Fuch's Heterochromic Iridocyclitis
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Case presentation

54 year old lady presented in Nov 1996, with history of a unilateral recurrent iritis since 1973 and PSC OS

Family history: One aunt had a degenerative corneal disease; mother had cataract. No uveitis.

Review of systems (R.O.S.): Bronchial asthma

Medication: Ocular: Maxidex Q.I.D.
Systemic: just off an antibiotic for a gyneacologic infection

Vision: best corrected vision
20/20 OD
CF OS

Anterior Segment: Kps lower cornea OS

Discoloration of iris OS
PSC OS
Phacodonesis OS

I.O.P. : 19 OD
20 OS

Posterior Segment: NA D OD, No view OS

Diagnosis: 1- Fuchs heterochromic uveitis
2- Posterior subcapsular cataract
3- Phacodonesis

Plan: ICCE, Vitrectomy, AC IOL (Due to the phacodonesis, the capsular bag would not properly support a PC IOL)
Surgery was done as above and the patient given Vexol and Ciloxan QID
Postoperative course: uneventful during the first month, vision improving to 20/25 OS.
One month postoperative, IOP OS started to rise, 38 mm. Hg.; Trusopt (carbonic anhydrase inhibitor) tid was added and pt referred to glaucoma service for evaluation.
The IOP OS was controlled by multiple topical drops but the pt lost some visual field.
Some flare-ups of the uveitis responded nicely to topical steroids.

The Disease:
Fuchs original paper on seven patients was published in 1906
Later he described 38 cases and reported the histopathology of six eyes
Today`s features of the disease are not significantly different from Fuchs` description

Epidemiology:
1.1 - 4.5 % of referred uveitis patients
M : F = 1 : 1
Age at presentation: 20 - 60 years, mean 40 y. Early symptoms are often subtle and diagnosis often delayed.
No ethnic or racial predilection.

Symptoms
Insidious onset and subtle complaints early. Pain, redness, photophobia and miosis are usually absent.

Commonest symptoms at presentation:
¾ VA (30-52%), Floaters (26-33), Discomfort (13-19), conj injection (11) and no symptoms (13).

Occasional symptoms:
heterochromia, - IOT, spontaneous hyphema

Signs

Classic triad: heterochromia, cataract and Kps.
Vitreous opacities in > 2/3 of cases and OAG in 26-59% of cases.
Dilated, poorly reactive pupil is occasionally seen.

Heterochromia
Present in 75-90%, difficult to see in bilateral cases (5-10%); in unilateral cases the hypopigmented eye is usually the affected eye.
Difficult to see in v pale blue or v dark brown irides and is no longer essential for diagnosis.

Best detected in daylight or bright overhead light, not on the slit lamp.

Iris characteristics

Depigmentation of the anterior border layer
Depigmentation and atrophy of the sroma
Smooth iris surface (loss of normal rugae).
Prominent normal radial iris vessels and iris sphincter.
Transillumination defects.
Iris nodules in up to 33%, at the pupillary border or over the entire iris.
PS are not a common feature.
Neovascularistion of iris and chamber angel (radial and circumferential v) in 6-22%.

Kps

Usually stellate or round, interspersed wispy filaments. Whitish, translucent and small to medium in size over the entire cornea with predominance inferiorly.

Anterior chamber

usually minimal reaction

Cataract

80-90% in a nine-year follow-up period and almost universal if longer. PSC which matures quickly

Vitreous

Anterior vit cells, aggregates of cells, stringy filaments and occ dense veils. White or translucent.
Cystoid macular edema and coexisting toxoplasma chorioretinal scars occasionally seen.

Differential Diagnosis

Nevus, MM, melanosis bulbi, metallic siderosis, idiopathic or secondary iris atrphy, Horner`s, chronic iritis(T.B., H.Z.,…). Rubeosis Î D.M., ischemia. Glaucoma Î Posner-Schlossman

Pathology

Chronic low grade inflammation of the iris three layers; anterior border layer, stroma and pigm epith. Lymphocytes, plasma cells and Russell`s bodies esp in ant border layer
Decreased number of pig cells in all layers esp ant border layer
Hyalinization of iris arterioles
Ciliary Body: atrophy and hyalinization of stroma
Trabecular mesh: Lymphocytes and plasma cells, rubeosis and fibrosis in advanced cases

Electron microscopy

inflammatory cells in close association with abnormal melanocytes
hyalinization of iris arterioles
C.B. atrophy and hyalinization of stroma
- total protein, oligoclonal IgG bands, immune complexes (local production of IgG & complexes) and IL-6 level.
defect in suppressor T cell function in PB
Initiating agent: ? infection (toxoplasma); autoimmune
chorioretinal lesions in 28-64%, majority of them are +ve for toxoplasma antibodies.

Treatment and Prognosis

If vision & IOT normal, Observe
Dense Kps or - IOT topical steroid but mild infl may persist & become refratory
Cataract: good prognosis for ECCE PC IOL
minor intraop hyphema (Amsler's sign),
postop uveitis & glaucoma, vit opacities,
occasional macular edema.
intensive perioperative steroid
Glaucoma: most ominous complication. Medical treatment or trabeculectomy with antimetabolite.
Vitreous opacities: significant in 18-50%.
If vision is severely affected, consider vitrectomy