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Selected Topics: Neurological Emergencies

ULTRASOUND-ASSISTED DIAGNOSIS OF OPTIC NEURITIS IN THE EMERGENCY DEPARTMENT: A CASE REPORT

Daniel Smith, MD, Jason Arthur, MD, MPH, Zachary Lewis, MD, and Gregory Snead, MD

Department of Emergency Medicine, University of Arkansas for Medical Sciences, Little Rock, Arkansas
Reprint Address: Jason Arthur, MD, MPH, Department of Emergency Medicine, University of Arkansas for Medical Sciences, 4301 W. Markham Street, Slot 584, Little Rock, AR 72205.

□ **Abstract—Background: Optic neuritis is a common cause of subacute unilateral vision loss, occurring in 1–5 per 100,000 persons per year. It is more common in Caucasians, women, and those from countries with northern latitudes. Those aged 20–49 years are at greatest risk. The condition arises due to inflammation of the optic nerve. Inflammation may occur due to systemic inflammatory disorders, most commonly multiple sclerosis. Case Report: A 21-year-old African-American male presented to our emergency department with a complaint of painful unilateral vision loss. On examination he was found to have a relative afferent pupillary defect and red desaturation. A bedside ultrasound suggested pseudopapilledema suggestive of optic neuritis. He was admitted to Neurology for confirmation of and treatment for optic neuritis. Magnetic resonance imaging confirmed optic neuritis. The patient was treated with i.v. steroids and discharged after improvement in visual function. Why Should an Emergency Physician Be Aware of This?: Optic neuritis is a clinical diagnosis. The subtle historical components and examination findings make it a diagnostic challenge for the busy emergency physician. Early diagnosis may improve visual outcomes. Discovery of pseudopapilledema on bedside ultrasound may be seen in optic neuritis, and is another finding that emergency physicians may assess for in patient presenting with unilateral vision loss.**

□ **Keywords—pseudopapilledema; optic disc elevation; optic neuritis; multiple sclerosis; relative afferent pupillary defect; RAPD; unilateral vision loss; unilateral eye pain; ultrasound; point-of-care ultrasound; bedside ultrasound**

INTRODUCTION

Optic neuritis is a common cause of subacute unilateral vision loss, occurring in 1–5 per 100,000 persons per year. It is more common in Caucasians, women, and those from countries with northern latitudes. Those aged 20–49 years are at greatest risk (1). The condition arises due to inflammation of the optic nerve. Inflammation may occur due to systemic inflammatory disorders, most commonly multiple sclerosis (MS) (2).

CASE REPORT

A 21-year-old African-American male with a medical history of migraines presented to the emergency department (ED) with painful unilateral vision loss. He reported 1 month of gradually worsening vision in his left eye, most notable when filling out paperwork associated with his job. During this time he also experienced a decrease in vibrancy of color in the left eye. In the 3 days preceding his ED presentation, he developed pain and photophobia in the left eye. The pain worsened with movement of the eye. Additionally, he reported mild numbness in the periocular region of the left face. He denied any trauma, chemical exposure, foreign body, or work around machinery. He had no history of eye problems and no family history of ocular or neurologic disease.

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On examination, he had 5/5 strength in his bilateral upper and lower extremities, finger-nose-finger was normal, he ambulated with a steady unassisted gait, and sensation was intact to light touch in all extremities. Cranial nerves II–XII were intact, with the exception of a left-sided relative afferent pupillary defect (RAPD). There was no tenderness on percussion over the temporal arteries. An eye examination was performed and was remarkable for the aforementioned RAPD OS, as well as left-sided red desaturation. Intraocular pressure was normal, fluorescein uptake was absent, and a slit-lamp examination was normal. A point-of-care ultrasound of the eyes, performed in lieu of fundoscopy, demonstrated left-sided optic disc elevation with a normal optic nerve sheath diameter (ONSD) of 3.1 mm, known as pseudopapilledema (Figures 1 and 2). Ultrasound of the right orbit showed neither optic disc elevation nor ONSD enlargement (Figure 3).

