

True Exfoliation in a Man with Retinitis Pigmentosa

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True exfoliation is the term given to lamellar splitting of the anterior lens capsule. Usually caused by exposure to infrared radiation, it may occur in uveitis, and rarely may be idiopathic. Retinitis pigmentosa comprises a group of retinal dystrophies involving photoreceptors and retinal pigment epithelium characterized by night blindness and progressive visual field loss. We present a case of idiopathic true exfoliation of the lens discovered on routine slit lamp examination, in an elderly man, discovered to have bilateral classic retinitis pigmentosa as well. True exfoliation coexistent with retinitis pigmentosa has not been reported so far in literature.

T rue exfoliation or capsular delamination¹ refers to thickening and splitting of superficial part the lens capsule from the deeper part with extension into the anterior chamber². Pathogenesis of this rare condition is unclear, and causes include exposure to intense heat or infrared rays, uveitis, cataract surgery or trauma²⁻⁷. Idiopathic^{7,8} true exfoliation of the lens has long been under reported.

Retinitis pigmentosa (RP)⁹ is the term used for a set of hereditary disorders of variable presentation, involving the photoreceptors and retinal pigment epithelium, resulting in progressive visual loss due to photoreceptor death, night blindness, and constriction of visual fields.

True exfoliation has never been reported in a patient with retinitis pigmentosa. Although, this combination could be a complete chance occurrence, but we decided to report this unique case.

CASE REPORT

A 73 year old man presented to our outpatient department of Fauji Foundation Hospital, Rawalpindi, with complaints of gradual, progressive decrease in vision of the left eye for the past four months. He also gave a vague history of having difficulties in night vision. He had undergone cataract surgery in the right eye, one and a half year ago, and had no problems

with it. Otherwise, he had never had any eye problems. He did not have any co morbid systemic illness. Family history was also negative for any ocular disease. He had never worked in a glass factory and did not give a history of exposure to infrared light or trauma.

On examination, best corrected distance vision in the right eye was 6/6, and in the left eye 6/36. Slit lamp examination revealed bilateral arcus senilis, bilateral mid-peripheral iris atrophy, pseudophakia OD, and the left eye showed a diaphanous membrane arising from the anterior capsule; attached on one end, folding of the lamella on itself, and floating and undulating in the anterior chamber (Figures 1 – 5). The lens had grade 3 nuclear sclerosis, and cortical cataract grade 2. The pupils dilated fully on mydriasis, with no signs what so ever of pseudoexfoliation. Intraocular pressures were 17 mm Hg OD and 18 mm Hg OS. Fundus examination revealed normal discs bilaterally, with a CDR of 0.2, bilateral mid-peripheral bony spicules of retinal pigment, beyond the arcades, retinal pigment epithelial atrophy extending from the mid-periphery to the macular periphery, with relative preservation on the maculae and a dull foveal reflex. The vessels were attenuated (Figures 6 – 8). Gonioscopy was done which revealed grade IV angles by Shaffer classification and prominent iris processes.

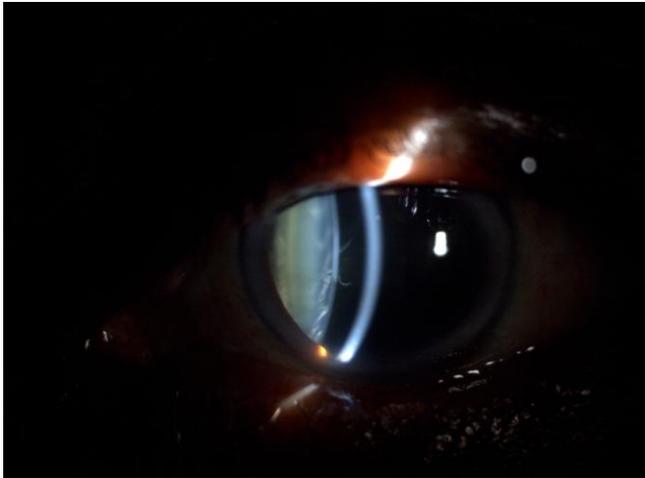


Fig. 1: Slit lamp photograph showing the capsular delamination, with the rolled lamella projecting into the anterior chamber.



