Where Did That Blood Come From? Atypical Presentation of Congenital Cataract With Hemorrhage

Basak BOSTANCI CERAN[1], Hasan ARIFOGLU[2], Gurcan VURAL[3], Emrullah TASINDI[4]

ABSTRACT

A 6-month-old baby was referred to our clinic for visually significant cataract in the left eye. Dens posterior lens opacity with hemorrhage was evident in the anterior segment evaluation. B-scan ultrasonography of the eye showed anechoic posterior segment. During the cataract surgery vascular remnants became evident after irrigation of the cloth in the anterior chamber. Histopathological examination of the vascular structure and cataract revealed fibrin formation, reactive lens epithelial hyperplasia and vascular endothelium, which were though to be remnants of tunica vasculosa lentis.

Keywords: Congenital cataract, Persistent fetal vasculature, Pediatric cataract surgery

INTRODUCTION

Persistent fetal vasculature (PFV), is a congenital ocular anomaly due to the incomplete regression of the embryonic hyaloid vascular system by the time of birth.[1] Diagnosis is usually made by observation or imaging of the persistent vascular organization. Ultrasonography (B-scan USG) may be helpful especially for cases in which the posterior segment is poorly visualized. Here we present a unique case of congenital cataract with PFV, where the remnants of the vascular system were clinically undetected before the surgery.

CASE REPORT

A 6-month-old boy presented with visually significant cataract in the left eye. Examination revealed dens posterior lens opacity with hemorrhage. (Figure 1) B scan ultrasonography was normal, showing anechoic posterior segment. (Figure 2)

Lensectomy and biopsy of the lenticular material was planned under anesthesia. During careful hydrodisection, the coagulated hemorrhage was washed out of the anterior chamber, revealing remnants of the vascular structure and a dense posterior plaque lying on the posterior lens

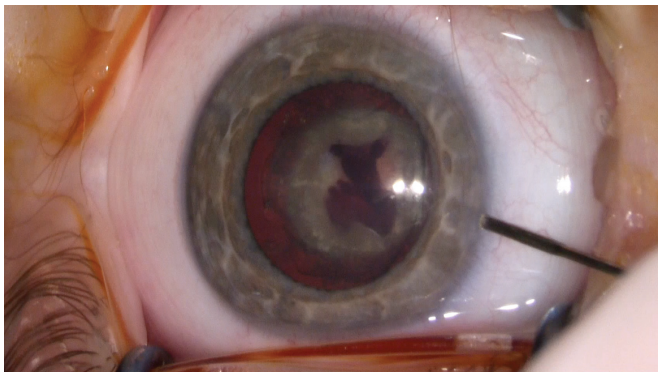

Figure 1. *Dens posterior lens opacity with hemorrhage.*

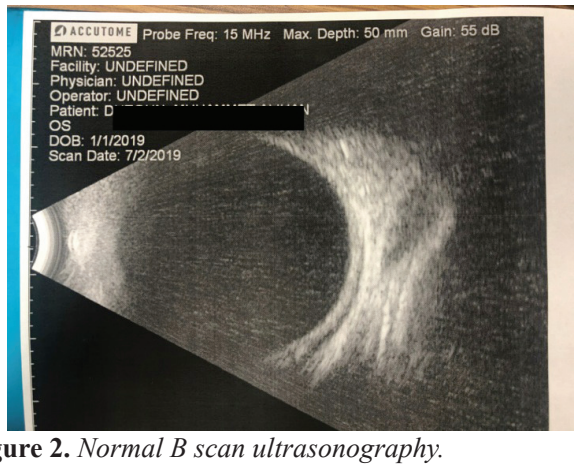

Figure 2. *Normal B scan ultrasonography.*

1- Assist. Prof., Okan University, Faculty of Medicine, Department of Ophthalmology, Istanbul, Turkey

2- Assoc. Prof., Okan University, Faculty of Medicine, Department of Ophthalmology, Istanbul, Turkey

3- Assist. Prof., Okan University, Faculty of Medicine, Department of Pathology, Istanbul, Turkey

4- Prof., Okan University, Faculty of Medicine, Department of Ophthalmology, Istanbul, Turkey

Received: 30.01.2020
Accepted: 05.05.2020
Glo-Kat 2020; 15: 194-196
DOİ: 10.37844/glauc.cat.2020.15.34

Correspondence Adress:
Basak BOSTANCI CERAN
Okan University Hospital, Aydınlı cad. No:2 İçmeler Tuzla, Istanbul, Turkey

Phone:
E-mail: drbbostanci@gmail.com

capsule. The lens was aspirated. The plaque was peeled off in a circular movement preserving the posterior capsule intact. Posterior capsulorrhexis and anterior vitrectomy was performed. Histopathological examination of the posterior capsular plaque (Figure 3a) revealed vascularized loose mesenchymal tissue with degenerated cystic space, hemorrhage and fibrin. (Figure 3b) Inflammatory cell infiltration including lymphocytes and macrophages and reactive lens hyperplasia of the lens epithelium are seen. (Figure 3c). Immunohistochemistry for CD31 (Leico Biosystems-Bond-Max) was done to demonstrate the endothelium of thin-walled vessels. (Figure 3d)

