Sir,
Posterior polymorphous dystrophy with polycoria and corectopia

Posterior polymorphous dystrophy (PPD) is a bilateral, inherited condition affecting the corneal endothelium and Descemet's membrane. The common features of iridocorneal endothelial syndrome (ICE) and PPD have been reported to co-exist.¹⁻⁴ The literature suggests that PPD and ICE may represent different parts of a spectrum of one condition.

There have been no reported cases of polycoria associated with PPD to date. We describe a case with PPD and unilateral glaucoma, corectopia, and polycoria, which strengthens the opinion that PPD and ICE are entities within the same spectrum of disease.

Case Report

A 63-year-old man presented with left transient corneal oedema and bilateral corneal endothelial changes consistent with PPD 16 years ago. This resolved spontaneously following which he developed right open angle glaucoma, with intraocular pressures (IOP) of 26 and 18 mmHg, right and left, respectively, increased right disc cupping, and superior arcuate field loss on 24-2 Humphrey's test. Glaucoma control, with no further field or disc deterioration, was achieved with topical beta-blockers. Within the last year, he developed right corectopia and polycoria (Figure 1a). Gonioscopy confirmed three full thickness iris defects near the iris root (Figure 1b). The defects had well-defined margins with no previously noted iris atrophy. These changes were in addition to band-like and vesicular endothelial changes.

Specular microscopy showed typical features of PPD including polymorphism and decreased endothelial cell count (cell density—657/mm²).⁵ Examination of his daughter revealed bilateral features of PPD. His two grandchildren had no corneal abnormalities.

There was no other familial evidence of either PPD or ICE.

Comment

Clinically, specular microscopy and histochemical features aid to distinguish between PPD and ICE syndromes.

The bilateral endothelial changes, inheritance, age, and sex predisposition suggest a diagnosis of PPD, however, the presence of unilateral glaucoma and iris abnormalities typical of ICE syndrome complicates the diagnosis in this case. Polycoria has not been previously reported in cases of PPD and similarly band like endothelial changes have not been reported in ICE. All other features are common to both conditions and appear to vary in severity.^{1,3,4}

We believe that a morphological spectrum involving both PPD and ICE may be explained by a 'two-hit hypothesis' similar to that suggested in a case with keratoconus, iris atrophy, and PPD.⁶ A first hit would cause 'epithelialization' of the corneal endothelial cells with multilayering and pleomorphism, correlating with the clinical signs of PPD. A second hit at a later stage of embryonic development, either environmental or viral, may lead to the migration of these cells, laterally giving rise to 'glass membranes', iridocorneal adhesions, glaucoma, and iris abnormalities, which are more consistent with ICE.⁷ Age, sex, and timing of these disruptions in normal embryonic development may therefore lead to variations within the clinical spectrum that has been described.³

The other possibility is that of an insult early in the development of the cornea.⁸ The posterior cornea, iridocorneal angle, and iris are all derived from a wave of neural crest tissue that occurs at 6 weeks of gestation. These structures are involved in both conditions, and may thus represent variation in clinical expression.

Immunohistochemical studies have shown endothelial cells in both conditions to demonstrate epithelial-like

Figure 1 (a) Corectopia with temporal pupillary distortion. (b) Three full thickness iris defects near the iris insertion.

features supporting the above theories of a common aetiological pathway.^{9,10}

This case supports the evidence that PPD and ICE are parts of a spectrum of one condition. We believe that patients with PPD should undergo thorough scrutiny of the endothelium, iris, and anterior chamber angle at follow-up, to aid the accurate management of these patients and their families.

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The authors have no proprietary interests.

Eye (2004) **18**, 856–857. doi:10.1038/sj.eye.6701327

Published online 30 January 2004

Sir, Ultrasound biomicroscopic documentation of traumatic cyclodialysis cleft closure with hypotony by medical therapy

A cyclodialysis cleft occurs when meridional ciliary body muscle fibres disinsert from the scleral spur. This can follow blunt trauma or intraocular surgery. Cyclodialysis creates a communication between anterior chamber and suprachoroidal space, resulting in hypotony and its resultant sequel. Shallow anterior chamber due to cyclodialysis makes the diagnosis difficult by gonioscopy. Ultrasound Biomicroscopy (UBM) is the best to diagnose and monitor cyclodialysis.¹

Several modalities for treatment of cyclodialysis have been described in the literature, like medical therapy, diathermy, cryopexy, direct and endoscopy-assisted photocoagulation, direct suturing, trans-scleral photocoagulation and vitrectomy.^{2–5} Cyclodialysis cleft closure has been documented after intervention on UBM.⁴ To the best of our knowledge, this is the first case to document resolution of cyclodialysis cleft on UBM following medical management.

Case report

A 17-year old male presented to emergency service with history of diminution of vision OD following trauma with stone for 7 days. Visual acuity was finger counting 2 m OD and 20/20 OS. Intraocular pressure (IOP) was 6 mm OD and 12 mm OS of Hg by applanation. Examination of OS was normal. On slit-lamp examination, OD showed corneal abrasions, shallow anterior chamber with 2+ cells and flare. Pupil had a sphincter tear at 11 o'clock. Fundus examination revealed retinal oedema involving inferior retina including the macula. Bscan ultrasonography revealed 360° choroidal effusion, echo-free vitreous, attached retina with normal globe contour. UBM showed the 360° choroidal effusion with cyclodialysis at 6–7 o'clock position (Figure 1). He was started on topical prednisolone acetate 0.1% eye