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Selected Topics: Neurological Emergencies

ACUTE OPTIC NEURITIS DIAGNOSED BY BEDSIDE ULTRASOUND IN AN EMERGENCY DEPARTMENT

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☐ **Abstract—Background: Transorbital ultrasound was used to diagnose acute optic neuritis (AON) at bedside in an emergency department (ED). Case Report: A 59-year-old female patient presented to an ED after 7 days of progressive unilateral visual loss while she was receiving outpatient treatment for relapsing-remitting multiple sclerosis. Transorbital ultrasound revealed a disparity between the optic nerve sheath diameters of the affected and nonaffected eyes and striking optic nerve edema in the affected eye. These findings led to a diagnosis of AON and early definitive treatment. Why Should an Emergency Physician Be Aware of This?: Given an absence of reliable diagnostic criteria for AON, comorbidity with multiple sclerosis, and limitations inherent to magnetic resonance imaging, transorbital sonography may facilitate diagnosis of this condition in emergent presentations.**

☐ **Keywords—transorbital ultrasound; acute optic neuritis; multiple sclerosis; optic nerve sheath diameter; optic nerve diameter; edema**

INTRODUCTION

Optic neuritis (ON) involves an inflammatory demyelination of the optic nerve that is associated with multiple sclerosis (MS). ON is the primary presenting feature in approximately 15% to 20% of MS cases, and 38% to 50% of MS patients go on to develop this condition (1). ON mostly affects female patients aged 18 to 45 years (2). A classic presentation of ON involves acute and painful monocular visual loss with resolution over several weeks to months (2,3). ON is diagnosed by clinical assessment from history and examination findings. Patients often report monocular pain over several days, after which they develop ipsilateral visual loss (1). During examination, patients consistently display decreased visual acuity in the affected eye with a relative afferent pupillary defect (4,5).

Although clinical findings may be useful in an assessment for ON, there are no uniform diagnostic criteria (6). As such, the level of clinical experience may influence a potential diagnosis, and suspected ON cases may be confused with similar or mimicking conditions (7). In emergent presentations, a diagnosis of acute optic neuritis (AON) may be very challenging. Dilatated fundoscopic examination is not always feasible in the emergency department (ED), and is only 35% sensitive when looking for the presence of optic disc edema (2). Although gadolinium-based magnetic resonance imaging (MRI) of the brain is highly sensitive, it is costly, time consuming, and not universally available (8). As such, MRI has been deemed "usually unnecessary" for diagnosis, "of unclear value" with respect to overall visual

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prognosis, and perhaps more suitable for "documentation of optic nerve inflammation" (1).

The treatment of ON has been investigated extensively. The Optic Neuritis Treatment Trial and subsequent randomized clinical trials have shown that pulsed high-dose corticosteroid therapy accelerates visual recovery in the first few weeks from symptom onset (1,9–11). Despite this, the clinical trials failed to demonstrate a long-term benefit of corticosteroid therapy due to the high rate of spontaneous resolution over time (1). It also has been suggested that aggressive corticosteroid therapy may decrease long-term complications in comorbid relapsing-remitting multiple sclerosis (RRMS) by impeding brain atrophy (1).

Timely diagnosis of AON followed by the administration of high-dose corticosteroids is, therefore, of prime importance. To date, a limited number of ED case reports and a newly published ED case series have described ultrasound-confirmed diagnoses of AON at bedside (12–15). In these presentations, emergency physicians identified increased optic nerve sheath diameter (ONSD) in the affected eyes of patients experiencing painful monocular visual loss. In our case report, we describe another clinical example wherein emergency physicians used transorbital ultrasound to diagnose and initiate treatment for a patient with AON. We also review data regarding transorbital sonography (TOS) in this setting.

CASE REPORT

A 59-year-old African American woman with a history of RRMS and hypertension presented to the ED of a university teaching hospital with progressive right-sided visual loss over a period of 7 days. The patient did not report unilateral ocular pain, but complained of a migratory headache. She denied eye redness, focal weakness, paresthesias, and slurred speech.

The patient had been diagnosed with RRMS 1 year prior to presentation after MRI confirmation of multiple spinal cord lesions. Despite compliance with multiple immunomodulatory regimens, she reported several recurrences. During these episodes, she reportedly experienced numbness, tingling, saddle anesthesia, bladder fullness, and episodes of visual loss. The patient had not been diagnosed previously with AON or with neuromyelitis optica. During an outpatient neurology workup, she received an aquaporin-4-autoantibody assay that returned within normal limits.

