Stunning Visualization of Fetal Hyaloid Artery

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Persistent hyperplastic primary vitreous (PHPV), also known as persistent fetal vasculature (PFV), is of considerable importance when evaluating congenital anomalies in infants and young children.[1] PFV results from a failure of the fetal hyaloid vasculature to involute.[2] This condition is typically unilateral.[2] Bilateral cases can be associated with systemic abnormalities or could represent overlooked similar conditions such as familial exudative vitreoretinopathy (FEVR), bilateral retinal folds, or Norrie disease. PFV can be divided into three types: anterior (persistent tunica vasculosa lentis), posterior (falciform retinal septum), or a combination of anterior and posterior.[2] Most patients with PFV have a combination of both anterior and posterior types.[2]

We report a case of PFV in a 12-month-old infant who was referred to our clinic because of a pigmented lesion at the optic disc.

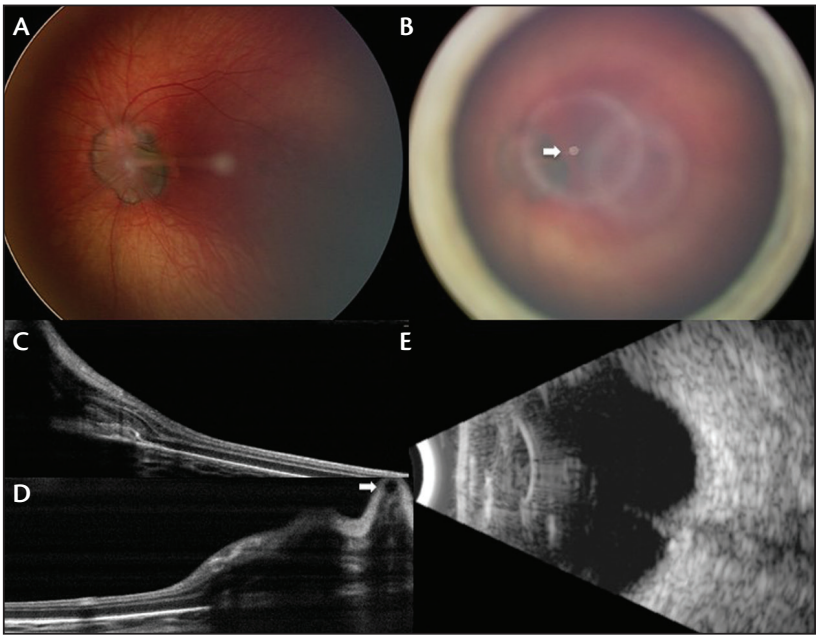

Figure 1. Persistent fetal vasculature (PFV) in the left eye of a 12-month-old female. Fundus image of the left eye showing fibrovascular stalk of persistent hyaloid artery toward the lens (A). The retina around the optic nerve was drawn up into the stalk, leading to a retinal pigment epithelial ring around the base. The disc was intact as seen through the dragged retina. Mittendorf dot from attachment of PFV to posterior lens capsule (B, white arrow). OCT horizontal cut through the optic nerve toward the foveola displaying retinal elevation and distortion (C). OCT vertical cut through the optic nerve displaying elevation of the fibrovascular stalk (white arrow) (D). Ultrasound showing fibrovascular stalk at disc region and thin hyaloid artery through the vitreous cavity (E).

CASE REPORT

A 12-month-old white female was referred to Wills Eye Hospital for evaluation of a pigmented lesion in her left eye. The patient was born full term without complications. A cutaneous capillary hemangioma of infancy was present at birth in the nasal region. With the exception of intermittent left exotropia beginning at age 3 months, there was no previous ocular history or trauma. Family history was unremarkable.

At the time of the ophthalmic examination, a left exotropia (40 ΔD), mild left hypertropia (15 ΔD), and a regressed cutaneous hemangioma on the left naris were observed. Visual acuity was fix and follow with the right eye, but the patient was unable to fix or follow with the left eye. Axial length was 21.5 mm in the right eye and 20.8 mm in the left.

