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Pseudoexfoliation Syndrome Showing Different Deposition Patterns in Pseudophakic Eyes: Two Case Reports

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Dear Editor,

Pseudoexfoliation (PEX) material is composed of basement membrane substances produced by lens epithelial cells and various ocular tissues and it deposits in the ocular tissues through aqueous humor [1]. Its pathogenesis has been explained as disturbances in the synthesis or assembly of microfibrils, oxidative stress, inflammation, disruption of blood-ocular barrier, various environmental factors, and high association with the lysyl oxidase-like-1 gene [2,3].

PEX is an extracellular matrix-related systemic disorder that increases in frequency with age. Therefore, it seems natural for eyes to gradually accumulate pseudoexfoliative materials over time, even after cataract surgery. However, PEX in pseudophakic eyes is actually very rare. We tried to explain the rarity of PEX in pseudophakic eyes through illustrations comparing two cases: one showed an asymmetrically wedge-shaped pattern of tiny and coarse PEX materials deposits in the tilted and a decentered intraocular lens (IOL); the other showed symmetrically concentric deposits on a well-positioned IOL.

The study was approved by the Institutional Review Board of Chung-Ang University Hospital (No. 2306-020-19476). Written informed consents for publication of the research details and clinical images were obtained from patients.

Case 1 is a 71-year-old man presented with glaucoma in his right eye since 2015 (Fig. 1A–1E). He underwent cataract surgery in 2005. Whitish-gray, tiny dandruff-like deposits were observed through the iris margin and were more prominent in the temporal than the nasal area. PEX deposits covered the temporal half of the pupil (Fig. 1A). After pupil dilatation, the IOL was observed to be tilted temporally and decentered nasally. PEX materials forming incomplete concentric circles with radial striations were prominent on the temporal side near the posterior surface of the iris (Fig. 1B).

Case 2 is a 76-year-old man underwent phacoemulsification in 2015 and at the time of cataract surgery, no significant abnormalities were observed, including PEX (Fig. 1F–1K). His next clinic visit was 8 years later, with glaucoma. No abnormalities were found in the pupil margins and angles; however, whitish-gray, fine and coarse granule-like deposits forming concentric ring with radial striations were observed on the anterior surface of the IOL. More deposits were found in the lower portion (Fig. 1I).

PEX deposits originate from lens epithelial cells, iris, and ciliary epithelial cells. Therefore, even if the lens epithelial cells are substantially removed by the capsulotomy, fibrillary deposits can still be formed by other intraocular cells. Reported rare cases of pseudophakic PEX syndrome occur between 2 and 20 years after cataract surgery, and progression to PEX glaucoma is uncommon [4,5]. Sulcus insertion of the IOL might increase the accumulation of deposits on the IOL surface [1,5].