On arrival, the patient was alert and oriented with mildly elevated blood pressure (166/77 mm Hg). On physical examination, both pupils were deemed reactive, although a possible right-sided afferent pupillary defect was noted later. Visual acuity was documented as 20/70 in the right eye and 20/30 in the left eye. Color vision was intact. Intraocular pressures were measured at 15 mm Hg in the right eye and 13 mm Hg in the left eye. The patient did not receive a formal fundoscopic examination. Neurological assessment revealed normal cranial nerves III–XII, including intact and symmetrical extraocular muscle movements, 5/5 strength in all extremities, intact sensation to light touch in all extremities, and normal "heel-to-shin" and "finger-pointing" tests.

Transorbital ultrasound was performed at bedside during the initial assessment. A linear array 13-6 MHz SonoSite HFL38xp probe (FUJIFILM SonoSite, Inc., Bothell, WA) was applied directly with minimal pressure over the eyes, which were scanned via sterile ultrasound gel medium in the longitudinal and short-axis orientations to visualize the orbits in full detail. The patient was asked to move her eyes to the left and right until proper orientation could be obtained for visualization of the optic nerves in cross section. Several still images and brief videos were recorded. The sharpest images were chosen for analysis and ONSD measurements were performed directly on these images via the built-in SonoSite X-Porte software (FUJIFILM SonoSite, Inc.).

The measurements yielded an ONSD of 6.5 mm in the affected right eye and an ONSD of 3.6 mm in the nonaffected left eye. Both measurements were taken at a distance of 3.0 mm posterior to the globe. In more descriptive terms, the images demonstrated striking optic nerve edema in the affected right eye that was not apparent in the nonaffected left eye. The images did not reveal evidence of vitreous hemorrhage, retinal detachment, or papilledema. Figures 1 and 2 present the ultrasound images and associated data.

The patient was admitted and received a dose of i.v. methylprednisolone 1000 mg. On the following day, the patient received a second dose of i.v. methylprednisolone 1000 mg and underwent brain MRI demonstrating "asymmetric signal … within the right greater than left optic nerve on T2-weighted sequences" with "minimal posterior enhancement." On the third hospital day, the patient received another dose of i.v. methylprednisolone 1000 mg and was discharged to home in stable condition. The patient was not placed on a course of oral corticosteroids. On outpatient brain MRI repeated 1 month later, the T2 enhancement previously identified in the right optic nerve was not evident. Six months later, the patient experienced a recurrence in ocular symptoms.

DISCUSSION

TOS is recognized as a core clinical competency of emergency physicians by the American College of Emergency Physicians (16). The effectiveness of this modality in ED settings has been documented with respect to the

evaluation of acute ocular conditions including elevated intracranial pressure; retinal, choroidal, and vitreous detachments; lens dislocations; globe ruptures; retrobulbar hemorrhages; and identification of retained foreign bodies (16).

Despite limited information regarding TOS in the diagnosis of AON, it has been shown that sonographic measurement of ONSD is feasible at bedside. Hassen et al. demonstrated that appropriately trained postgraduate and attending physicians could obtain sonographic ONSD measurements in close approximation to retrospective ONSD measurements obtained by board-certified radiologists on computed tomography scans (correlation coefficient 0.9; 95% confidence interval 0.8846–0.9303) (17). On the other hand, limitations to bedside ONSD measurement included operator dependence, lack of interpersonal reliability, variable patient compliance, misidentification of ocular landmarks, and challenges in identifying the edges of the optic nerve sheath (17).

Over the last few years, a growing body of useful data has emerged from outpatient neurology settings with respect to TOS and ON. A 2014 case-control study by Lochner et al. compared the ONSD in 21 patients previously diagnosed with unilateral optic neuritis to the ONSD of 21 matched controls (18). This study detected a statistically significant disparity between the ONSD for affected eyes of ON patients and the ONSD for nonaffected eyes of patients and healthy controls (18). The median ONSD was recorded as 6.3 mm for affected eyes (interquartile range [IQR] 5.9–7.2 mm) compared with 5.5 mm for nonaffected eyes (IQR 5.1–6.2 mm), and similarly, 5.2 mm for controls (IQR 4.8–5.5 mm) (18).

The median optic nerve diameter (OND), in distinction from ONSD, did not seem to vary significantly between patients and controls in Lochner et al.'s 2014 study (18). However, in a more expansive 2017 longitudinal study, Lochner et al. demonstrated that median OND was, in fact, significantly increased among ON patients (3.4 mm; 2.9–3.8 mm) relative to healthy controls (2.7 mm; 2.5–2.9 mm) (19). Lochner et al. further correlated this finding with information regarding visual acuity and biomarkers (19).