Fig. 4: Folded anterior capsule visible on nasal side

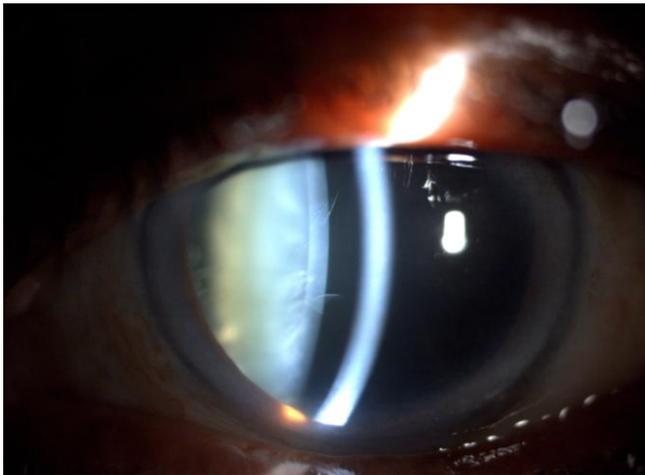


Fig. 2: The folded inferior part of the true exfoliation

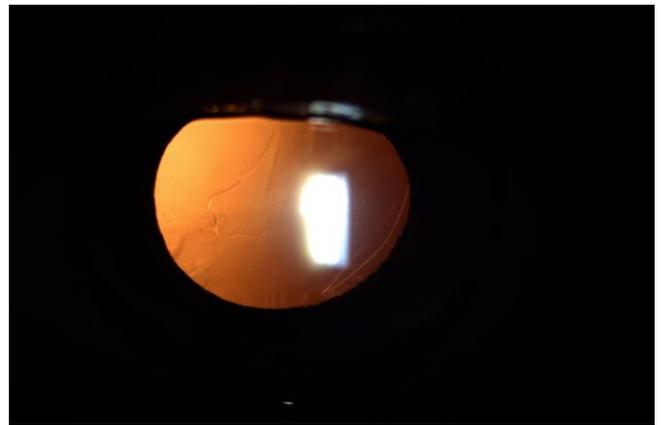


Fig. 5: Retroillumination showing entire extent of the true exfoliation



Fig. 3: Nuclear sclerosis Grade 3 and margins of the split layer

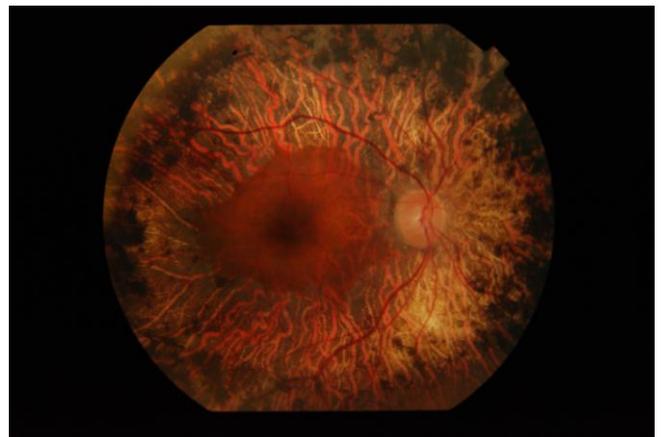


Fig. 6: Fundus photograph of the right eye showing Classic Retinitis pigmentosa, with mid-peripheral bony spicules, baring of RPE, vessel attenuation, and sparing of central macula.

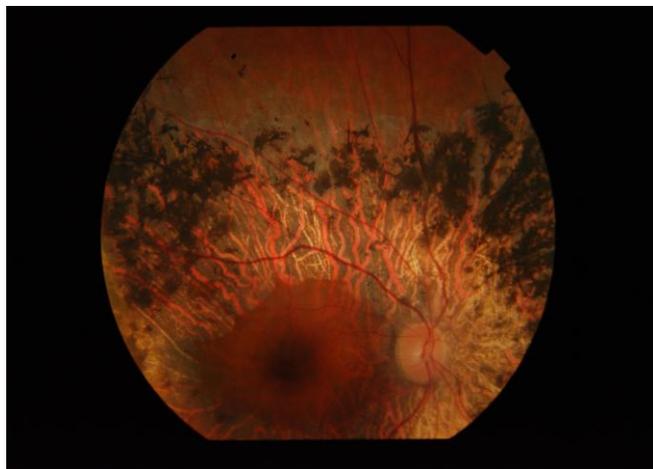


Fig. 7: A superior view of the right fundus showing the mid-peripheral involvement

