An 11-Year-Old Male with History of Premature Birth Referred for Lattice Degeneration

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Introduction:

An 11 year-old male was referred for evaluation of bilateral lattice and retinal holes. He was born at gestation age of 27 weeks with a birth weight of 539 gm. His past medical history was significant for an extended NICU stay requiring oxygen supplementation and multiple gastric surgeries. He did not carry a diagnosis of previous or treated for retinopathy of prematurity (ROP).

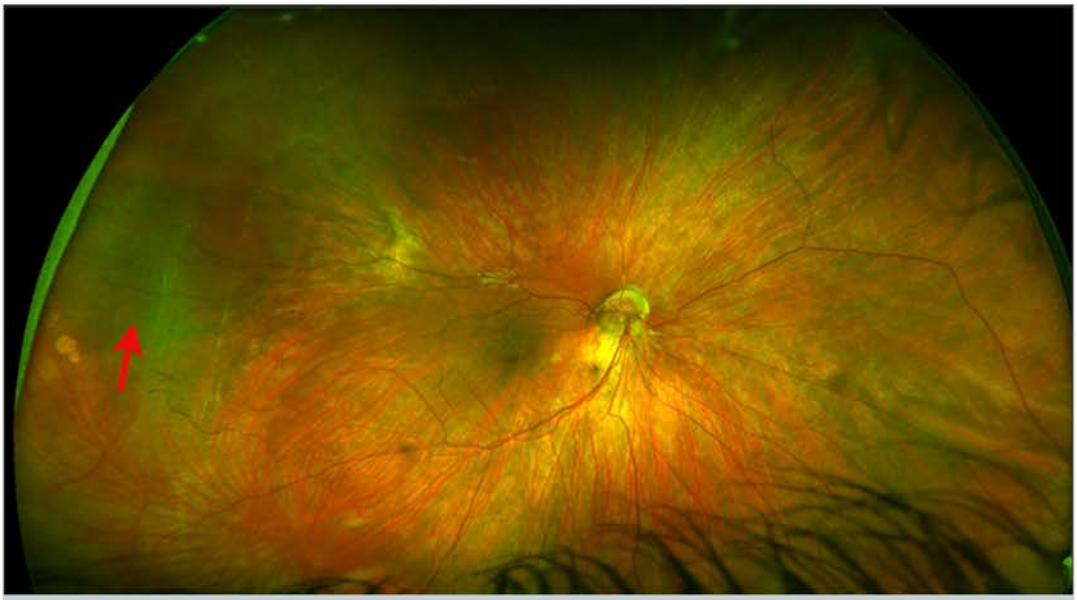

Figure A: Widefield scanning laser photo of the right eye revealing temporal dragging of the macula and temporal vasculature, and tractional retinal detachment (red arrow).

Exam:

Best corrected visual acuity was 20/80 in the right eye and 20/200 in the left eye. Intraocular pressures, visual fields, and extraocular motilities were normal. No afferent pupillary defect was present. Nystagmus was present in both eyes. Anterior segment exam was normal.

Dilated fundus examination revealed temporal dragging of the macula and vasculature of the right eye (Figure A). Pigmentary changes and tractional retinal detachments in the temporal periphery were present in each eye (Figure A & B). Exudate or abrupt termination of retinal vessels was not seen.

Discussion

The differential diagnosis of these findings includes ROP, familial exudative vitreoretinopathy (FEVR), Norrie disease, Coats disease, incontinentia pigmenti (in a female patient), and persistent fetal vasculature (PFV). In FEVR, the patient's findings can range from only peripheral non-perfusion (often in a V shape with the apex towards the maula) to temporal dragging of the macula and vasculature, vitreous opacities, exudate, abrupt termination of vessels, macular edema, radial retinal folds, and recurrent hemorrhages. These patients typically will have a normal birth history and positive family history although affected relatives may be asymptomatic.[1] Findings in Coats disease include male sex predominance, and vascular changes that can be unilateral although often can be highly clinically asymmetric with fellow eye involvement documented with fluorescein angiography. Retinal telangiectasias are seen in the mid-peripheral to peripheral fundus with associated exudation.[2] In Norrie Disease, the retinal is bilaterally severely dysplastic with a pseudoglioma

appearance, retrolental fibrovascular tissue, and a residual hyaloidal stalk connecting them.[3] Findings of PFV include persistent pupillary membranes, incomplete regression of the hyaloid artery, retrolental membranes, hyaloid stalk, and attached retinal traction or detachment.[4] Incontinentia pigmenti can present with skin lesions, teeth defects, peripheral retinal avascularity, and macular occlusive disease in female patients.[5]