Fundoscopic examination of the right eye was normal. Examination of the left eye revealed persistence of the hyaloid artery as a stalk of fibrovascular tissue extending from the optic nerve to the posterior lens capsule. The retina around the optic nerve was drawn up into the stalk in the prepapillary region as a Bergmeister papilla,

causing retinal dragging toward the disc. The disc, visualized through the dragged retina, appeared intact (Figure 1A). A Mittendorf dot, the anterior attachment of the hyaloid artery, was observed at the nasal part of the posterior lens capsule as a small, circular opacity (Figure 1B). Optical coherence tomography (OCT) showed an elevated retina drawn into the vitreous, and the foveola was not visible (Figure 1C-D). Ocular ultrasonography disclosed a fibrovascular stalk at the disc region and a thin hyaloid artery visible through the vitreous cavity (Figure 1E).

Fluorescein angiography (FA) of the right eye revealed a patent hyaloid artery within the fibrovascular stalk, documented with initial filling of the vessel in the full venous phase and completed filling in the recirculation phase (Figure 2A-B). FA demonstrated staining of the tissue encircling the optic disc with a delineating blocking effect of retinal pigment epithelium at the border of retinal elevation. There was peripheral nonperfusion temporally, likely from the posterior retinal dragging (Figure 2D). There was no tunica vasculosa lentis, neovascularization, or vitreous hemorrhage. Surgical treatment was not performed because of the disrupted foveal architecture.

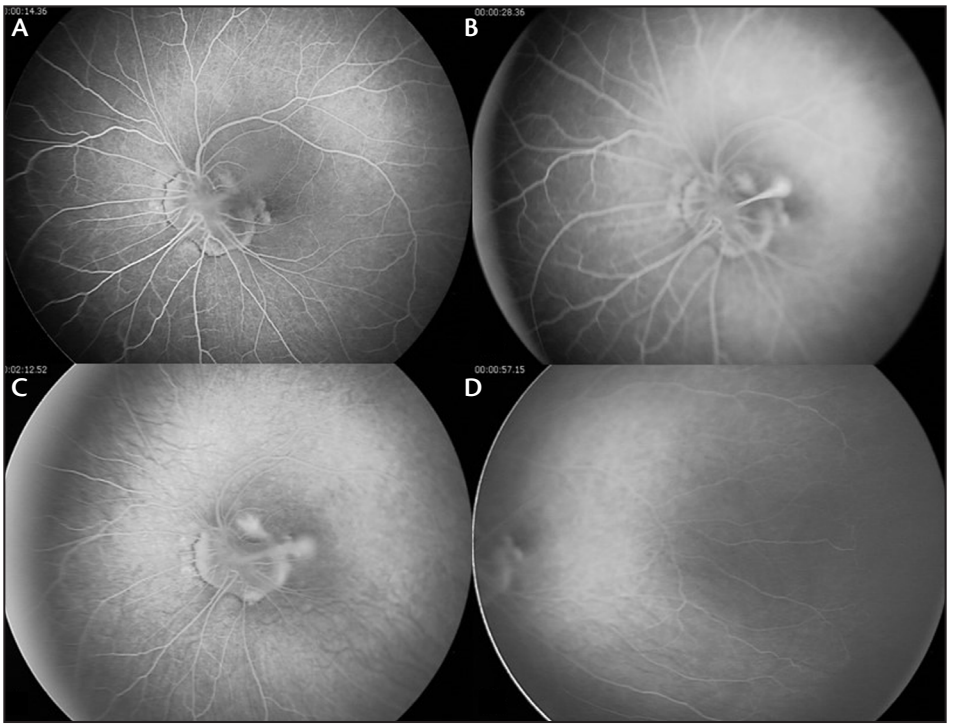

Figure 2. Fluorescein angiography (FA) of PFV in the left eye of a 12-month-old. Full venous phase of the FA demonstrating initial filling at the base of the hyaloid artery (A). Later phase of the FA showing filling of the entire hyaloid artery and staining of the juxtapapillary tissue area, with delineating blocking effect of retinal pigment epithelium at the border of retinal elevation (B). Recirculation phase of FA of the retina, showing increased staining at the margin of the scar tissue (C). Temporal peripheral nonperfusion was evident (D).

