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Case report

Neuroretinitis in a young woman

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A 33 year-old healthy woman presented with 4-days history of blurry vision in the right eye. She saw an optometrist yesterday who noticed a swollen optic nerve in the right eye and referred her to the emergency department. There she had a CT scan of the brain performed which was interpreted as normal. She was then seen by an ophthalmologist who diagnosed her with optic neuritis and referred her to our service.

Central vision was 20/40 in the right eye and 20/20 in the left eye. There was no relative afferent pupillary defect on careful testing (RAPD). Ophthalmoscopy revealed a hyperemic and swollen right optic nerve without peri-papillary hemorrhages (Fig. 1A). There was no vitritis. Formal visual fields demonstrated a central scotoma in the right eye and was normal in the left eye. Ocular coherence tomography of the macula demonstrated macular edema with the fluid tracking to the macula from the swollen optic nerve (Fig. 1B), explaining decreased central visual acuity and central scotoma on visual field testing.

The patient was not taking any medications and had no recent history of febrile illness. She mentioned that she recently gave away two feral cats who lived in her backyard for several months. Because of at least moderate optic nerve head swelling and the absence of RAPD, it was thought that the most likely site of pathology is intra-ocular portion of the optic nerve and peri-papillary retina, with sparing of optic nerve axons. Provisional diagnosis of neuro-retinitis was made. Bartonella Hensellae and Borrelia Burgdorferi antibody titers, syphilis serology, quantiferon gold, angiotensin-converting enzyme levels, and chest X-ray were ordered. Empiric treatment with 500 mg of oral ciprofloxacine for 2 weeks was prescribed.

One week later visual acuity worsened to 6/30 in RE. There was still no RAPD. Right optic nerve was still swollen but there was now a ring of hard exudates around the macula (Fig. 1C). Bartonella Hensella IgG antibody titers were elevated but the rest of the work-up was unrevealing. When repeated 2 weeks later, titers of IgG antibodies increased three-fold thus confirming acute infection. Treatment with 40 mg of oral prednisone commenced and continued for 3 weeks. Four weeks later, visual acuity improved to 6/9 and optic nerve swelling has almost completely resolved (Fig. 1D).

The diagnosis of neuroretinitis can be made in the presence of optic nerve head edema and the delayed (by 2–4 weeks) appearance of macular star that develops secondary to leakage from peripapillary capillaries causing the exudates to settle in the circular pattern in outer plexiform layer of the retina around macula. After the macular star develops, the diagnosis can be made easily easy thus obviating the need for neuro-imaging as the etiology of neuro-retinitis is not related to optic nerve inflammation beyond its intraocular portion. The diagnosis though can be difficult for a neurologist to make as evaluation of the macula with direct ophthalmoscope is not easy and initial presentation with a swollen optic nerve and variably decreased central visual acuity can resemble that of demyelinating optic neuritis. The important distinguishing features is that patients with neuroretinitis never experience pain on eye movements which is present in over 95% of patients with demyelinating optic neuritis [1,2]. Optic nerve head edema can be present in up to 1/3 of patients with optic neuritis, however, it is usually very mild and peripapillary hemorrhages are distinctly uncommon and seen in less than 5% of cases while they are almost universally present in patients with neuroretinitis [1–3]. As the diagnosis of neuro-retinitis can be confirmed only by the development of the macular scar, it is prudent for all patients presenting with optic nerve head swelling in whom the diagnosis of optic neuritis is entertained, be examined by an ophthalmologist in order to exclude alternate etiologies.

The etiology of neuro-retinitis is usually either idiopathic or infectious. Various infectious agents have been identified as culprits, most common ones being Bartonella Hensella, syphilis, and Lyme disease but other infections have also been described as causes of neuroretinitis [3,4].

