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Case Report

Persistent Fetal Vasculature Detected by Prenatal Ultrasound

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Abstract

Ultrasound is usually performed during pregnancy in order to assess fetal anatomy. We report a case of a newborn patient with unilateral congenital cataract diagnosed by prenatal ultrasound at 22 weeks' gestation. Postnatal ophthalmic B-scan ultrasound and doppler echography disclosed a small, spherical lens, with cataractous changes, and a tubular stalk of fibrovascular tissue attached to the posterior surface of the opacified lens, connecting to the optic nerve, consistent with persistent fetal vasculature (PFV) in the right eye. Left eye was otherwise normal. After born, the patient was submitted to surgery by the 12th week of life. Twenty-five-gauge sutureless lensectomy technique was performed through the pars plicata approach. The lens nucleus and cortex were aspirated in the bag. Anterior vitrectomy and removal of the PFV stalk was performed. At follow up, by 26 months of age, the best corrected visual acuity was 20/100 in the affected eye. Prenatal ultrasound with accurate evaluation and analysis of the eyes can provide early ophthalmic diagnosis and early referral, providing a prompt surgical approach soon after birth in order to avoid amblyopia and childhood blindness.

Keywords: Congenital Cataract; Persistent Fetal Vasculature; PFV; Prenatal Ultrasound

Abbreviations

PFV: Persistent Fetal Vasculature; RE: Right Eye; LE: Left Eye

Case Report

A 36-year-old pregnant woman was referred at 22 weeks' gestation for routine obstetric ultrasound evaluation. During pregnancy she developed idiopathic thrombocytopenic purpura and needed treatment with steroids. The pregnancy had been spontaneously conceived. The intrautero ultrasound performed at 22 weeks' gestation disclosed a unilateral congenital cataract in the fetus right eye (RE) and a dense cone behind the lens (Figures 1 Left and Right) without other congenital anomalies identified in the fellow eye or elsewhere. There was no consanguinity and no history of retinoblastoma in the family, but due to the ultrasonographic findings, differential diagnosis of retinoblastoma, as well as Persistent Fetal Vasculature (PFV), must be considered. Parents were aware and the mother was referred for ophthalmic consultation.

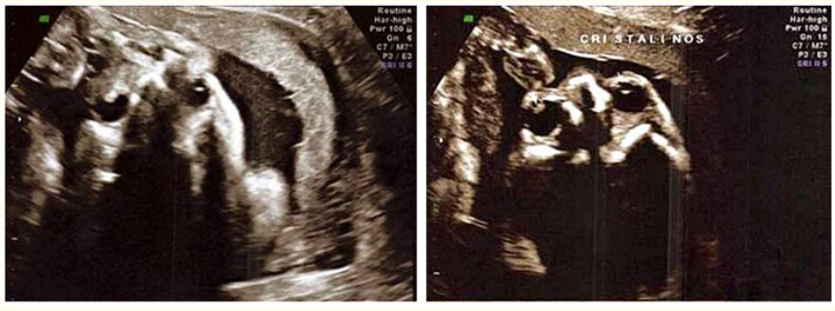

Figure 1: Left: Obstetric intrauterine ultrasonographic imaging of the fetus at 22 weeks' gestation. Figure 1 Right: A coronal ultrasound at 22 weeks' gestation detecting congenital cataract in the fetus's right eye (the arrow is pointing to the cataractous lens.

The patient, a white girl, was born at 36 weeks gestational age weighting 2,800grams. Leukocoria in RE was present at birth. At external observation, the RE was smaller than the left eye (LE). One week after birth the patient underwent her first ophthalmological evaluation. Red reflex was absent in the RE. At slit lamp evaluation it was detected a total white cataract and shallow anterior chamber. Intraocular pressure was 8 mmHg in the affected eye. Biometry disclosed an asymmetric axial length as 16.65 mm for the RE and 17.65 mm for the LE.

Ophthalmic ultrasound B-scan and doppler ecography of the RE, conducted with Philips's equipment with a linear transductor of 5-12 MHz, revealed small, spherical lens with cataractous changes and a stalk of fibrous tissue extending from the optic nerve, inserting as a white mass at the posterior lens capsule, consistent with a combined form of PFV (Figures 2 Left and Right, and Figure 3). Color Doppler imaging disclosed no blood flow inside the fibrovascular stalk. No retinal detachment was detected. Left eye was otherwise normal.

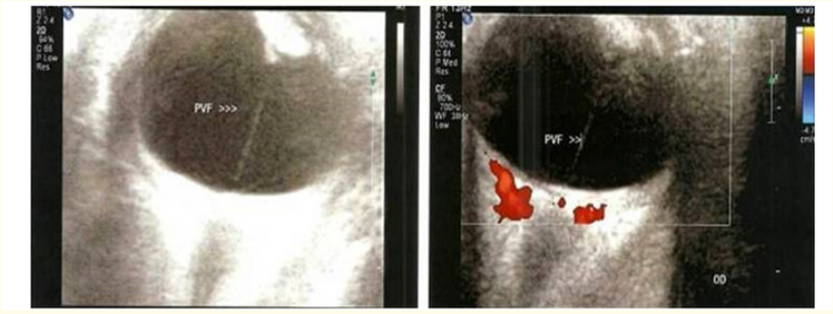

Figure 2: Left: Postnatal B-scan ultrasound demonstrating a tubular stalk of fibrous tissue extending from the optic disk toward the opacified lens, as seen in PFV. Figure 2 Right: Color Echo-Doppler imaging disclosed blood flow in choroidal and retinal vessels but disclosed no blood flow inside the stalk.

