CLINICAL STUDY

Unilateral capsular glaucoma after long-standing bilateral pigmentary glaucoma

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Abstract

In 1964 we treated a 42-year-old woman diagnosed to have classical bilateral pigment dispersion syndrome combined with an intraocular pressure (IOP) of up to 28 mmHg in both eyes. The patient now has a daughter, also with pigment dispersion syndrome. Miotic treatment brought the IOPs to normal, and 8 years later when the disease was in the inactive phase all treatment could be discontinued. At the age of 67 years, exfoliation deposits became visible in the right eye only, but the IOPs were still below 21 mmHg. Four years later, the pressure of the right eye rose to 31 mmHg. During the next few years all available IOP-lowering medications and laser trabeculoplasty failed, and finally trabeculectomy had to be performed to keep the pressure of the right eye under control and to halt visual field changes, which had already appeared. This case illustrates that development of exfoliation syndrome may take place irrespective of pigment dispersion, and that their simultaneous occurrence may lead to an IOP rise that is resistant to medical therapy and laser trabeculoplasty. It additionally provides further clues to the pathogenesis of capsular glaucoma.

Key words Capsular glaucoma, Exfoliation syndrome, Pigment dispersion syndrome, Pigmentary glaucoma, Pseudoexfoliation, Secondary glaucoma

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Pigment dispersion and exfoliation syndromes are well-defined separate clinical entities, both of which may lead to secondary glaucoma. Pigment dispersion is fairly rare and seen mostly in young to middle-aged and myopic males, whereas exfoliation syndrome is much more common and found in older individuals irrespective of their refractive error. In pigment dispersion syndrome, pigmentation of the trabecular meshwork is dense around the entire chamber angle circumference, while in exfoliation syndrome the pigmentation may be more irregular. Krukenberg's sign is spindly only in pigment dispersion,[1] while Sampaolesi's line can be found in both conditions. The typical iris transillumination defects of pigment dispersion are radial, slit-like and develop in the mid-peripheral iris, whereas those of exfoliation syndrome, if visible, are circumferential and lie close to the pupillary margin.

The combined occurrence of pigment dispersion and exfoliation syndromes has been reported[2] in a paper describing 5 cases. These authors stressed the uncontrollable nature of chronic glaucoma in eyes harbouring both conditions. We describe an additional patient who has been followed up by the senior author for 34 years, and in whom these two syndromes occurred sequentially.

Case report

A 42-year-old white woman was seen by the senior author in 1964. Her refraction was close to emmetropia, but she nevertheless had a positive bilateral Krukenberg's sign, heavy pigment dispersion throughout the anterior segment and numerous radial iris transillumination defects. Dense uniform pigmentation was evident in the chamber angle. On repeated examinations the intraocular pressure (IOP) was between 27 and 28 mmHg in both eyes. Her optic discs and Goldmann visual fields were normal. She has now a daughter in whom bilateral pigment dispersion syndrome was later diagnosed at the age of 31 years.

Due to her young age the patient was placed on 2% pilocarpine three times a day, a therapy which she tolerated well. On repeated examinations the IOPs remained below 21 mmHg (Fig. 1). After 10 years all therapy was discontinued and the IOPs remained at the same level. The patient was now followed on no treatment.

In 1989 at the age of 67 years, the right eye showed definite exfoliation deposits for the first time. After pupillary dilatation, a clear central disc/peripheral band type of exfoliation on the anterior lens capsule was detected in the right eye, while no exfoliation was visible in the left eye. The IOP measured 21 and 20 mmHg in the right and left eye, respectively. In 1993, the IOP

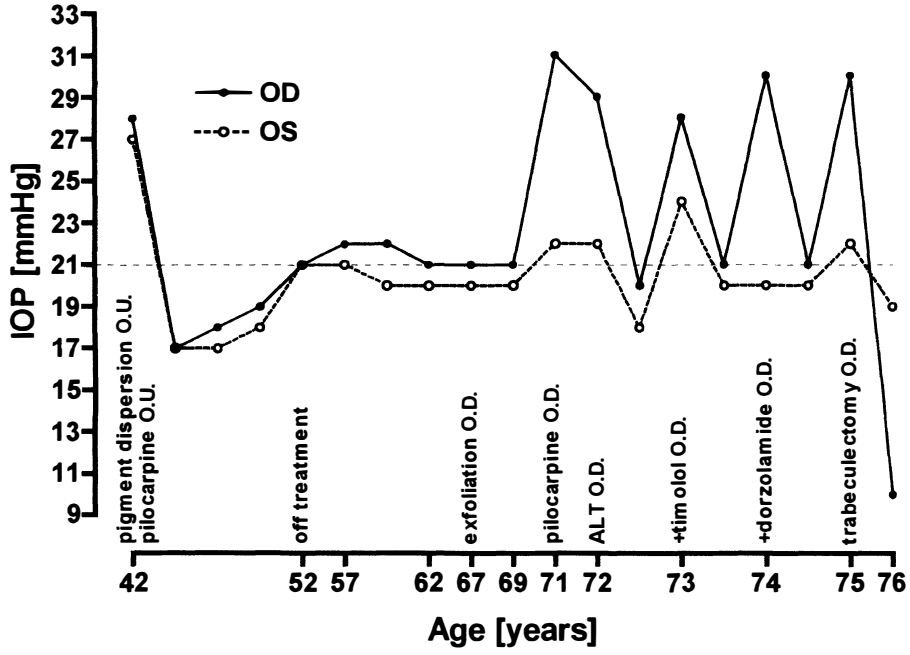

Fig. 1. *A 34-year-long follow-up of a female aged 42 years in 1964 when ocular hypertension associated with bilateral pigment dispersion syndrome was first diagnosed. After 8 years the patient was followed on no medication. In 1989, clinically unilateral exfoliation syndrome was diagnosed in the right eye, followed by a marked ipsilateral intraocular pressure rise 4 years later. Subsequent attempts to control the intraocular pressure by various medications and laser were unsuccessful. Trabeculectomy brought the intraocular pressure to the low target pressure level. O.D., left eye; O.S., right eye; ALT, argon laser trabeculoplasty.*

