Persistent Fetal Vasculature: The Importance of Early Detection and Intervention

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Abstract:
Children born with persistent fetal vasculature generally have a poor visual prognosis. This report highlights the importance of early detection and timeliness of intervention in a child initially referred for strabismus.

A. Case History:
Pediatrician requested consultation of 7 month old male for suspected strabismus. A full term pregnancy and uncomplicated delivery was reported with no health concerns at birth and no family history of strabismus or amblyopia. Ocular misalignment of left eye was previously noted by the mother but was dismissed as pseudostrabismus.

B. Pertinent Findings:
Acuity/Fixation: Central, Steady, Maintained OD; Not Central, Steady, Not Maintained OS
Alignment: 25 Δ LET° per modified Krimsky
Anterior Segment: prominent epicanthal folds OU; corneal diameter: 11mm OD, 10mm OS; leukocoria OS: obstructing visual axis
Fundus Exam: stalk extending from optic nerve head to posterior lens capsule OS

C. Differential Diagnosis:
Pseudostrabismus, infantile esotropia, congenital cataract, retinoblastoma, Coat’s disease, toxocariasis, toxoplasmosis, retinal/choroidal/optic nerve coloboma, myelinated nerve fibers, Norrie’s disease, retinal dysplasia, cyclitic membrane, retinal detachment, incontinentia pigmenti, retinoschisis, and medulloepithelioma (Friedman & Kaiser, 2009)

D. Diagnosis and Discussion:
Diagnosis included persistent fetal vasculature (PFV) associated with microcornea, central retrolental plaque, sensory esotropia and deprivation amblyopia, all affecting the left eye. A tractional retinal detachment with absence of the macula was subsequently diagnosed during surgery. PFV is caused by the failed regression of the primary vitreous during the 3rd and 9th month of gestation (Shastry, 2009). It is a sporadic malformation of the eye that most commonly involves the anterior and posterior segment, and occurs unilaterally 90% of the time (Taylor and Hoyt, 2009). Visual prognosis worsens in cases of posterior segment involvement and surgical invention within the first 3 months yields the greatest visual potential (Surbert & Gurwood, 2000). In absence of timely treatment, retinal detachment, ciliary body detachment, intraocular hemorrhages, and angle closure glaucoma can occur. Typical management includes surgical intervention to release tractional components and clear the optical path for light (ex: removal of central plaque, cataract, and/or stalk), optical rehabilitation with contact lens, and aggressive management of concomitant strabismus and amblyopia. Frequent monitoring for glaucoma and retinal detachments is also required.

E. Treatment and Management
Occlusive patch therapy was initiated at the time of diagnosis (1 hour per day, right eye) and surgical intervention was recommended with guarded prognosis for visual recovery. B-scan ultrasound initially revealed an intact retina, but this was found not to be the case during surgery. Pars plana lensectomy, vitrectomy and capsulotomy were performed 26 days following diagnosis. A contact lens fitting took place 8 days after this and dispensed a week later at 15 days postsurgery. Occlusive patch therapy was re-initiated 22 days post-surgery and increased to 2 hours per day.
F. Conclusion:
This case highlights the importance of early detection of sight threatening conditions in the pediatric population, and provides an example as to the potential significance of early onset strabismus in an otherwise healthy child. Recent literature indicates that 20/125 vision or better can be achieved in PFV patients if surgical intervention takes place prior to 6 weeks of age followed by compliant contact lens wear and amblyopia therapy (Chen et al. 2010). In the present case, the span of time between initial diagnosis and contact lens dispensal was 41 days, this in addition to the 7 month span that ocular misalignment was present but not addressed. Unfortunately for this child, 20/400 to counting fingers can be hoped for as the best visual outcome, a further example as to the importance of due diligence in recognizing and promptly addressing potential cause of permanent vision loss in children.