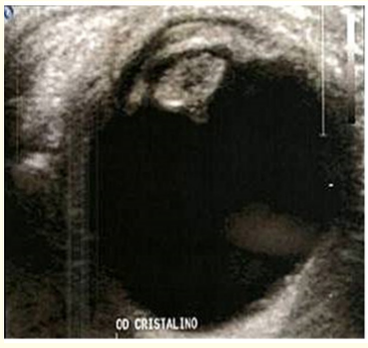

Figure 3: Ophthalmic B-scan ultrasound performed one week after birth detected congenital cataract and a small cone behind the lens in the right eye.

Surgical treatment was planned to be performed at the 7th week of life, but surgery had to be postponed due to the occurrence of nasolacrimal duct obstruction, and local treatment was performed. The patient underwent cataract surgery by the 12th week of life. Twenty-five-gauge sutureless lensectomy with bimanual vitrectomy technique was performed through the pars plicata approach. The opaque lens nucleus and cortex were aspirated in the bag. Posterior capsulorhexis, anterior vitrectomy, fibrovascular stalk section and management, and anterior capsulotomy were performed via pars plicata.

Contact lens fitting and partial time occlusion of the LE were prescribed with good compliance of the patient and family. Secondary opacification of visual axis occurred four months after initial procedure. An additional anterior vitrectomy was performed to clear the visual axis.

At last follow up, at 26 months of age, the best corrected visual acuity was 20/100 in the affected eye and 20/20 in the LE.

Discussion

Early detection of ophthalmic pathology is important in prenatal and in postnatal management decisions [1]. In the related case, a pregnant woman was referred to the ophthalmologist at 22 weeks' gestation due to the fetal ultrasonographic abnormalities detected in the RE without other congenital anomalies identified in body, brain, and in the fellow eye.

The prenatal ultrasonographic imaging of the RE showed an opaque lens and a dense opaque cone behind the lens. Differential diagnosis with intraocular tumor as retinoblastoma is very important in obstetric decision making. Due to the ultrasonographic characteristics of the reported case, the images detecting opacity in the lens, anterior portion of the eye and posterior face of the lens, retinoblastoma and PFV must be considered as differential diagnosis. Although ultrasound is unable to provide definitive diagnosis, the increasing sensitivity in detecting ocular abnormalities in utero can provide to physicians and expectant parents more time to make informed decisions, effective treatment planning, and early ophthalmologic referrals. The baby born premature and one week after birth the postnatal ocular eco-doppler ultrasonography disclosed cataract associated with PFV.

In this case, early surgical intervention was necessary. Congenital cataract is an amblyogenic condition and PFV can cause secondary glaucoma; intraocular hemorrhages; and synechiae may ensue [1,2]. The patient underwent a 25G sutureless pars plicata vitrectomy and lensectomy at 12 weeks of life. The surgical technique was previously described [3].

Persistent fetal vasculature is a congenital developmental anomaly of the eye resulting from failure of the embryological primary vitreous and hyaloid vasculature to regress that typically presents unilaterally without association with systemic findings or may be associated with rare systemic syndromes. Bilateral cases of PFV account for less than 10% of the cases, and it can be classified as anterior, posterior and combined forms, according to the affected intraocular structures [4,5]. The patient here reported presented the combined form of PFV.

The heterogeneity of clinical presentation makes PFV a challenge to surgical management. A novel classification of PFV based on high-resolution B-mode ultrasound and color doppler imaging was proposed by Hu *et al* in order to help in the planning of surgical treatment [5]. Recent advances in surgical instrumentation and techniques have changed the indications for PFV surgery [3,5].

Cataract surgery in patients with PFV needs retinal surgery skills and is related to higher rates of postoperative complications as: retinal detachment, hyphema, intraocular hemorrhage, glaucoma, secondary opacification of the visual axis and extensive inflammatory response with pupillary block [6-9]. Initial surgical planning may alter depending on the pre and intraoperative ocular conditions. Ocular eco-doppler findings detecting blood flow from the optic nerve to the posterior face of the lens is a predictive factor of intraoperative hemorrhage. In some cases, endo-diathermy of the blood vessels is necessary. Intraocular hemorrhage is a common cause of bad prognosis and poor outcomes. High-resolution intrauterine ultrasound expedited ophthalmic referral soon after birth [1,9,10].

Eyes with PFV and cataract may be associated with variable degrees of microphthalmia that could interfere in the visual improvement. Early surgical intervention with microsurgical 25G lensectomy technique combined with anti-amblyopic therapy may result in favorable visual outcomes. Further studies concerning cumulative factors that could predict surgical prognosis for PFV cases are necessary [2].

Conclusions

The case related achieved good anatomical and functional outcomes after surgery. Visual rehabilitation and occlusion therapy were mandatory. Prenatal ultrasound can help to early detect fetal ocular malformation, congenital cataract, microphthalmia, intraocular tumors (retinoblastoma) and PVF. High-resolution intrauterine ultrasound expedited ophthalmic referral soon after birth and influenced postnatal ophthalmological surgical planning helping to avoid amblyopia and blindness.

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The authors certify that the content has not been published or submitted for publication elsewhere. Authors also certify that the protocol for the research project has been approved by a suitably constituted Ethics Committee of the institution within which the work was undertaken, and that it conforms to the provisions of the Declaration of Helsinki in 1995 (as revised in Edinburgh 2000).

Declaration of Patient Consent

The authors certify that they have obtained appropriate patient consent form. In the form the patient has given his consent for his images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity

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Conflictis of Intrerest

There are no conflicts of interest.

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