Pseudoexfoliation Syndrome
—Case Report and Review of Clinical Features—

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Pseudoexfoliation syndrome is characterized by the presence of gray-white flakes on the pupillary borders and anterior lens capsule, increased trabecular meshwork pigmentation, and association with glaucoma. We describe 3 patients with this syndrome seen at Asan Medical Center Department of Ophthalmology in 1989, and we focus on their clinical features and management.

We believe that patients with this syndrome are not as rare in Korea as has been thought, judging by scant report of cases in the past.

Key words: pseudoexfoliation syndrome, glaucoma.

INTRODUCTION

Pseudoexfoliation syndrome (PXS) was first described by Lindberg in 1917. Contrary to an earlier belief that PXS is largely confined to the Scandinavian region, it is now known to be worldwide in distribution, although prevalence rates vary among different geographic locations.

Reports of cases in Korea have been few, and therefore it is considered to be an extreme rarity in this part of the world.

Recently, we at Asan Medical Center in Seoul, Korea, detected and managed 3 cases of PXS. All 3 patients visited our department with complaints of visual disturbances due to senile cataract. Two of them were found to be associated with glaucoma, one with acute angle closure, and the other with open angle glaucoma.

CASE REPORT

Case 1

An 86-year-old woman presented with a two-year history of declining vision in both eyes. Her past medical and ocular history was unremarkable. Her best correctable visual acuity was 0.2 OD, intraocular pressure of 9 mmHg OD and 13 mmHg OS, grade IV open angle with 3+ pigmentation of trabecular band OU, advanced nuclear and cortical cataract OU, and poorly visible fundi with no grossly visible abnormality. The most notable finding was some white flaky material seen on the pupil border (photo 1). Upon pupil dilatation, the whitish-flaky material was observed on the anterior capsular surface with the characteristic 3 zones of translucent central disc, intermediate clear zone, and granular girdle in the periphery with radial striations (photo 2). Routine cataract extraction was recommended.

Case 2

A 72-year-old woman presented with the complaint of slowly declining vision. Her past history
was unremarkable. Best correctable visual acuity was 0.5 OD and 0.6 OS; IOP of 10 mmHg OD and 15 mmHg OS; deep anterior chamber and grade II open angle all around without synchiae; 2+ pigmentation of trabecular band OU, and pigment deposits along and anterior to the Schwalbe’s line (photo 3); moderate cataract OU; and normal fundi with healthy discs.

The pupil borders lacked whitish deposits, but there were suspicious pupillary ruff defects (photo 4). The anterior capsule surface showed a thin layer of whitish granular material that lacked the distinct pattern of Case 1 and could escape detection without full dilatation and careful observation. Some time later, the patient returned with the symptoms and signs of acute angle closure glaucoma in the right eye, which was successfully managed by argon laser iridectomy.

**Case 3**

A 75-year-old man was first seen in June 1989 with a 4-year history of unilateral glaucoma in the right eye. The past record indicated a progressive and erratic course with wide fluctuations in IOP under medical therapy. One episode of an acute rise of IOP to 50mmHg with corneal edema, but without evidence of angle closure, was recorded, and laser iridectomy was subsequently performed. Meanwhile, visual field loss steadily increased. At the time of our examination, the right eye showed a visual acuity of 0.2, an IOP of 25, deep anterior chamber, miotic pupil with whitish granular material on the pupil border, and pupillary ruff defects, grade III open angle with 3+ pigmented trabecular band, advanced nuclear and cortical cataract, and a severely cupped disc with a C/D ratio of 0.8. Visual field showed advanced field loss with central and inferior temporal quadrant remaining.

Due to years of pilocarpine use, the pupil could not be dilated wide enough to observe the exfoliative features of the anterior capsule. The left eye was entirely normal with visual acuity of 1.0, an IOP of 18, and no signs of any abnormality. The IOP in the right eye remained in the high 20s under maximal medical therapy over the following weeks, and in October, a 180-degree argon laser trabeculoplasty (LTP) was performed.

Subsequently, the IOP dropped to the low 20s and high teens, except for one spike up to 38 when the patient discontinued medication for several days. Two months later the eye underwent ECCE combined with trabeculectomy. Despite the failure to form a lasting bleb, the IOP has remained controlled to this day, due presumably to the effects of medication and LTP.