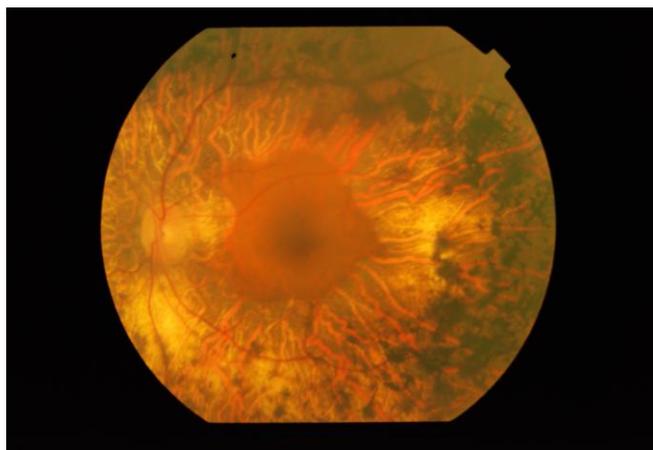


Fig. 8: Fundus photograph of the left eye, showing retinitis pigmentosa, the view being hazy due to the cataract

The history of night blindness and associated signs in the fundus led to a clinical diagnosis of classic RP, and since the maculae were spared, with good vision in the right eye, and because often patients of RP give a vague history of night blindness, and present to us when maculopathy occurs; no further investigation was deemed necessary by us. The mid-peripheral iris atrophy can also occur in elderly patients, and we consider it another chance occurrence, since there were no signs of either pigment dispersion or pseudoexfoliation.

He underwent successful cataract surgery for the left eye, and best corrected distance vision is 6/6 to date.

DISCUSSION

True exfoliation is a so rare condition, that most textbooks do not explain it. It was described for the first time¹⁰ in 1922 by Elschnig in glassblowers, and later by Punder, who noticed floating anterior capsular folds in a patient with a complicated cataract. The pathogenesis of this entity is obscure. Usually, it is classically seen in people who have been exposed to intense heat and infrared radiation over a long period, like glassblowers or blast furnace operators. This may result in rupture of the lens capsule. Recently, true exfoliation has been associated with trauma, ocular inflammation, glaucoma, hypermetropia, senility, cataract surgery, and capsular protein abnormalities.^{2-7,10,11} However; no one has ever described an association with a pigmentary retinopathy.

Transmission electron microscopy (TEM)¹¹ has demonstrated loss of lens epithelial cells along with abnormal fibrils which indicated age related degeneration as a causative influence. Heat activated proteolysis, abnormalities in capsular proteins and cellular abnormalities have been proposed as possible pathogenetic mechanisms².

True exfoliation needs differentiation¹² from the more common pseudoexfoliation syndrome; the former being a splitting of the anterior capsule with serrated or glistening, curled or scrolled margins, and the latter being a dandruff like material deposited widely across the anterior segment, and associated more frequently with an open angle glaucoma.

The term 'retinitis pigmentosa'¹³ is a misnomer due to the absence of inflammation; and encompasses all retinal dystrophies with photoreceptor loss and pigmentary retinal deposits. It has a prevalence of around 1:3000 to 1:5000. It is typically characterized by the classic triad of waxy disc pallor, arteriolar attenuation, and bone - spicule retinal pigment. Atypical RP has many forms: pericentral, central, sectorial, sine pigmento, RP punctata albescens, RP with exudative vasculopathy, and unilateral RP^{9, 13-15}.

Diagnosis is established by the presence of night blindness, fundus changes, progressive visual field loss and diminished ERG 'a' and 'b' waves. Inheritance pattern of RP may be autosomal dominant, autosomal recessive, X-linked or digenic. Ocular associations of RP are many fold being; posterior sub-capsular cataract, open angle glaucoma, myopia, keratoconus, optic disc drusen, vitreous cells, and intermediate uveitis^{9, 13-15}.

However our search of literature has revealed that true exfoliation has never been reported in a patient with RP, typical or atypical. Although, this may very well be a chance occurrence, we believe that we are the first to report this instance.

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