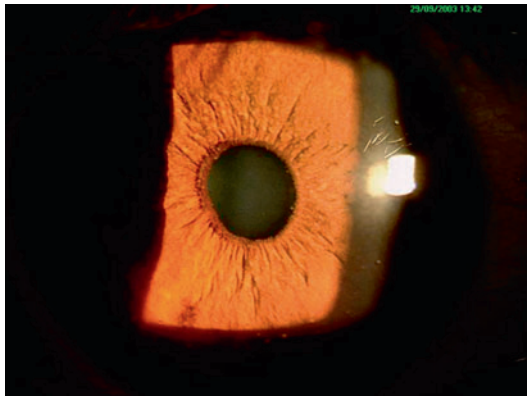

Fig. 3 The right eye of case 1 demonstrated small iris nodules near the pupil

history of trauma, redness, or pain. The left eye had similar complaints for which she underwent cataract surgery and intraocular lens implantation 4 years back with good postoperative visual gain. There was no history of topical or oral medication, joint pain, or tuberculosis. The BCVA was 2/60 in the right eye and 6/9 in the left eye. The intraocular pressure was 16 mm of Hg in either eye. Both pupils were reacting briskly to light, and there was no RAPD in the right eye. Anterior segment of the right eye showed small white KPs present diffusely over the endothelium (Fig. 4) including in the superior cornea. Posterior subcapsular cataract was present and there was no posterior synechia. Diffuse stromal atrophy of iris was noted with some loss of iris collarette (Fig. 5a). Vitreous showed cells. There were occasional anterior chamber cells and no flare was identified. The left eye showed minute fine diffuse KPs, mild diffuse iris stromal atrophy, posterior chamber intraocular lens, and no synechiae (Fig. 5b). Vitreous cells were also noted in the left eye. Fundi of both eyes were unremarkable with the absence of snow balls or snow banks, chorioretinal scars, and cystoid macular edema. Right eye showed some vitreous opacity. A differential diagnosis of intermediate uveitis was excluded by the absence of snowballs, snowbanks, peripheral vasculitis, and cystoid macular edema. Chronic anterior uveitis was ruled out by the absence of synechiae, absence of granulomatous keratic precipitates, and absence of any episode of redness, pain, or photophobia. Bilateral

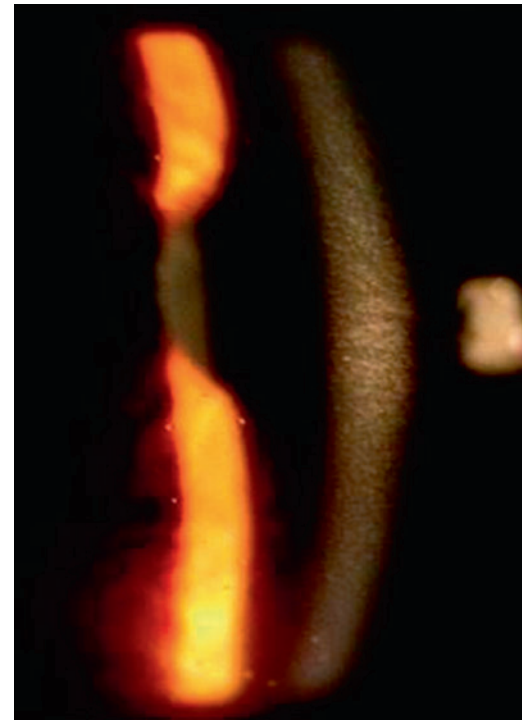

Fig. 4 Slit lamp photo of the right eye of case 2 showed keratic precipitates involving both the superior and inferior cornea

presentation is not usually seen in FUS but can occur in 10% patients. Heterochromia may not be seen in bilateral cases, and in pigmented iris detection of heterochromia in unilateral cases may be difficult. Thus, a clinical diagnosis of bilateral Fuchs' uveitis syndrome with right eye posterior subcapsular cataract and left eye pseudophakia was made. The patient underwent uneventful surgery (phacoemulsification with hydrophobic posterior chamber intraocular lens implantation) in the right eye after a short course of preoperative topical steroids. Immediate postoperatively, she was put on 1 hourly steroid topically which was tapered gradually. At 2 weeks after surgery, the patient gained a BCVA of 6/6 in the right eye. In FUS, intraoperative hyphema may be expected. Early postoperative inflammation and anterior capsular opacification may be commoner or more severe with silicone intraocular lenses. In the late postoperative period, posterior capsular opacity may be noted in some cases.

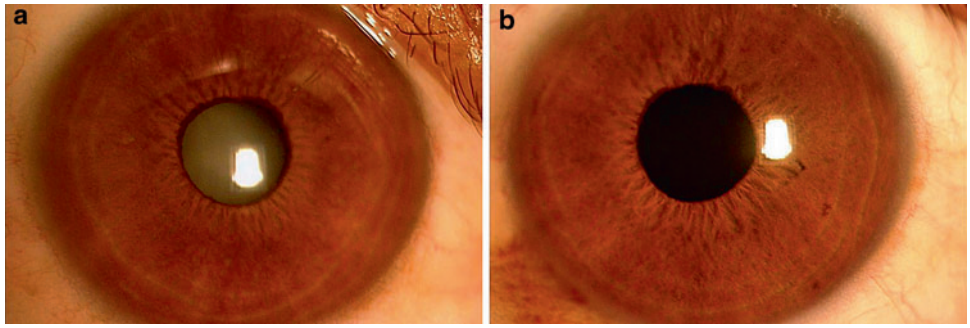

Fig. 5 The slit lamp photograph of the right eye (**a**) and the left eye (**b**) of case 2 showed cataract in the right eye and posterior chamber intraocular lens in the left eye. There was some loss of definition of iris surface in both the eyes

Key Points

- FUS is usually silent without redness, photophobia, or pain.
- Diffuse stellate keratic precipitates with minimal anterior segment inflammation are seen.
- Unilateral glaucoma, cataract, and vitreous floaters are common.
- There is diffuse stromal atrophy which may or may not lead to heterochromia.
- Steroids and cycloplegics are usually not required, though perioperative steroid is vital for successful cataract surgery.
- Phacoemulsification with hydrophobic intraocular lens implantation within the capsular bag usually provides good visual outcome.
- Glaucoma can cause severe vision loss and may be difficult to manage.

Suggested Reading

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