**DISCUSSION**

Pseudoexfoliation syndrome is now known to occur in all parts of the world, and the frequency of its occurrence has been reported to vary widely from country to country and from one part of the same country to another. Due to scant reports of this syndrome in Korea, it is considered to be a rarity in this region. The 3 cases reported here were all seen in the first 6 months of the opening of the Asan Medical Center, during which period the average number of outpatient visits to the Department of Ophthalmology numbered less than 40 per day. This represents a high prevalence rate of PXS among our outpatient group, but it probably does not reflect the true rate for this region, and it is equally probable that the condition is not an extreme rarity either. Assuming the latter assumption to be correct, the reason for the scant findings and reporting of cases may lie in the low index of suspicion by ophthalmologists and relatively infrequent pupil dilatation studies during routine clinical examinations. Some cases of PXS may escape detection easily be casual and less-than-complete and thorough examination, since the main feature of exfoliation material can be subtle and require full pupil dilatation for detection on the lens capsule, a condition not always possible in eyes under miotic therapy or in other situations where pupil dilatation is omitted for various reasons.

The second case is an example of a subtle exfoliative feature that could have gone unnoticed and been regarded as a simple angle closure glaucoma. Given the increased awareness and interest of PXS, plus conditions that allow more routine pupil dilatation studies, more cases may come to light.

Recognizing and diagnosing PXS is not merely of causal or academic interest only, for it has significant implications in prognosis and management when cataract or glaucoma is associated. Its
association with various aspects of glaucoma is well-established,3-5 and the open angle glaucoma associated with PXS is considered a true secondary glaucoma apart from the primary open angle glaucoma.6 The second of our cases exhibited a not-so-rare association with angle closure glaucoma.7 Although the management and immediate outcome of such angle closure glaucoma are not different from other angle closure glaucoma, the long-term outlook should be considered potentially different in that the open angle form is more likely to develop later, since pseudoexfoliation is present and, therefore, must be watched more closely for an extended period.

Our third case demonstrated the characteristic of increased difficulties in management of PXS glaucoma, particularly when the condition is unilateral.8 The eye showed erratic and poor response to medical therapy with fluctuating IOP and progressive field loss, despite maximal medical therapy. One episode of acute glaucoma with high pressure and corneal edema resulted in someone performing a peripheral iridectomy, although the angle was open. It did respond favorably to laser trabeculoplasty, since it has been widely regarded as a particularly effective treatment for PXS glaucoma. However, longer and closer follow-ups seem to indicate relatively shorter period of effectiveness from laser therapy compared with primary open angle glaucoma, and it is recommended that full 360-degree treatment in two divided sessions be done rather than the more popular single 180-degree treatment. Our patient may need a second laser treatment should the IOP rise again, since no lasting filtration bleb was established by ECCE trabeculectomy procedure. It is held that surgical therapy for PXS glaucoma is as effective as for primary open angle glaucoma, notwithstanding the example of our case.

Another significant aspect in PXS management relates to weak zonules and cataracts. Lens zonules are not only the site of exfoliative material deposits, but they have been suspected as being one of the possible sources of the exfoliative material, although the unequivocal source is yet to be determined.9 Clinically they can present problems by zonular breaks during or after cataract surgery as has been experienced by many.10 Therefore, due care must be exercised in preventing the break, in recognizing it when it occurs, and perhaps deciding wisely to implant the IOL in the ciliary sulcus rather than in the capsular bag, even though all the zonules are thought to be intact, which no one can be certain of during surgery. Such care may include use of continuous circular capsulorhexis instead of capsulotomy, use of hyrodissection, and avoiding phacoemulsification.

REFERENCES

**Photo 1.** Case 1 shows abundant gray-white flaky material deposited along the entire pupil border.

**Photo 2.** Case 1 shows the anterior capsular surface with clear intermediate zone on the right and peripheral zone on the left with heavy deposits of radial striations and curled-up edge.

**Photo 3.** Case 2 shows the open angle with moderate pigmentation of trabecular band and multiple pigmented lines (Sampaolesi lines) on and anterior to the Schwalbe's line.

**Photo 4.** Case 2 shows no pupil deposits but with suspicious pupillary ruff defects and subtle whitish-granular deposits on the anterior capsular surface.