DISCUSSION

During the embryonal development, primary vitreous holds a vascular structure for nourishing the lens and anterior segment structures.[1] This hyaloid vascular structure which enters the optic cup from the dorsal ophthalmic artery forms in the fourth to fifth gestational week and proceeds anteriorly to the posterior pole of the lens. Hyaloid artery gives two brances; the vasa hyaloidea propria proceeding into the vitreous cavity; and the tunica vasculosa lentis which covers the surface of the lens.[2]

Persistent fetal vasculature is a congenital ocular anomaly, which occurs secondary to a disorder of this embryological hyaloid vasculature to regress when a child is born.[3]

Regression of the vascular structure is suggested to occur as a result of apoptosis and macrophage activation. The process begins with the smallest vessels and progresses to the larger ones. Occlusion of the hyaloid artery is the final step that occurs during the third trimester.[4,5]

The etiology of PFV is being under investigation. Defective apoptosis due to abnormal expression or abnormal timing for normal expression of genes going along with unsteady angiopoietin-2, VEGF, bFGF and growth factor levels have been proposed.[4,5]

Although most of the cases are sporadic, there have also been autosomal dominant and recessive inheritance patterns, which were reported in the past.[6]

Anterior, posterior and combined types are the three subtypes of the disorder. Existence of a retrolental opacity with lengthy ciliary processes or cataract are the common findings of the anterior sybtype. A vitreous membrane originating from the optic nerve as well as the formation of

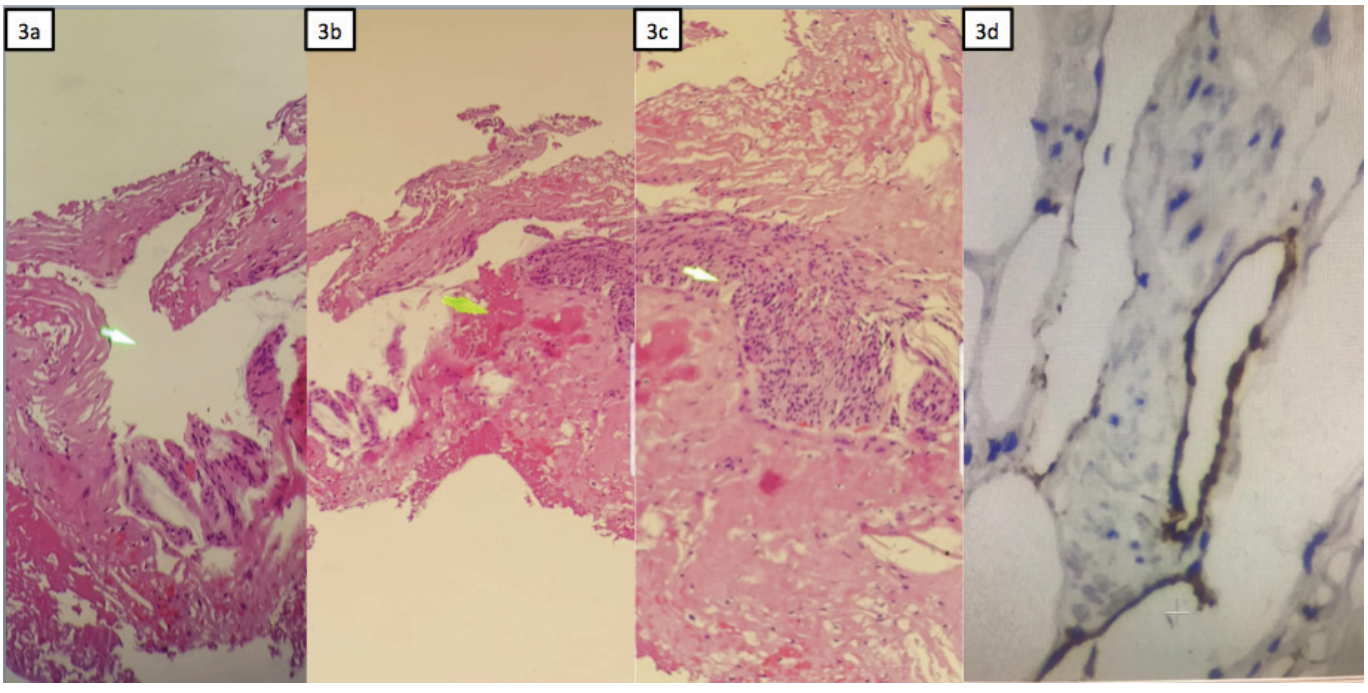

Figure 3a. *Full thickness tissue including loose mesenchymal areas with cystic degeneration, hemorrhage and epithelial hyperplasia (Haematoxylin and Eosine, magnification x40).*

