PRESUMED FUCHS' UVEITIS SYNDROME – A Case Report and Review of Literature

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SUMMARY

Fuchs' uveitis syndrome (FUS) is an uncommon form of uveitis. A case of presumed FUS seen in a patient at the eye clinic of the University of Ilorin Teaching Hospital (UITH), Ilorin, Nigeria is presented. An update on FUS is provided. A high index of suspicion is advocated for cases of asymptomatic, unilateral uveitis associated with cataract. Cataract surgery in these types of patients is not associated with a fulminant post-operative inflammation common with other forms of uveitis.

Key words: Fuchs' uveitis syndrome, cataract

INTRODUCTION

Lens opacities with associated changes in iris colour (heterochromia) had been observed as early as the 19th century[1]. Later, a series of patients with heterochromic uveitis were described by Ernst Fuchs.[2] This condition has undergone extensive review since then.[3-5] Fuchs' uveitis syndrome (FUS) (previously called Fuchs' heterochromic iridocyclitis) typically affects young people, without pain, redness or photophobia. They may present with floaters (as a result of the occurrence of posterior vitreous detachment), symptoms of blurred vision from cataract. The syndrome is rare, with an incidence of between 1.5% and 4.5%.[4,5] Often the diagnosis of uveitis is an incidental finding from examination for a different clinical entity causing blurred vision such as refractive error or cataract. FUS is almost universally associated with cataract,[4] initially posterior subcapsular, but can rapidly progress to complete mature cataract.

The classical findings in FUS are heterochromia, keratic precipitates (KPs) – fine, stellate and involving the whole of the endothelial surface – low-grade iridocyclitis and iris atrophy. More importantly is the absence of cystoid macular oedema (CMO) and posterior synechiae despite chronic inflammation. FUS is mostly unilateral; bilateral in 5-10% of cases.[3,7] It is one of the most under-diagnosed uveitis syndromes, often misdiagnosed as intermediate uveitis, posterior uveitis, panuveitis and granulomatous uveitis.[6] This is made worse by the fact that heterochromia is not a universal feature in FUS, hypochromia is mild or absent especially in darker irides (Africans); even in paler irides, the darker inner iris pigment can be exposed from the loss of the anterior iris stroma (inverse heterochromia).[5] Also, the heterochromia could antedate the features of FUS long before the visual symptoms develop.

A case of presumed FUS was seen at the eye clinic of the University of Ilorin Teaching Hospital (UITH), Ilorin, Nigeria. We are not aware of any other similar report from this hospital.

CASE REPORT

A 32-year-old female tailor presented at the eye clinic of the University of Ilorin Teaching Hospital (UITH), Ilorin, Kwara State, Nigeria in June 2005 with blurring of vision in the right eye of 2 months duration. The blurred vision was insidious in onset, painless, progressive and worse with distant vision. There was no antecedent trauma, redness, eye discharge or photophobia. She denied any history of topical eye or systemic medication. She was not a known diabetic or hypertensive patient. No history of joint pains or swelling, genital or mouth ulceration.

On examination, unaided visual acuity (VA) using the Snellen chart was 3/60 and 6/5 in the right (RE) and left (LE) eyes respectively. Her vision did not improve with pinhole in the RE. About 15° exotropia of the RE was noticed. Further examination of the RE showed normal lid appearance, white conjunctiva, clear cornea, normal anterior chamber depth, round and active pupil, no posterior synechiae. The lens was opaque with no view of the fundus. There was good light projection in

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the RE. The intraocular pressures (IOP) were 18 and 16mmHg in the RE and LE respectively. The LE was essentially normal. Slit lamp biomicroscopic examination (SLE) of the RE revealed uniform and evenly distributed fine, round keratic precipitates (KP) involving the whole corneal endothelium; there was moderate aqueous flare and absence of inflammatory cells in the anterior chamber. SLE of the vitreous in the RE was precluded by the lens opacity. The SLE of the LE was normal.

