



Asymmetric Optic Nerve Head Edema in a Patient with Neuro-Behçet Disease

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Background

Behçet's Disease is a systemic vascular disease that can affect multiple organ systems and classically presents as a triad of oral aphthous ulcers, genital ulcers, and ocular disease. Ocular manifestations include but are not limited to: non-granulomatous anterior uveitis with or without hypopyon, vitritis, retinitis, papillitis, and occlusive retinal vasculitis. Neuro-Behçet Disease (NBD), which occurs in approximately 10% of patients, is a well recognized cause of papilledema due to venous sinus thrombosis, and can also cause a variety of other ocular symptoms, including cranial nerve palsies and strokes. The literature contains many cases of patients with either uveitic disc edema or papilledema, but there are far fewer published reports of patients with both entities.

Case

A 29 year-old male with a past medical history of suspected Behçet's disease secondary to erythema nodosum, aphthous ulcers, knee arthritis, and HLA-B51 positivity presented to the eye clinic at the Syracuse VA Medical Center in December 2020 with decreased vision in both eyes. He was found to have asymmetric optic nerve head edema and vitritis.

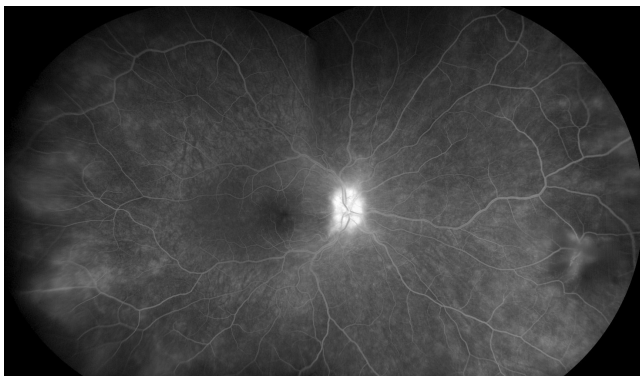
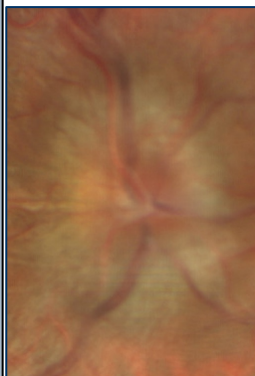
The patient was referred to the ED and found to have a venous sinus thrombosis on neuroimaging, for which he was admitted and started on high-dose IV solumedrol and anti-coagulation. He was later discharged on azathioprine 50 mg daily, prednisone 70 mg daily, celecoxib, meloxicam, pantoprazole, and enoxaparin and referred to Upstate University Center for Vision Care for further management.

Case Continued

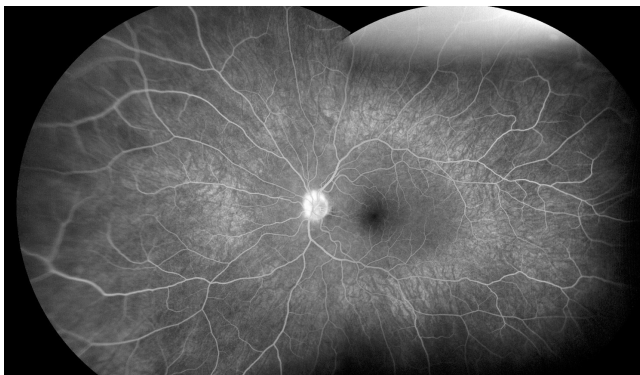
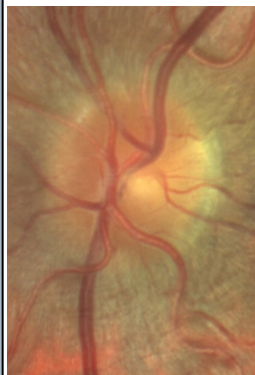
Follow-up examination showed no active inflammation in the anterior chamber or vitreous, but there was still asymmetric optic nerve head edema with associated asymmetry of optic nerve staining and leakage on fluorescein angiogram along with peripheral choroidal leakage and temporal venous leakage in the right eye. These findings were consistent with a unilateral papillitis and vasculitis along with papilledema secondary to the patient's venous sinus thrombosis. The patient was started on adalimumab and slowly tapered down on oral prednisone with continued improvement of visual acuity, vasculitis, and optic nerve edema on subsequent visits.

Ancillary Ophthalmic Testing

Right Eye



Left Eye



Discussion

- Behçet's Disease is a diagnosis based on clinical criteria
- 30% of patients with ocular involvement will have neuro involvement, but only 10% of patients with NBD will have ocular manifestations
- Asymmetric papilledema is uncommon and should prompt additional testing to more thoroughly evaluate the findings
- Fluorescein angiogram remains an invaluable tool in ophthalmology and should be performed in all patients with Behçet's Disease with ocular symptoms

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