A diagnosis of optic neuritis was made, Ophthalmology and Neurology were consulted, and the patient was admitted to the Neurology service for high-dose steroids. A dilated eye examination by Ophthalmology showed left-sided grade 3 disc edema, RAPD, pain with extraocular movement, and dyschromatopsia consistent with optic neuritis. Magnetic resonance imaging (MRI) of the brain and orbits without contrast demonstrated T2 hyperintensity and mild enhancement of the left optic nerve consistent with optic neuritis. The remainder of the MRI was normal. A lumbar puncture was performed to investigate potential causes of the patient's symptoms, however, opening pressures were not obtained. Cerebrospinal fluid (CSF) cell count suggested a traumatic tap at 49 red blood cells and 1 white blood cell; however,

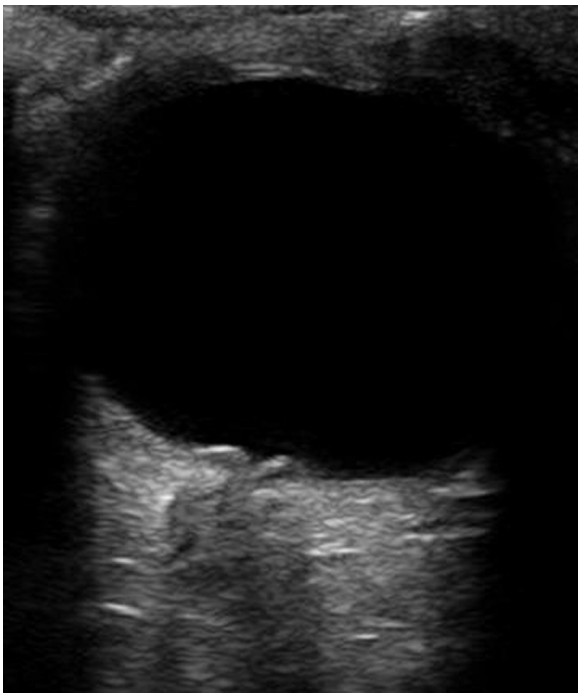

Figure 2. Ultrasound of the left orbit in the long axis demonstrating optic disc elevation with a normal optic nerve sheath diameter.

Figure 1. Ultrasound of the left orbit in transverse axis demonstrating optic disc elevation with a normal optic nerve sheath diameter, known as pseudopapilledema.

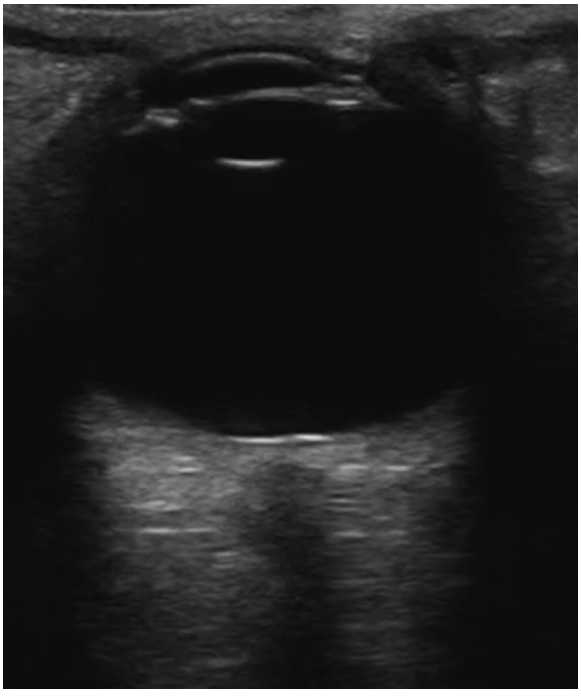

Figure 3. Ultrasound of the right orbit demonstrating normal anatomic features.

protein and glucose were normal. Gram stain and cultures were negative. Aquaporin-4 receptor antibody, neuromyelitis optica antibody, CSF oligoclonal bands, CSF IgG, CSF VDRL, an antinuclear antibody screen, serum human immunodeficiency virus, and a serum hepatitis panel were negative. He was admitted and treated with 1 g i.v. methylprednisolone for 5 consecutive days. During his hospitalization, his vision improved subjectively and visual acuity at the time of discharge was finger counting in the left eye. He was discharged with a plan for Neurology and Neuro-opthalmology follow-up as an outpatient.