Figure 1 is a photograph of patient with the RE showing a matured cataract and fine round KPs on the corneal endothelium in a white quiet eye.

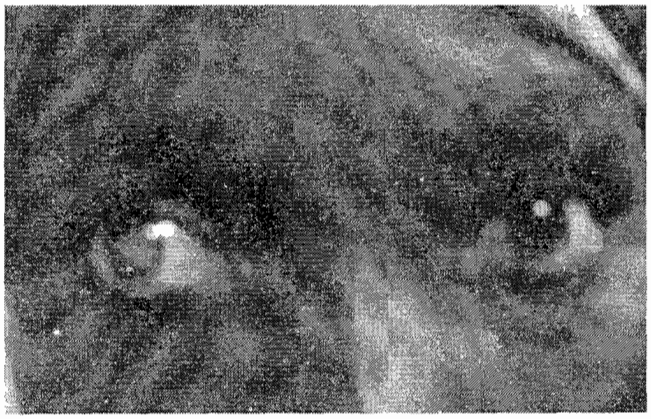

Figure 1. A 32-year old patient with RE mature cataract with fine, round KPs in a white quiet eye.

Ocular ultrasonography (US) of the RE showed an increased echogenicity in the lens region, suggestive of cataract. No focal vitreous/retinal lesion was seen. Orbital soft tissues were within normal limits. US for the LE demonstrated no focal lesion within the eye and the orbit. Haematological investigations were within normal limits – FBS - 5.5mmol/l; PCV - 37; WBC - $5.3x10^9$; ESR - 12mm/hr. The VDRL test was negative. The patient's blood pressure was 110/80mmHg.

The patient had a posterior sub-tenon injection of depo-medrol 40mg without any change in the clinical picture. No other topical ocular anti-inflammatory medication was administered. After biometric examination, the patient had a RE extracapsular cataract extraction with the insertion of a posterior chamber polymethylmetacrylate (PMMA) intraocular lens of 20.00 diopters (RE ECCE + PC IOL) under local anaesthesia with retro bulbar and facial block of 1% xylocaine (Lignocaine) injection. The patient had a sub-conjunctival injection of gentamycin 20mg and dexamethasone 2mg. There was no intra-operative complication.

On the 1st post-operative day, the visual acuity (VA) in the RE was 6/60 with an intact wound, mild striae keratopathy, round pupil and in situ PC IOL. The vision in the RE improved to 6/36 on the 2nd post-operative day and the patient was subsequently discharged on topical eye medications including an antibiotic, a mydriatic and a steroid eye drops. The SLE of the RE on discharge showed minimal anterior chamber activity with IOP of 16mmHg in both eyes. On the first follow-up visit two weeks after discharge, the VA in the RE improved to 6/24 unaided and 6/9 with pinhole. The fundus of the RE was found to have a pink healthy disc with a cup disc ratio of 0.3, clear vitreous and a flat retina with normal vasculature. No macula oedema was noticed. The IOP in both eyes was 12mmHg. The topical eye medications were gradually tailed off. The VA in the RE remained at 6/9 with refraction 6 weeks after discharge.

DISCUSSION

FUS is an uncommon, chronic uveitis with an incidence of between 1.5% and 4.5% of diagnosed uveitis cases.[4,5] It is usually found in young people, unilateral in most cases and bilateral in about 5 – 10% of cases.[3,7] It is often complicated by cataract.[4,5] Heterochromia is not a helpful component of the diagnosis and when FUS is bilateral, subtle iris changes become difficult to detect. This patient had the characteristic low grade uveitis with the absence of posterior synechiae and no signs of iris colour changes. Loss of vision in this patient was due to mature cataract.

