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LETTER TO THE EDITOR

Persistent hyperplastic primary vitreous (PHPV)

Hyperplasie et persistance du vitré primitif

A 14-year-old otherwise healthy boy presented with right-sided small eye with leukocoria since the age of four. At that time, his parents neglected these signs until the leukocoria bothered the patient who asked for treatment.

His visual acuity was reduced to hand motion on the right eye. External examination revealed a right esotropia with leukocoria. Slit-lamp examination of the right eye revealed small corneal diameters and a shallow anterior chamber (Fig. 1A). Furthermore, we noted a posterior synechia (Fig. 1A, white arrow), elongated and stretched ciliary processes (Fig. 1A, white star) with a white and partially resorbed cataract preventing funduscopy. The left eye was normal. Ocular B-scan and Doppler ultrasonography of the right eye showed a short axial length (19.7 mm, normal > 21 mm), a cataractous lens and an echogenic dense band extending from the lens to the optic disc (Fig. 1B). These clinical and ultrasound data were consistent with anterior form of PHPV. Our patient was scheduled for a lensectomy and anterior vitrectomy of the right eye.

PHPV is an ocular developmental disorder resulting from incomplete apoptosis of the embryonic hyaloids vasculature [1]. This malformation induces the growth of a retrolental fibrovascular mass, which is often accompanied by cataract and microphthalmia. Clinically, PHPV presents in several ways depending on the severity and extent of the involvement (anterior form, posterior form, combination of anterior and posterior forms). Strabismus, leukocoria, microphthalmia, microcornea, elongated ciliary processes (anterior form), retinal fold, papillary abnormalities and macular hypoplasia (posterior form) are the common clinical signs of PHPV. The diagnosis is confirmed by a B-scan and Doppler ultrasonography [2,3]. If left untreated, this condition can lead to angle-closure glaucoma, vitreous hemorrhage, retinal detachment and phthisis. Early surgical intervention combined with aggressive antiamblyopic therapy are critical to the final outcome [1].

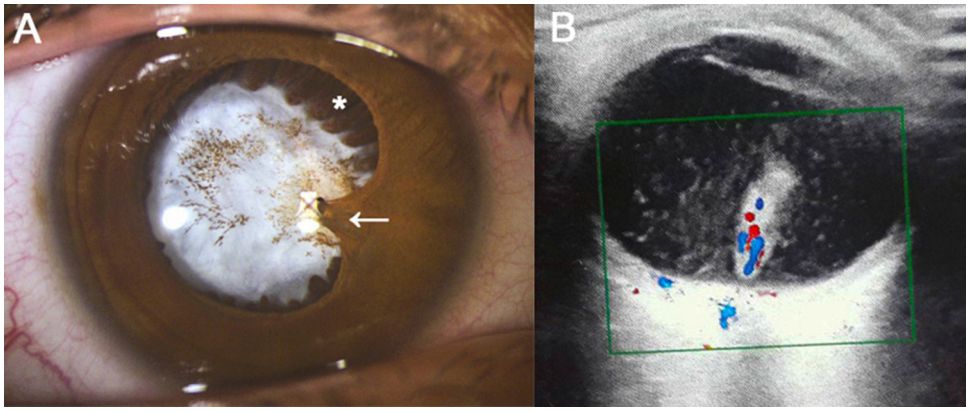

Figure 1. A. Slit-lamp image of an anterior form of PHPV showing a posterior synechia (white arrow), an elongated ciliary process drawn to the center (white star) and a white cataract. B. Ocular Doppler ultrasound showing a cataractous lens, vitreous echoes and a persistent hyaloid artery.

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Disclosure of interest

The authors declare that they have no competing interest.

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