DISCUSSION

Optic neuritis is a clinical diagnosis. Visual loss is typically unilateral and progresses over hours to days (2). Symptoms of optic neuritis include orbital pain, pain with eye movement, and progressive visual loss. Flashes of light with eye movement, known as photopsias or phosphenes, may also be present (1). Patients may report difficulty distinguishing colors, with red in particular being reported frequently. Patients may also note worsening symptoms with exercise, exposure to warm temperatures or to warm water, known as Uhthoff's phenomenon (2).

Examination typically shows decreased visual acuity, decreased color vision and contrast sensitivity, visual field defect, and RAPD. Absence of RAPD can occur if the eye is unhealthy secondary to either retinal disease or disease of the optic nerve in the unaffected eye (2). There are several methods for identifying red desaturation. Many Snellen eye charts include red and green lines and therefore allow for gross comparison between the asymptomatic and symptomatic eyes. Pseudoisochromatic color plates may also be used for gross evaluation. More objective testing can be achieved with the use of Farnsworth Panel D-15 or Lanthony desaturated 15-hue tests, among others (3). In our case, red desaturation was detected by having the patient view the treating physician's hospital identification badge, which has a red border, with the asymptomatic eye followed by the symptomatic eye. Optic disc swelling is seen in roughly one-third of cases (1,2). Retinal examination is typically normal (1). Although optic neuritis is a clinical diagnosis, MRI of the brain and orbits provides diagnostic confirmation and assessment for the presence or risk of MS (4).

Pseudopapilledema is optic nerve head swelling that is not associated with increased intracranial pressure. Pseudopapilledema is also known as papillitis or optic disc or optic nerve edema. ONSD is commonly measured 3 mm posterior to the retina. A cutoff of 5.7 mm as the upper limit of normal results in 100% sensitivity for pseudopapilledema (5). Our institution uses 5 mm as the upper limit of normal of ONSD averaged between both eyes.

Treatment of optic neuritis centers on reduction of the risk of developing MS and restoring vision loss. The mainstay of treatment is high-dose i.v. corticosteroids. There is some evidence that this reduces the risk of MS development and reduces time to vision recovery, although it has not been shown to improve long-term vision outcomes compared to oral steroids or placebo (6–8). Additionally, evidence suggests that treatment with oral prednisone increases the risk of developing recurrent symptoms (7,8).

Prognosis of the condition is variable. Left untreated, vision improvement is seen after a few weeks and may continue, and 90% of affected patients improve to 20/40 vision or better at 1 year (9). Following resolution of the acute symptoms, chronic signs of optic neuritis may persist. These can include persistent vision loss, RAPD, temporary exacerbations by Uhthoff's phenomenon, continued color desaturation, and optic atrophy. Many of these symptoms may persist even after recovery of functional vision (10). Patients with optic neuritis are at risk for developing MS. The risk of developing clinically definite MS at 5 years was shown to be 30% in the Optic Neuritis Treatment Trial following the first episode of demyelinating optic neuritis (11).

WHY SHOULD AN EMERGENCY PHYSICIAN BE AWARE OF THIS?

Optic neuritis is a rare but important diagnosis to make in the ED. The subtle historical components and examination findings make it a diagnostic challenge for the busy emergency physician. Failure to recognize these subtleties may result in loss of vision. Bedside ultrasound of the orbit to assess for optic disc elevation may be a helpful aid in the diagnosis of optic neuritis. Findings suggestive of pseudopapilledema should cue the emergency physician to re-evaluate the history and examination, spur emergent Ophthalmology consultation, and increase diagnostic accuracy.

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