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PHOTO ESSAY

Unilateral persistent fetal vasculature coexisting with anterior segment dysgenesis

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Abstract Persistent fetal vasculature (PFV) is a common congenital developmental anomaly of the eye which results from failure of the embryological primary vitreous and hyaloid vasculature to regress by the time of birth (Int Ophthalmol Clin 48: 53–62, 2008). Typically, it is divided into anterior, posterior or combined types and is characterized by the presence of a vascular stalk located between the optic disc and the posterior lens capsule (Int Ophthalmol Clin 48: 53–62, 2008). Although it has been reported to manifest itself differently, in our case it presented in a microphthalmic eye as anterior segment dysgenesis with broad-based mid-peripheral synechiae, posterior embryotoxon, iridoschisis, ectropion uveae, hypotony and subluxated cataractous lens with a taut anterior hyaloid face which are rare associations with PFV.

Keywords Persistent fetal vasculature · Anterior segment dysgenesis · Mid-peripheral synechiae

A 9-year-old boy presented with vision of hand movement close to face, a smaller eye and dissociated horizontal deviation in the left eye. Circumferential posterior embryotoxon, iridoschisis, ectropion uveae and peripheral anterior synechiae and corneal opacity overlying the area of iridocorneal adhesions were evident biomicroscopically. A total subluxated cataract was observed in the pupillary zone. Anterior segment optical coherence tomography (AS-OCT) depicted circumferential broad-based mid-peripheral anterior synechiae, more prominent temporally (Fig. 1a). Intraocular pressure was 8 mmHg in the left eye. Confocal microscopy of the cornea showed normal endothelial cells in both eyes. A fully dilated evaluation revealed a subluxated total cataract and stretched ciliary processes appreciable nasally (Fig. 2a). The axial length was 22 mm in the right eye compared to 19 mm in the left eye. Ultrasonography initially showed anechoic posterior segment. However, a repeat careful sonography showed a persistent stalk joining the disc and the posterior lens capsule which was initially overlooked as it was too thin (Fig. 1b). A diagnosis of persistent fetal vasculature (PFV) was thus confirmed.

Lens aspiration, primary posterior capsulorrhexis, coagulation and cutting of the stalk with the help of a Fugo plasma blade (MediSURG R&MC) and anterior vitrectomy were performed under general anesthesia. The patient was left aphakic due to a markedly decentered bag and was fitted with a contact lens and prescribed occlusion postoperatively (Fig. 2b).

PFV results from failure of the embryological primary vitreous and hyaloid vasculature to regress at birth [1]. Coagulating the vascular stalk with a plasma

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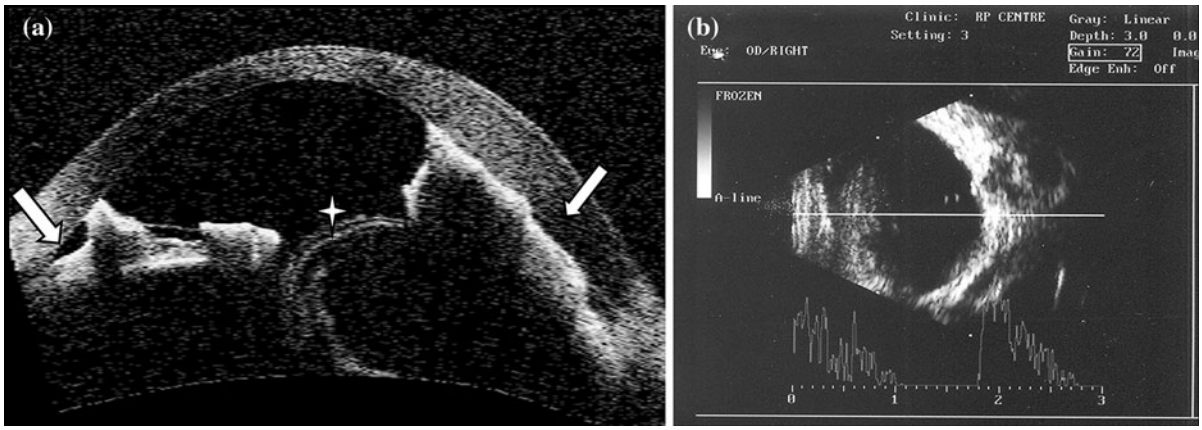

Fig. 1 **a** AS-OCT showing broad-based anterior mid-peripheral synechiae (*white arrows* showing clear peripheral zone with no synechiae), and outline of the subluxated lens (*asterisk*). **b** B-scan ultrasonography showing a thin membrane extending from the disc

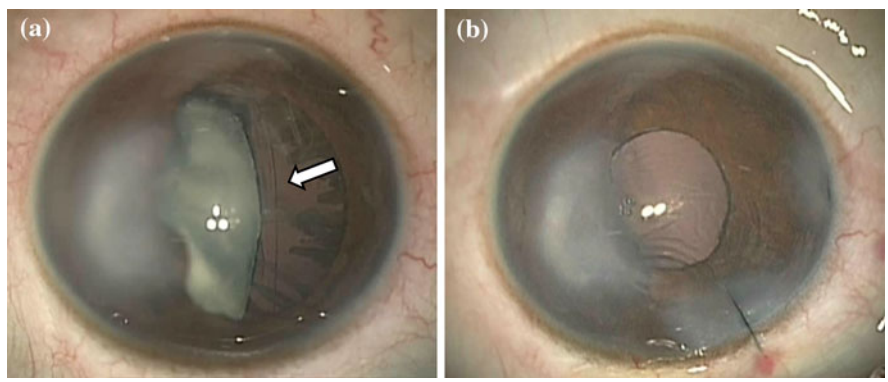

Fig. 2 **a** Intra-operative photograph of corneal opacity overlying iridocorneal adhesions, subluxated total cataract, stretched ciliary processes and visible bag outline (*arrow*). **b** Post-operative photograph depicting clear media

blade helps perform the surgery without risk of intra-operative hemorrhage [2]. Iris anomalies like striated and cryptless pattern, dilated vessels, and ectropion uveae are associated with PFV [3]. However the presence of mid-peripheral anterior synechiae akin to those observed in iridocorneal endothelial syndrome is unreported in PFV. Genetically, PFV has been associated with Axenfeld–Rieger syndrome and chromosomal 6p25 deletion [4] and digenic mutations in *PITX2* and *FOXC1* genes [5] have been found. Traction by the fibrovascular membrane on the posterior capsule probably caused lenticular subluxation, elongated ciliary processes and hypotony in our patient.

Thus, PFV may overlap with the clinical presentation of unilateral anterior segment dysgenesis even in the absence of family history of Axenfeld–Reiger or genetic association.

PFV may be under-diagnosed in cases presenting as atypical clinical picture like unilateral anterior segment dysgenesis. Careful ultrasonography should be performed along with biomicroscopy in a dilated pupil in suspected cases of PFV.

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