

Congenital membranous cataract associated with persistent fetal vasculature

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Abstract

• A 15-year-old boy underwent phacoemulsification for a membranous cataract during which the presence of anterior persistent hyaloid artery and elongated ciliary processes were discovered. A posterior capsulorhexis was performed and the anterior part of the persistent hyaloid artery stalk was resected together with the posterior capsule. A foldable intraocular lens was implanted and the optic was captured in the posterior capsulorhexis margin. This is an unusual case of congenital membranous cataract associated with malformations of persistent fetal vasculature and elongated ciliary processes.

• **KEYWORDS:** congenital membranous cataract; persistent fetal vasculature; elongated ciliary processes

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CASE REPORT

A 15-year-old boy was referred to our hospital with blurry vision, strabismus and leukocoria of the right eye. The patient had always had poor vision in the right eye since birth without ocular trauma or surgical history, any family history or systemic disease. The pregnancy and delivery of the patient was normal. On examination, visual acuity was hand motion at 10cm in the right eye and 20/20 in the left eye.

Extraocular muscle movement examination showed 30° constant exotropia of the right eye. The corneal diameter was 12.0mm bilaterally. The keratometry was 45.06@156/46.30@60 and 41.41@15/42.08@105, and the axial length was 23.85mm and 23.93mm in the right and left eye, respectively. Corneal endothelial cell density was 3610cells/mm² in the right eye. The right eye slit lamp examination revealed a white fibrotic membrane opacification in the central and nasal peripheral of the lens (Figure 1A). The anterior chamber depth was normal. The

fundus could not be clearly viewed, the intraocular pressure (IOP) was 8.5mmHg, and a slightly posterior scleral staphyloma was detected by B-scan ultrasonic tomography (Figure 1B). These anterior and posterior segment abnormalities found in the right eye were not found in the left eye. Phacoemulsification was performed in the right eye under topical anesthesia. Corneal incisions were made, and a continuous curvilinear capsulorhexis was created in the intact anterior capsule under an ophthalmic viscosurgical device (sodium hyaluronate 1%, Healon GV). A flattened lens capsule with calcified cortex and a small amount of residual cortex in the peripheral part of the membranes was revealed. During irrigation and aspiration, the presence of anterior PFV and elongated ciliary processes were discovered, the persistent hyaloid artery appeared as vascular tissue extending from the posterior capsule behind the posterior capsule opacity to the vitreous cavity (Figure 1C). So a posterior capsulorhexis was performed and the anterior part of the hyaloid artery stalk was then resected together with the posterior capsule using intraocular scissors (Figure 1D). A +19.0D foldable intraocular lens (IOL, 3-piece AcrySof MA60BM, Alcon Laboratories Inc.) was implanted and the optic was captured in the capsulorhexis margin (Figure 1E).

Two weeks later, the right eye's visual acuity was counting fingers at 1 meter with the normal IOP, and IOL was well centered. Fundoscopy showed the normal retina and optic disc, and the remnant persistent hyaloid artery was apart from the visual axis (Figure 1F). During the 12-month follow-up examination, the right eye was quiet and vision remained stable at counting fingers, and no complications were found.

DISCUSSION

As a rare congenital disease, congenital membranous cataract is characterized by a collapsed, flattened capsule with little or no cortex or epithelium on the lens [1]. Müllner-Eidenböck *et al* [2,3] reported that varying degrees of persistent fetal vasculature (PFV) syndrome are a frequent cause of unilateral congenital cataracts. But a membranous cataract case was not found in their report of 31 eyes and only two membranous cataract cases were found in 62 cases reported by Haddad *et al* [4]. We describe an unusual case of unilateral congenital membranous cataract associated with PFV and elongated ciliary processes as well as its surgical management.

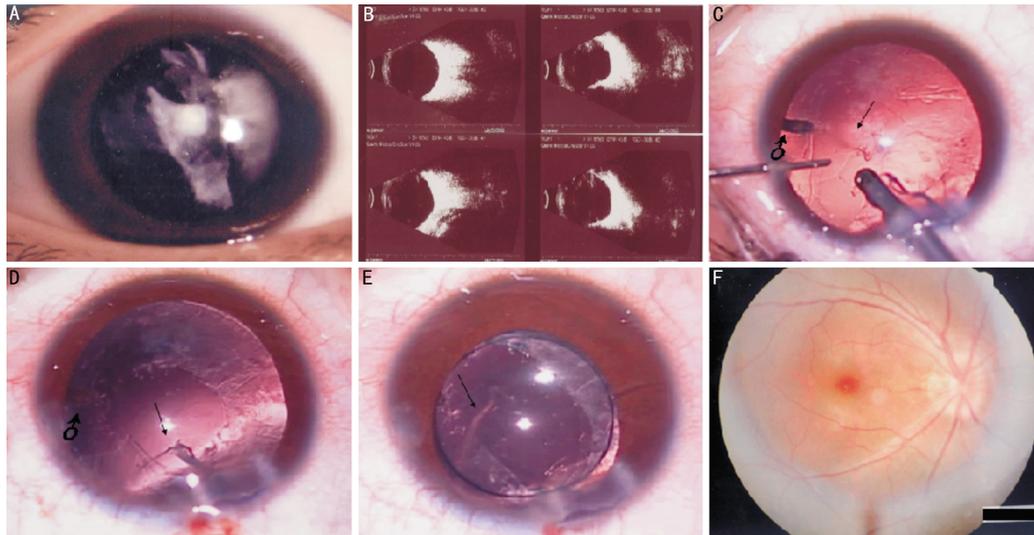


Figure 1 A: Slit-lamp photograph of a flattened lens capsule with a calcified cortex in the central and nasal peripheral of lens of the right eye; B: B-scan ultrasonic tomography of a slightly posterior scleral staphyloma; C: Surgery video showing the anterior persistent fetal vasculature (↘) and stalk extending ciliary processes (♂); D: Surgery video showing a posterior capsulorhexis and the resected persistent hyaloid artery tissue (↘); E: Surgery video showing that the IOL optic was captured in the capsulorhexis margin; F: Fundus photograph showing that the normal retina and the optic disc, and the remnant persistent hyaloid artery was apart from the visual axis

The PFV apparently plays a causative role in the pathogenesis of congenital cataracts, which occurs when the posterior/anterior pole of the lens is invaded by the fibrovascular membrane or by vessels of the tunica vasculosa lentis^[5,6]. However, the congenital membranous cataract is a rare type of congenital cataract, and its exact aetiology remains unknown. In our case, as without ocular trauma or surgical history, any family history or systemic disease, the membranous cataract maybe result from the persistent hyaloid artery and the tunica vasculosa lentis which may cause rupture of the lens capsule and further absorption of the cortex^[1,7,8].

Because of impossible full visualization due to membranous cataract and helpless result by B-scan ultrasonic tomography, preoperative diagnosis was a congenital membranous cataract, the diagnosis of the anterior PFV and elongated ciliary processes was based on the intraoperative findings. The posterior capsulorhexis with the IOL optic capture and removal of the fibrovascular tissue might allow a clear visual axis and prevent posterior capsule opacification^[9-15]. Although the final visual acuity was unsatisfactory due to amblyopia possibly caused by visual deprivation, the successful management result shows the technique described above is an effective method to remove the posterior capsule opacity and the anterior portion of the persistent hyaloid artery. In additional, our case shows that, PFV appears to play an important role in the pathogenesis of congenital membranous cataract, the surgeon must consider the possibility of the anterior PFV when congenital membranous cataract is noted and prepared for surgical management.

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