Figure B: Widefield scanning laser photo of the left eye revealing peripheral retinal lattice-like pigmentary degeneration (small red arrow) and tractional retinal detachment (large red arrow).

Fluorescein angiography is a helpful tool to differentiate the underlying condition. Symptomatic patients with FEVR exhibit peripheral non-perfusion and telangiectasias in the macula and vascular-avascular junction. Over 50% of asymptomatic family members will have peripheral vascular anomalies.[1] Leakage is indicative of active progressive disease. Patients with Coats disease may exhibit unilateral telangiectatic vessels, "light bulb" aneurysms, capillary nonperfusion, and leakage. Mild or involutional findings may also be seen in the fellow eye.[2] Peripheral non-perfusion, hyperfluorescent hyaloidal stalk, or popcorn hyperflourescence may be seen in eyes affected by PCV and 90% will show peripheral non-perfusion in the fellow eye.[6] ROP findings include abnormal vascular patterns, vascular-avascular junctions, shunts, and neovascular nets. These findings are present regardless of if the patient was treated with anti-VEGF therapy or not.[7]

Our patient's birth history, lack of familial inherited condition, and bilateral findings support ROP. Several signs of the sequalae of ROP are present. These include temporal dragging of the macula and vasculature, retinal pigmentary changes, lattice-like degeneration, myopia, amblyopia, and tractional retinal detachments. Additional signs may include cataract, glaucoma, retinal folds, retinal breaks, and rhegmatogenous and exudative retinal detachments. Late term findings are variable depending on the severity of ROP. Those with Stage 1 ROP may exhibit minimal no findings later in life whereas those with a history of Stage 5 ROP may have phthisis.

Studies by Smith, Kaiser, and colleagues have shown a higher-than-normal likelihood of retinal pathology long into adulthood in patients with a history of ROP. Retinal dragging occurs in 34%. 14-26% experience retinal detachments, 8-11% have retinal tears, 9-11% have lattice degeneration, and 7-8% have retinal folds.[8,9] Even normal appearing eyes may be at risk with retinal tears observed in eyes with minimal cicatricial changes.[10]

In a study comparing patient characteristics of retinal detachments in a pediatric referral practice, ROP accounted for the highest percentage of TRDs at 16.8%.[11] ROP associated TRDs also showed the lowest surgical (PPV +/- SB) primary success rate of 33% and a high rate of PVR of 77%.[11] Thankfully, most TRDs without a retinal tear show long-term stability and may be closely observed if there is no threat to the macula. With regard to rhegmatogenous retinal detachments, these can occur at any time in life and the patient must be warned of detachment symptoms. Rhegmatogenous retinal detachments (RRD) require surgical intervention and like ROP associated TRDs, exhibit a higher primary-repair failure rate. Review of several studies shows mixed results in terms of surgical treatment of choice with scleral buckle alone having a 23-50% failure

rate and combined pars plana vitrectomy + scleral buckle around 43%. [8,10,12] The variable and higher than average failure rate is most likely the result of widespread vitreoretinal adhesions along with number and location of the retinal breaks.[10]

To increase the success of repair, meticulous release of any vitreoretinal adhesions is necessary to prevent postoperative contracture, retinal tearing, and recurrent detachment. Laser must be carefully applied posterior to the vitreous base which may be located more posteriorly than in the normal eye. Similarly, for scleral buckle to be effective, placement must be more posterior in order to relieve traction at the posteriorly-inserted vitreous base.

This case highlights the importance of lifelong and careful evaluation of the posterior segment in patients with a history of ROP. In addition, awareness of the sequalae of ROP and history of prematurity informs the need for further work up and surgical planning.

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