DISCUSSION

PFV is a spectrum of congenital ocular malformations, often resulting in vitreoretinal fibrosis with grey-white scar tissue. This condition is an important simulator of retinoblastoma, a malignant intraocular tumor of children.[1-4] Persistence of components of the fetal intraocular vasculature from the optic disc up to and including the lens have been documented.[1]

In fetal life, the hyaloid artery provides nutrition to the developing lens. The primary vitreous, containing branches of the hyaloid artery, is formed during the first month of fetal development. The hyaloid artery begins to regress during the formation of the avascular secondary vitreous at 9 weeks. During the third trimester, the hyaloid artery undergoes involution with regression of the Bergmeister papilla and tunica vasculosa lentis.[2] However, if the mechanisms that normally inhibit vascular growth or maintenance are suppressed, certain vessels can persist. Hyaloid vascular remnants are observed in more than 90% of infants born at fewer than 36 weeks gestation and more than 95% of infants weighing less than 5 pounds at birth.[1,2]

Clinical features of PFV can vary from mild to severe. Mild cases include those with a Mittendorf dot, Bergmeister papillae, faintly visible persistent hyaloid artery that may be patent or closed, and/or remnants of tunica vasculosa lentis. Severe cases can include tractional retinal detachment, centrally dragged ciliary body pars plicata processes, retrolenticular fibrosis, cataract, shallowing of the anterior chamber, elevated intraocular pressure, pain, and/or phthisis bulbi. Rarely, patients with PFV can experience mild to severe vitreous hemorrhage, which can prohibit further visualization of the retina. It

is postulated that the vitreous hemorrhage is due to the tractional force of eye movement, which can lead to rupture of the unsupported hyaloid artery.[5]

PFV should be differentiated from other similar conditions including retinoblastoma, FEVR, retinopathy of prematurity, Norrie disease, incontinentia pigmenti, and retinal detachment from other causes. Unlike PFV, retinoblastoma rarely produces a microphthalmic eye unless there is phthisis bulbi. Ultrasonography is useful in evaluating intraocular calcification in children with retinoblastoma.[6] FEVR, Norrie disease, and incontinentia pigmenti are bilateral conditions, whereas PFV is almost always unilateral. Furthermore, those conditions show no stalk but rather can lead to retinal dragging and occasionally retinal folding.[7]

In our patient, areas of mild peripheral retinal nonperfusion and looping of retinal vessels were seen on fluorescein angiography in the affected eye (Figure 2D). The vascular maldevelopment and secondary retinal dragging into the stalk likely led to the peripheral nonperfusion.[9] In other conditions such as retinopathy of prematurity, FEVR, and Norrie disease, defective retinal angiogenesis leads to ischemic peripheral nonperfusion.[6] PFV, however, is an abnormality of hyaloid vasculature regression, and the peripheral nonperfusion is likely due to retinal dragging and not true ischemia.[9,10] These mild peripheral abnormalities generally do not require laser photocoagulation.

Visual acuity is usually reduced in PFV because of primary and secondary effects of the persistent hyaloid system on the developing macula and optic nerve.[1] Reasons for reduced vision can include structural problems such as lens opacity, vitreous hemorrhage, retinal dragging or tractional retinal detachment, microphthalmia, secondary glaucoma, phthisis bulbi, or refractive problems such as refractive error and amblyopia. Treatment of PFV depends on the pathogenesis of vision loss and on the anticipated visual outcome. Surgical intervention can include coagulation of the hyaloid artery, repair of the retinal detachment, or extraction of the lens. Following repair, amblyopia therapy should be considered.[1,8,11] Many cases, particularly those with intact visual acuity or those at the opposite extreme with complex tractional detachment and disorganized retina, are managed conservatively with observation.[11]

CONCLUSION

When evaluating infants with leukocoria, the clinician should consider PFV in the differential diagnosis. Overlooked cases could suffer debilitating loss of vision, eye pain, and possible loss of the eye. There are a variety of diagnostic modalities that can aid in the detection of PFV, including FA to delineate the hyaloid artery and tunica vasculosa lentis, OCT to show vitreoretinal traction, and ultrasonography to rule out retinoblastoma. Early detection using these modalities could assist in protection of the globe and vision in patients with PFV. ■

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