Other features of FUS are iris stroma smoothening with loss of normal corrugated texture, iris nodules – found in more than 30% of the patients[3,7,8] either on the pupil margin (Koeppe nodules) or on iris surface (Busacca nodules). This can lead to initial misdiagnosis as granulomatous uveitis. Classically, KPs are fine, stellate, involving the whole corneal endothelium; however, various types and distribution of keratic precipitates have also been described including 'mutton fat', such that KPs are not always stellate and generalized. Refractile iris crystals – Russell bodies – are also found in FUS as well as in other chronic uveitides.[9,10] Vitritis is a common finding, usually mild, and without associated retinal vasculitis. The absence of cystoid macular oedema (CMO) distinguishes FUS from other uveitis syndromes with chronic vitritis. Glaucoma is found in 15 – 59% of cases of FUS and the mechanism is poorly understood as it has not been associated with a steroid-responsive trabeculitis, and the drainage angle is usually open.[4,11] When glaucoma does occur, it is often resistant to treatment, with wide fluctuations in intraocular pressure (IOP). The use of optic disc photography and retinal tomography imaging has been advocated to detect progression. The glaucoma treatment should also be aggressive, incorporating both medical and surgical methods.[11] This patient did not present with glaucoma, neither was there any increase in the intraocular pressure post-operatively and on follow up; however, an assumption of the incidence of glaucoma with FUS in our environment cannot be made with this single case report.

FUS should be differentiated from herpetic uveitis and glaucomatocyclitic crisis (Posner–Schlossman syndrome) – conditions with hypo-pigmentary heterochromia and increased IOP. In contrast with FUS, raised IOP in glaucomatocyclitic crisis is responsive to steroids whereas glaucoma in FUS is usually resistant to treatment, both medical and surgical. Some similarities exist in the initial presentation of FUS and intermediate uveitis – young patients with vitreous floaters – however, the absence of CMO in the former and the presence of peripheral snow banking and retinal vasculitis in the later differentiate them.

Different theories[12] – sympathetic/neurogenic, infections, hereditary and immunological – have been propounded in the aetio-pathogenesis of FUS, but none has proved that FUS results from a single pathogenic process. Histopathological findings have revealed the presence of lymphocytes and plasma cells, indicating that true inflammation actually occurs;[13] however, the inflammation in FUS, as distinct from other uveitides, is low-grade, less aggressive and unresponsive to steroids.[14] The urge to treat the inflammation should be resisted except in differentiating the entity from other conditions like glaucomatocyclitic crisis and idiopathic anterior uveitis with trabeculitis, in which there is a similarity in the initial clinical appearance.

FUS rarely leads to the typical long-term complications of chronic uveitis (posterior synechiae, CMO causing foveal damage). The course of the disease does not change with any known anti-inflammatory drugs; treatment should be directed to the sight-threatening complications of cataract and glaucoma. Our patient had a seemingly uneventful post-operative period. Jakeman et al.[15] in their study, reported a 20% incidence of severe uveitis post-operatively which resolved within two weeks on intensive topical steroids; Also, Baiyeroju-Agbeja[16] reported increased uveitis post-operatively in 3 of 4 patients that resolved within the first post-operative week on application of topical steroids. Thus, even though steroids do not have a role pre-operatively, they become invaluable in the peri and post-operative periods.

Excellent results have been reported following cataract surgery with IOL in FUS than in other forms of chronic uveitides.[17, 18] The choice of IOL for insertion depends on the uveal (aqueous cells and cellular deposits on the lens) and capsular (lens epithelial migration, posterior capsular opacification) biocompatibility of the IOL. Hydrophobic acrylic lenses have been found to have better capsular biocompatibility than IOLs of other materials.[19]

The diagnosis of FUS does not prevent cataract surgery if necessary and the post-operative period is not associated with the fulminant inflammation common with other forms of uveitides.

CONCLUSION

Though an uncommon form of uveitis, the diagnosis of FUS remains clinical; careful examination and a high index of suspicion is necessary. FUS was presumed to be the diagnosis in this patient and did not prevent a planned cataract surgery. A hydrophobic acrylic lens if available gives a better post-operative result than silicone lenses.

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