Several conditions (malignant hypertension, asymmetric papilledema, non-arteritic anterior ischemic optic neuropathy (NAION), diabetic papillitis and atypical optic neuritis) can also masquerade as neuro-retinitis and present with unilateral or asymmetric optic nerve edema and macular star [3–5]. In malignant hypertension, the key findings on retinal examination are multiple hard exudates and the findings are almost always bilateral. In atypical optic neuritis again visual function is almost always severely impaired and RAPD should be always be present, even if it is bilateral. In NAION visual function is almost always abnormal and RAPD should be present in all cases. In diabetic papillitis visual function is preserved and it occurs in patients

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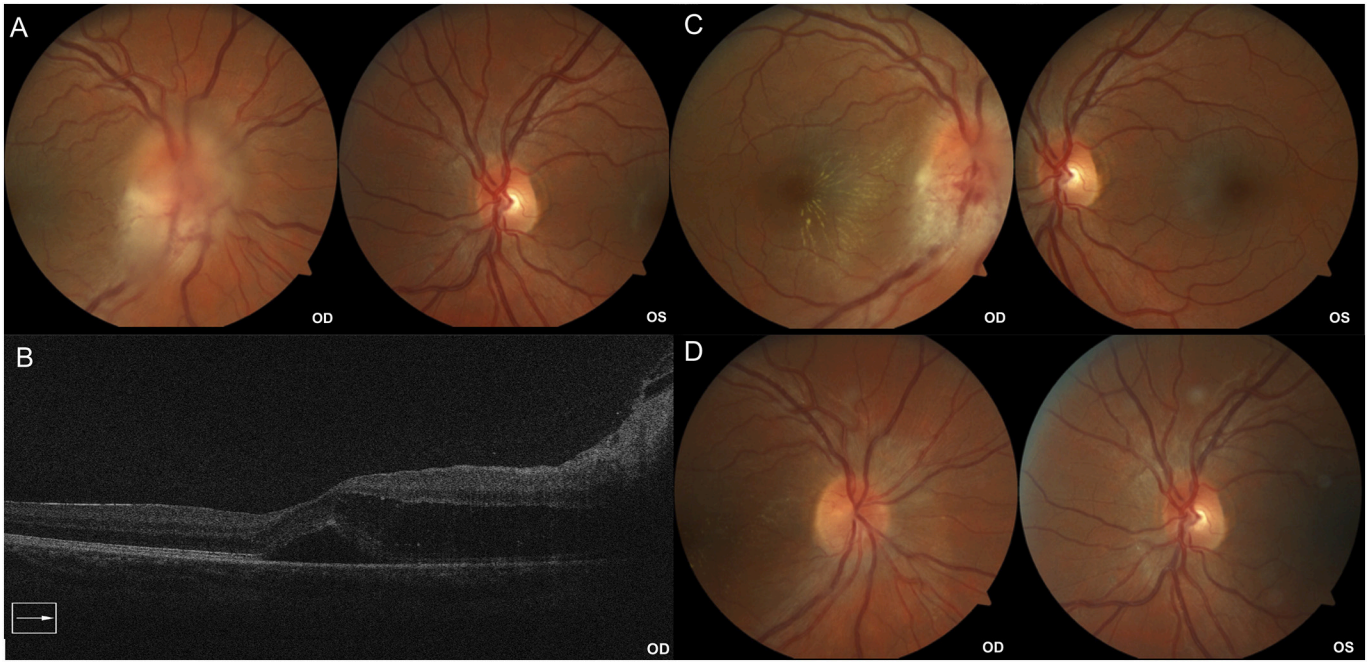

Fig. 1. (A) Fundus photographs of the optic nerves at presentation demonstrating swelling of the right optic nerve head. (B) Ocular Coherence Tomography of the macula demonstrating presence of submacular fluid tracking there from the swollen optic nerve. (C) Fundus photographs of the optic nerves 2 weeks after initial presentation demonstrating evolving right optic nerve head swelling and development of macular star around right macula. (D) Fundus photographs of the optic nerves four weeks after initial presentation demonstrating almost complete resolution of right optic nerve head edema and macular scar.

with known type 1 diabetes mellitus. Most masqueraders can be distinguished from neuroretinitis by the fact that retinal abnormalities are present at the onset and most have abnormal visual function at presentation.

It is important for a neurologist to identify the patient with neuroretinitis as they are not at risk of developing multiple sclerosis, do not require neuro-imaging and should instead be tested for a panel of specific infectious diseases [1,3–5].

Declaration of competing interest

The authors declare that they have no conflict of interest.

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