of the right eye rose to 31 mmHg, while that of the left eye was 23 mmHg. Treatment with 2% pilocarpine four times a day and argon laser trabeculoplasty brought the pressure down to 20 mmHg. In 1995, the IOP of the right eye was again 28 mmHg, whereas that of the left eye remained normal. Topical 0.25% timolol twice a day was added. This combination kept the pressures around 20 mmHg for an additional year.

In 1996, however, the IOP of the right eye was again 30 mmHg. Topical 2% dorzolamide three times a day was added, bringing the IOP once again down to 20 mmHg. Automated perimetry revealed a clear depression in the visual field of the right eye and a mean sensitivity that was clearly below the normal range. The findings in the left eye were considered to be normal. One year later, trabeculectomy had to be undertaken because of loss of IOP control with maximum tolerated medical therapy and the development of glaucomatous optic nerve damage. After 8 months the IOP of the right eye is 10–12 mmHg on no therapy.

Discussion

Pigment dispersion and exfoliation syndromes are separate clinical entities, even though pigmentation of the anterior segment and chamber angle is a unifying feature. The dispersed pigment may accumulate along Schwalbe's line (Sampaolesi's line). The main differential diagnostic criteria for the two syndromes are presented in Table 1. Pigment dispersion is diagnosed in young to middle-aged individuals and tends to remit with time, whereas exfoliation emerges by the age of 50 years and tends to increase with advancing age.

In two North American populations, pigment dispersion was detected in 18 of 934 individuals (2.5%) screened for glaucoma.[3] The prevalence of exfoliation syndrome in Finland has been reported to be 14% in the age group of 60–69 years, 22% in the age group of 70–79 years and 35% in the age group of 80 years or more.[4] Patients with pigment dispersion tend to be males, whereas exfoliation syndrome is unrelated to gender. Pigment dispersion syndrome is mostly bilateral, whereas in up to 60% of patients exfoliation is at least

Table 1. *Differential diagnosis of pigment dispersion syndrome and exfoliation syndrome*

	Pigment dispersion syndrome	Exfoliation syndrome
Age	25–50 years	Mostly over 60 years
Male:female ratio	2:1	1:1
Laterality	Bilateral	Initially unilateral in 30–60%
Refraction	Myopic	No predilection
Chamber angle	Wide open Sampaolesi's line	No predilection; Sampaolesi's line can be narrow
Retrocorneal pigment deposition	Positive Krukenberg's sign	No particular pattern; Krugenberg's sign less spindly
Iris transillumination defects	Radial and slit-like, in mid-peripheral iris	Moth-eaten pupillary rim plus concentric furrows
Iris blood vessels	Normal	Leaky by angiography

initially clinically unilateral.[5] Finally, in pigment dispersion syndrome the chamber angle is wide open with a typical posterior concavity of the iris, while in exfoliation syndrome the angle is mostly normal and may even be occludable. Similarities include even the presence of Sampaolesi's line.[1]

Elevation of the IOP in capsular glaucoma may be the result of blockage of the trabecular meshwork by exfoliation deposits and pigment, or may reflect tracbecular cell dysfunction. Specular microscopic findings of the corneal endothelium lend support to the theory of endothelial cell dysfunction.[6–8] Decreased endothelial cell density has been correlated with the extent of pigment dispersion in exfoliation syndrome.[9] It has also been suggested that in some cases it might be due to primary open-angle glaucoma. The negative topical corticosteroid provocative test in exfoliation syndrome has not favoured frequent association with primary open-angle glaucoma.[10–12]

Clinical studies of patients with clinically unilateral exfoliation have provided support for the mechanical theory. Of 25 patients with unilateral exfoliation, 58% developed glaucoma only in the eye involved by exfoliation during a follow-up of 1–5 years.[5] In the patient now described, exfoliation superimposed on pigment dispersion led to a marked elevation of IOP and short-lived success of medical therapy in the eye with exfoliation. This lends further support to the central role of exfoliation deposits in blocking the aqueous outflow.

Recently, the concept of truly unilateral exfoliation syndrome has been challenged.[13] In an autopsy analysis of 5 patients with clinically unilateral exfoliation syndrome, classic exfoliation deposits were indeed absent from the clinically uninvolved eye even by routine histopathology, but immunohistochemical and lectin histochemical analysis revealed exfoliation-like deposits around many iris blood vessels.

When a patient with bilateral pigment dispersion syndrome develops asymmetrical IOPs, one should consider other causes for the pressure elevation such as cleavage of the chamber angle after previous blunt trauma or the advent of exfoliation syndrome. One should be aware of the coexistence of pigment dispersion and exfoliation for the proper management of the resulting difficult secondary glaucoma. In our clinical experience, bilateral pigment dispersion and subsequent capsular glaucoma is not a rare occurrence.

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