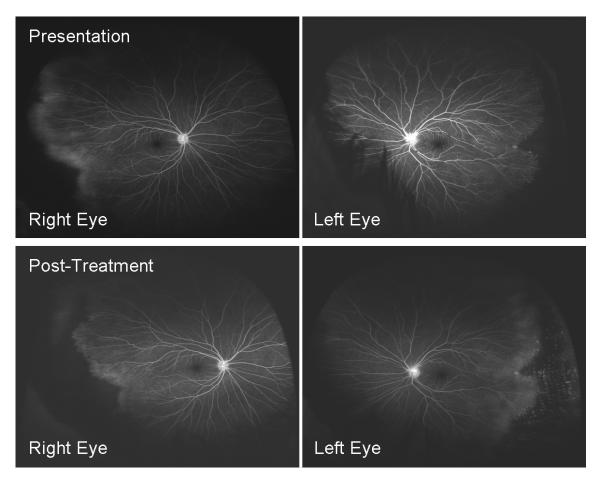


Clinical case blog

Title: Ultra-widefield Fluorescein Angiography of Familial Exudative Vitreoretinopathy

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A 44 year-old healthy female presented with 1 day of blurred vision in the left eye. Her visual acuity was 20/20 in the right eye and 20/50 in the left. On examination, the right eye appeared normal and there was a vitreous hemorrhage in the left. Fine vessels on the left optic disc were suspicious for neovascularization, but there were no other signs of retinopathy. She had no history of diabetes or other vasculopathies. Ultra-Widefield Fluorescein Angiography (UWFA) was obtained using an Optos (Dunfermline, Scotland, UK) scanning laser ophthalmoscope (Figure 1). Both eyes show an avascular zone of peripheral retina, peripheral arteriovenous anastamoses, and supernumerous vascular branching. Leakage peripherally and at the left optic disc is consistent with retinal neovascularization. No neovascularization is seen on the right. In each eye, there is mild late leakage at the border of the peripheral avascular zone but no evidence of vasculitis. The patient denies being born prematurely and is not aware of any family history of eye problems. A review of systems was unremarkable. Given the absence of prematurity or inflammatory conditions, the patient was diagnosed with Familial Exudative Vitreo-retinopathy (FEVR). This is typically an autosomal dominant disease with variable expressivity, and while the majority of affected patients retain good vision, complications include retinal neovascularization, vitreous hemorrhage, and retinal traction and detachment [1]. Most patients with FEVR are asymptomatic and are thus unaware of their disease until complications develop. In one study, while only 8% of newly-diagnosed patients reported a positive family history, 58% of asymptomatic relatives were subsequently found to have retinovascular abnormalities [2]. As UWFA be-

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comes increasing available, diseases affecting the peripheral retina may be more commonly diagnosed. In the present case, areas of retinal ischemia in the left eye were treated with laser photocoagulation (Figure 2), and the neovascularization subsequently regressed.

References

- 1. Ebert EM, Mukai S (1993) Familial exudative vitreoretinopathy. Int Ophthalmol Clin 33: 237-247.
- 2. Kashani AH, Learned D, Nudleman E (2014) High prevalence of peripheral retinal vascular anomalies in asymptomatic family members of patients with familial exudative vitreoretinopathy. Ophthalmology 12: 262-268.