Persistent Fetal Vasculature with Atypical Presentation

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Abstract
Persistent fetal vasculature is an uncommon condition, presenting clinically as Leukocoria (white pupillary reflex), micro-ophthalmia, and cataract. The authors report a case of an infant with atypical normal corneal diameter and long axial length sizes.

Keywords: Persistent Fetal Vasculature; Leukocoria; Micro-ophthalmia; Cataract

Introduction
Persistent fetal vasculature (PFV) is an uncommon condition of failure in ocular development due to incomplete regression of the fetal hyaloid vasculature. Here we report of a unique presentation of PFV.

Case Report
A 21-month-old boy presented to the Tel Aviv Sourasky Medical Center Pediatric Ophthalmic Unit due to suspected high myopia, unilateral right leukocoria and a new right eye strabismus. He was born at term following an uneventful pregnancy and birth and was otherwise healthy. Family history was unremarkable.

At initial visit he presented with signs of amblyopia in his right eye by fixating pattern, a moderate right esotropia (20PD) was diagnosed in the primary position. Pupil were equal and reactive to light, there was no palpable mass or swelling. He was systemically well with no other focal neurological signs on slit-lamp biomicroscopy examination a central posterior sub capsular cataract was seen (Figure 1). Due to the media opacity a fundus examination to the right eye was unsuitable. The left eye including the anterior and posterior part appeared to be normal except hypermetropia of 3.50 diopter (D).

As mentioned, since there was no clear view of the fundus, a B-scan ultrasound study was performed in which a combined PFV was found (Figure 2). Under anesthesia, the horizontal cornea diameter was 12 mm bilaterally (normal 10.00-12.00 mm), and the axial length of the left eye was 20.76 mm on the left eye and 23.55 mm on the right eye (average of 10 serial measurements with 0.10 mm standard deviation, OcuScan RxP, Alcon; normal values to this age, 19.70 - 23.00 mm1).

Lensectomy, posterior capsulotomy, core vitrectomy, removal of the remnant fibrovascular stalk and intracocular lens implantation of AcrySof 14.5 diopter lens were successfully performed

Discussion
PFV is a disorder of failure in ocular development due to incomplete regression of the fetal hyaloid vasculature [1,2]. It is usually divided into anterior, posterior or combined manifestation, and is characterized by the presence of a vascular stalk between the lens capsule and the optic disc [3]. The condition is usually unilateral and bilaterally is rare (< 3%) [4]. Anterior PFV is considered the most frequent cause of unilateral lens opacification,
and it is associated with both short axial length and reduced cornea size. In posterior PHPV is also associated with reduced cornea size and more than two thirds of cases are microphthalmic [5,6]. A grey-scale ultrasonographic study in an eye with PFV will show an echogenic band extending from the posterior surface of the lens capsule to the optic disc [1].

Descriptions of normal axial length and normal corneal diameter in PFV patients are sparse [7–9]. Most affected individuals have a mild form of the disease, and some of them do not have a classic leukocoria presentation and therefore tend to seek help later on in life [7–9]. The herein described classic young age presentation of PFV is unique for having an atypically longer than usual and then the second normal eye axial length and an atypically normal corneal diameter. To our knowledge this is the first report describing a unilateral case of classic presentation of PFV with a longer axial length in the affected eye.

References


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