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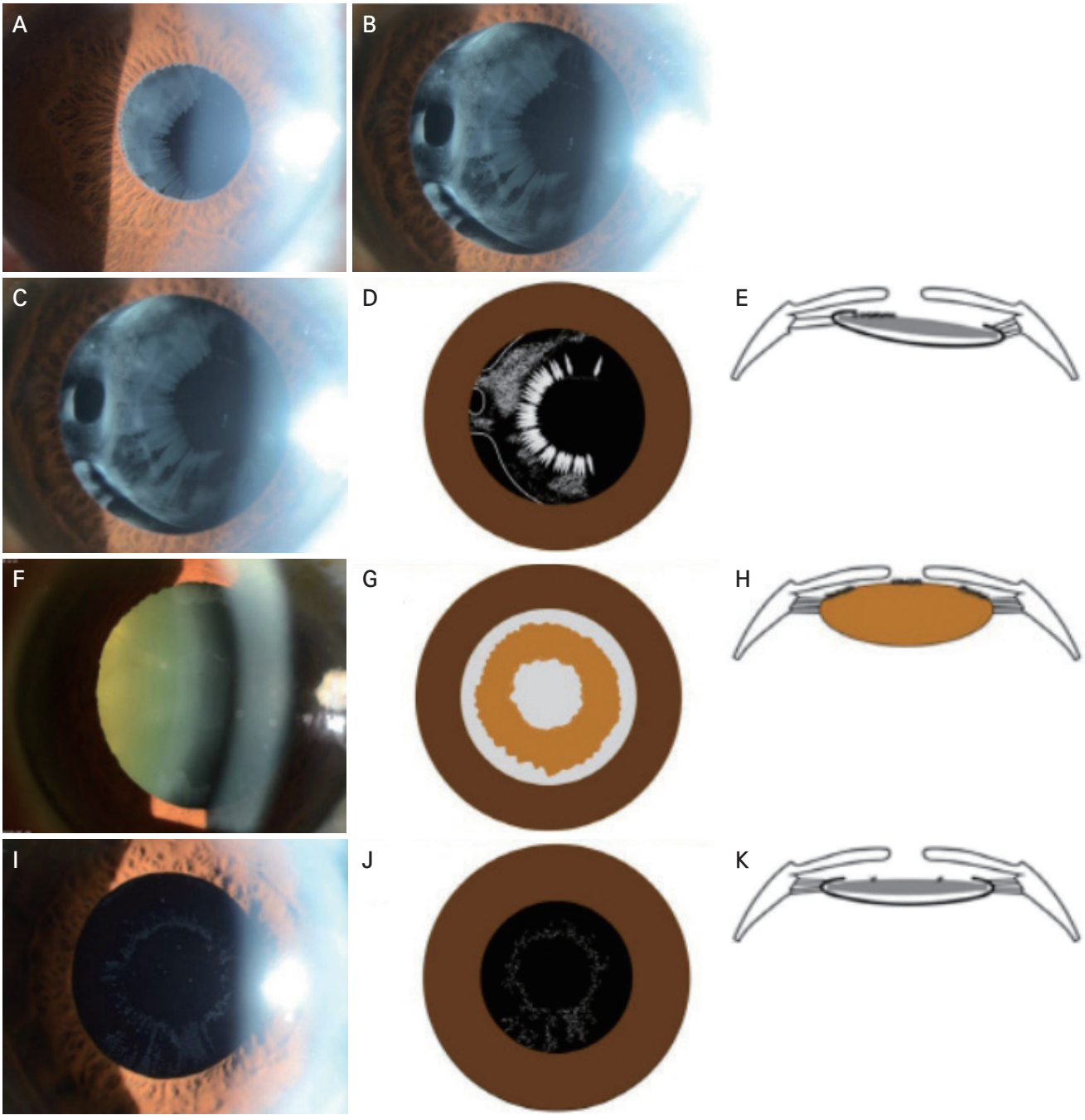

Fig. 1. Slit-lamp images of the right eye of (A–E) case 1 and (F–K) case 2. (A) Before pupil dilatation, grayish opaque tiny flakes of dandruff-like pseudoexfoliation (PEX) materials are seen around the iris margin, with more deposits on the temporal side than the nasal side. PEX deposits are covering the temporal half of the pupil. (B) After pupil dilation, the intraocular lens (IOL) was observed to be tilted temporally and decentered nasally. Prominent deposits of fine and coarse opaque PEX materials have formed incomplete concentric circles with radial striations on the temporal side, closer to the posterior surface of the iris. (C–E) In case 1, tiny dandruff-like fakes with wedge-shaped, imperfect concentric circles and radial striations on the temporal side of the IOL. (F–H) PEX syndrome in the lens capsule exhibits a characteristic three-ring patterns: a central disc, middle clear zone, and peripheral cloudy ring. (I–K) In case 2, concentric deposits of fine granule-like PEX materials are seen in the paracentral area. The accumulation of deposits slightly thicker in the inferior portion of IOL.

Concentric and radially striated PEX deposit patterns on the IOL are similar to the crystalline lens of PEX syndrome (Fig. 1F–1H). This suggests that deposition occurs in the areas most exposed to aqueous humor in which high

concentrations of deposits exist. By contrast, PEX deposits on the IOL are rare and the patterns are distinct from those seen with a crystalline lens: a clear center and a radially striated middle zone (Fig. 1C, 1I). The reason for this difference is because most of the lens epithelial cells, which are the source of deposits seen on a crystalline lens, are removed in pseudophakic eyes, and the distance between the iris and the IOL increases after surgery, which makes the flow of aqueous humor freer than in phakic eyes. Consequently, the concentration PEX materials is lower and easier to wash out. Therefore, deposit accumulation on the IOL takes a long time and PEX deposits in the middle area where the distance between the IOL and the iris is relatively short (Fig. 1I–1K).

Case 1 showed an interesting sectoral distribution of PEX deposits, with asymmetric concentric circles with radial striations in temporal area. We suggest that the PEX materials were piled up thicker at the anteriorly tilted temporal side of IOL, and then later IOL decentered nasally (Fig. 1C–1E)

The clinical significance of PEX material deposits on the IOL may be a sign of progression from PEX syndrome to PEX glaucoma. It is important to carefully observe the iris margin because PEX deposits are difficult to see in IOLs. Furthermore, because PEX deposits accumulate in the mid-periphery rather than in the center, it is not easy to detect the loss of visual acuity. Therefore, it is needed that patients with PEX syndrome should be regularly checked using a slit-lamp biomicroscopy with pupil dilation to early detect the PEX material deposits on the IOL surface.

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