Figure 3b. *High power view of loose mesenchymal areas with cystic degeneration.*

Figure 3c. *Inflammatory cell infiltration including lymphocytes and macrophages.*

Figure 3d. *High power view verifying CD31 immune study stains endothelial cells. (CD31 immunohistochemical staining, magnification x400).*

retinal folds, retinal detachment or optic nerve hypoplasia can be counted as some of the findings of the posterior subtype.[7]

A persistent pupillary membrane may form as a tunica vasculosa lentis remnant and cause pupil deformities, congenital ectropion or entropion in rare cases. If these vessels are still perfused, they also might may cause spontaneous hyphema.[8] According to our opinion, the vascular organization, which was visible anterior to the lens in our case, was the remnants of tunica vasculosa lentis.

Direct visualization of the persistent vascular remnant is usually the most common method for diagnosis. Ultrasonography is very useful especially when the posterior segment is poorly visualized. It is also used to rule out masses and retinal detachments in such cases. In rare cases like ours, B scan ultrasonography can be normal and show anechoic posterior segment. Computerized tomography with contrast may also be helpful for aiding the diagnosis.

When cataract surgery is planned in patients with suspected PFV, surgical decision is made according to preoperative and intraop status of the eye. If there is active perfusion of these vessels arising from the optic nerve to the lens, spontaneous intraoperative hemorrhage might occur. The flow of those vessels must be evaluated with ocular ultrasonography and eco-Doppler preoperatively. If blood flow is observed, endocoagulation instruments must be prepared to prevent massive intraocular hemorrhage.

Micro-scissors or micro forceps must be kept underhand since those might be needed for membranous cataracts or fibrovascular plaques to clear the visual axis.

Prognosis depends on the subtype of PFV and associated microphthalmia. Visual and functional results are associated with the age of the baby at the time of surgery and that of lens opacity occurrence, as well as existence of macular involvement. Results of early surgical intervention and anti-amblyopic therapy are often more desirable compared to the results of the eyes which were operated later and received unsatisfactory anti-amblyopic treatment.[9] In a study of Hunt et al, the cases who were diagnosed at 2.4 months average had a final visual acuity of 20/200 or better, while those presented at an average of 4.3 months were able to see 20/300 or worse.[7]

Possible postoperative complications include retinal detachment and phthisis bulbi leading to loss of vision. The rate of complications is reported to be higher in eyes with posterior retinal folds, combined PFV subtypes, microphthalmia, a persistent vascular flow and a large stalk.[10]

Parents of the PFV patient must be informed about the possible complications of cataract surgery in such eyes. In addition to that, importance of anti-amblyopic treatments and evaluation of the patients postoperatively are as important as the time of diagnosis and surgery itself . Our case is a unique form of persistant fetal vasculature with cataract, where only the anterior tunica vasculosa lentis was persistent. Although severe complications of cataract surgery are reported,we were managed to finish the case without any peroperative and post operative complication.

REFERENCES

1. Pau H. Hypothesis on the pathogenesis of retinopathy of prematurity – it is not VEGF alone but anatomical structures that are crucial. Graefes Arch Clin Exp Ophthalmol. 2010;248:1–3.
2. Lambert SR, et al. Congenital Fibrovascular Pupillary Membranes: Clinical and Histopathologic Findings. Ophthalmology 2012;119:634-64
3. Cero'n O, Lou PL, Kroll AJ, et al. The vitreo-retinal manifestations of persistent hyperplasic primary vitreous (PHPV) and their management. Int Ophthalmol Clin, 2008 48:53–62 (Review)
4. Goldberg, MF. Persistent Fetal Vasculature (PFV): An Integrated Interpretation of Signs and Symptoms Associated with Persistent Hyperplastic Primary Vitreous (PHPV) LIV Edward Jackson Memorial Lecture. Am J Ophthal 1997;124:587-626.
5. Saint-Geniez M, D'Amore P. Development and Pathology of the Hyaloid, Choroidal, and Retinal Vasculature. Int J Dev Biol 2004;48:1045-58.
6. Shastry BS. Persistent hyperplastic primary vitreous: congenital malformation of the eye. Clin Experiment Ophthalmol. 2009; 37:884–90.
7. Hunt A, Rowe N, Lam A, et al. Outcomes in persistent hyperplastic primary vitreous. Br J Ophthalmol 2005;89:859-63.
8. Sisk RA, Berrocal AM, Feuer WJ, et al. Visual and anatomic outcomes with or without surgery in persistent fetal vasculature. Ophthalmology. 2010;117:2178-83.e1-2.
9. Anteby I, Cohen E, Karshai I, et al. Unilateral persistent hyperplastic primary vitreous: course and outcome. J AAPOS. 2002;6:92-9
10. Fortes Filho JB, Tartarella MB, Takahagi RU, et al. Persistent fetal vasculature: ocular features, management of cataract and outcomes. Arquivos Brasileiros de Oftalmologia. 2013 19;76.