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Sir,
Ultrastructure of anterior lens capsule in Peters' plus syndrome

The Peters' anomaly is characterized by central corneal opacity (leukoma), thinning of the posterior aspect of the cornea, iridocorneal adhesions, and keratolenticular adhesion or cataract. The presence of lens abnormalities in Peters' anomaly is more frequently associated with systemic anomalies.¹ Peters' plus syndrome (PPS) is a rare entity that combines anterior chamber abnormality of Peters' anomaly with other systemic anomalies such as cleft lip and palate, short stature, broad hands and feet, and variable mental delay.² The etiology is unknown, but may involve abnormal neural crest development. Peters' anomaly has been reported to be caused by mutations in several genes such as *PAX6*, *PITX2*, *PITX3*, and *CYP11B1*.² PPS follows an autosomal-recessive pattern of inheritance,² but the causative gene of this syndrome remains unknown. Previous pathologic reports in Peters' anomaly revealed attenuation or absence of Descemet's membrane and corneal endothelium in the central posterior cornea, with iridocorneal adhesion or

keratolenticular adhesion.^{3–5} We report, herein, the ultrastructure of anterior lens capsule in a patient with PPS.

Case report

A 4-year-old boy was found to have bilateral cataract and referred to our department. Bilateral nystagmus was found. We observed a poor ambulatory activity of the patient while occluding his left eye with eye patch. Slit-lamp examination showed bilateral cataract with bulging anterior lens surface that adheres to the posterior cornea (Figure 1). Mild opacity was detected in the corneal stroma in front of the adhesion site. Microcornea and posterior synechia were also found in both eyes. Pediatric evaluation revealed short stature (<3%), low body weight (<3%), speech, and motor developmental delay. Growth hormone deficiency was also detected by stimulation test. The patient received extracapsular lens extraction in the right eye, with keratolenticular adhesion separated by viscoelastic material, and the anterior lens capsule removed by can-opening capsulectomy. Aspirated lens material was sent for virus culture and Herpes virus polymerase chain reaction (PCR), with both test results came out to be negative. Anterior lens capsule was fixed in 2% glutaraldehyde/phosphate buffer, and then washed three times in phosphate buffer. The specimen was post-fixed with 1% osmium tetroxide. After dehydration with alcohol series, it was embedded with epoxy/resin and sectioned. The section was examined under a JEOL electron microscope (JEM 1230). We observed villus-like processes emanated from the anterior lens capsule, with two cells still attached to the process tips. These cells were presumed to be sloughed-off corneal endothelial cells, as the lens capsule adhered to the corneal endothelial side before surgical separation (Figure 2, En). Intact lens epithelial cells could be found adjacent to the lens capsule (Figure 2, Ep).

Comment

Keratolenticular adhesion is characteristic in PPS. Various pathologic results have been reported in the literature.^{3–5} The lens may be adherent to the corneal stroma with absence of Descemet's membrane and lens capsule.⁴ A stalk-like connection between lens and cornea has been reported as well, and the Descemet's membrane was deflected over onto the stalk.⁵ The lens may be in contact or embedded in the defective posterior corneal surface, but with an intact anterior lens capsule.⁴ The lens may be only opposed and not adherent to the posterior surface of the cornea, with both the anterior

Figure 1 Peters' anomaly, (a) Slit-lamp examination of right eye showed keratolenticular adhesion in the central cornea and posterior synechia. Inset showed a slit view of the contact region between the anterior bulging lens surface and the posterior cornea. The cornea was clear except the contact region, where there was mild haze in the corneal stroma. (b) Slit-lamp examination revealed similar findings in the left eye.

Figure 2 Ultrastructure of anterior lens capsule. (a) Villus-like processes were observed to emanate from the anterior lens capsule (cap). Several cells attached to the processes were detected. (b) These processes emanated from the anterior lens capsule, aligned regularly along the margin, with a narrow connecting neck. (c) Attached cell showed a dense nuclei with irregular cell margin, which may result from the mechanical separation of the adhesion. It is presumed to be a sloughed-off corneal endothelial cell (En). (d) In other region of the lens capsule, intact lens epithelial cells (Ep) could be found adjacent to the lens capsule. Focal degeneration of lens capsule was found as well (asterisk). Scale bar is 5 μm in (a), 2 μm in (b), 1 μm in (c) and (d).

lens capsule and cornea intact.³ To our knowledge, villus-like processes extending from the anterior lens capsular surface has never been reported in literature.

These processes align regularly along the margin, with a narrow connecting neck. These processes were about 0.5 μm in width and 10–15 μm in length, with some

electron-dense particles between them. Since the syndrome is genetically heterogeneous, this finding may not be generally applicable.

In literature, several developmental theories have been proposed for the pathogenesis of Peters' anomaly, and none of them explains all the clinical and histopathologic findings in all forms of Peters' anomalies.² A failure of neural crest cell differentiation destined for corneal endothelium and Descemet's membrane is considered the most reasonable explanation for the keratolenticular or iridocorneal adhesion. We propose that these villus-like processes might represent the developmental remnants that fail to regress while corneal endothelium separates from the underlying lens epithelium.

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Sir,

Progression of diabetic retinopathy following coronary artery bypass graft

We read with interest Mansour *et al*'s¹ paper highlighting the risk of anterior ischaemic optic neuropathy (AION) in diabetics after coronary artery bypass graft (CABG) surgery. We present a diabetic patient who suffered significant loss of vision owing to rapid progression of retinal ischaemia following CABG.

A 58-year-old man was diagnosed with mild non-proliferative diabetic retinopathy (NPDR) in October 2002. His vision was 6/6 in both eyes. In July 2004, his vision decreased to 6/18 in the right eye and 6/9 in the left eye. A fundus fluorescein angiogram (FFA) detected ischaemic maculopathy in both eyes (Figure 1.) Six months later he developed new vessels in the left eye and had two sessions of panretinal photocoagulation. His vision at this stage was 6/12 in both eyes. Two months later he underwent a three-vessel CABG. The preoperative haematocrit was 0.432. Postoperatively the lowest haematocrit was 0.164. The patient received one unit of packed red blood cells. The surgery and postoperative period was uncomplicated. Six weeks following the surgery his vision reduced to 6/60 in his right eye and 6/24 in left eye. Clinically the right eye had severe NPDR. The left eye showed new vessels at the disc. Both discs were healthy. FFA showed large areas of capillary dropout and increased macular ischaemia (Figure 2). Two months later his right eye developed rubeotic glaucoma causing the vision to drop to hand movements. Since then he has had extensive panretinal photocoagulation in both eyes and cycloablation of the ciliary body in the right eye. At present he has no perception of light in the right eye and hand movements in the left.

The retinal ischaemia in this patient was probably precipitated following CABG. It may be argued that his retinopathy was progressing preoperatively and may have

Figure 1 FFA of the left eye showing ischaemic maculopathy.