In this case report, we demonstrated an ultrasound-guided diagnosis of AON. As seen in Figures 1 and 2, the ONSD measurement was pronounced in the affected eye (6.5 mm). This observation is in keeping with the reported median ONSD measurement (6.3 mm) for affected eyes of ON patients included in Lochner et al.'s 2014 case control study and 2017 longitudinal study (6.3 mm; 6.0–6.5 mm) (15). In contrast, the ONSD measurement was small in the nonaffected eye (3.6 mm) and below the median and interquartile range for the healthy controls included in Lochner et al. 2014 and 2017 (5.2 mm; 4.8–5.5 mm) (18,19). This measurement also was below the lower end of the

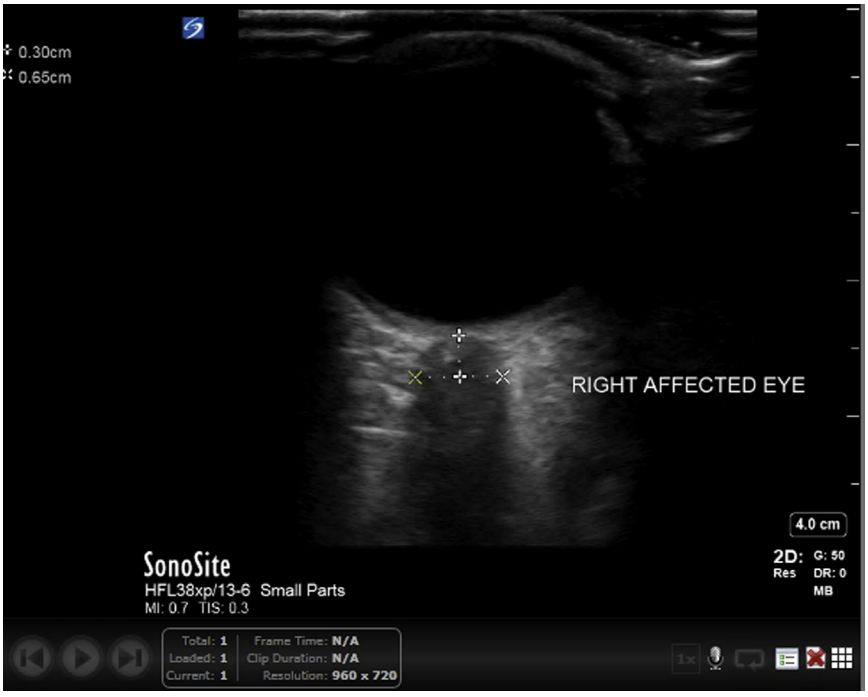

Figure 1. Transorbital ultrasound of the affected right eye. Optic nerve sheath diameter was 6.5 mm at a distance of 3.0 mm posterior to the globe. Optic nerve edema is demonstrated.

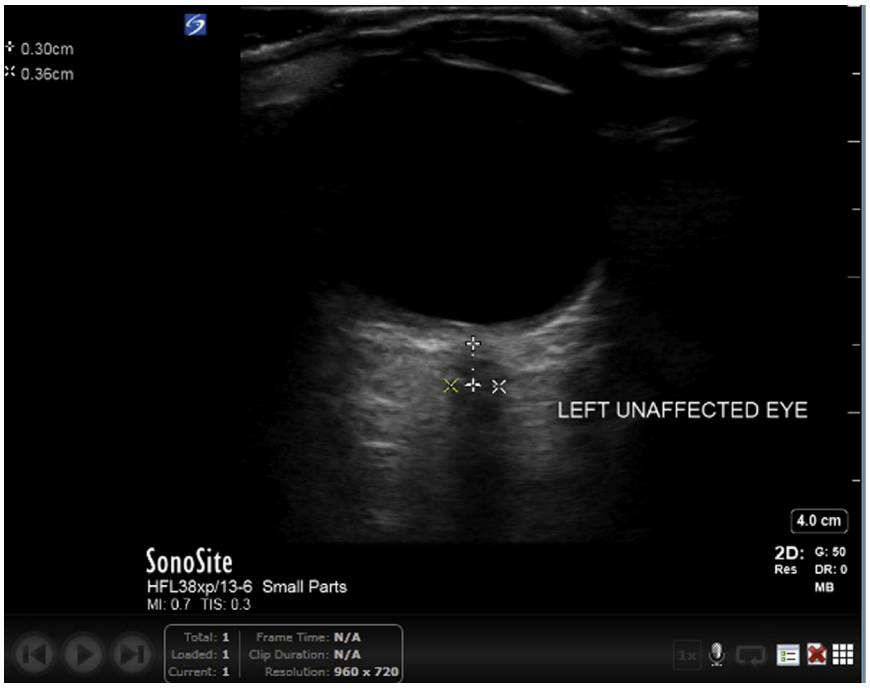

Figure 2. Transorbital ultrasound of the nonaffected left eye. Optic nerve sheath diameter was 3.6 mm at a distance of 3.0 mm posterior to the globe. Optic nerve edema is not present.

range of ONSD measurements (4.3 mm) observed in 40 healthy subjects in Bäuerle et al. (20).

This discrepancy suggests a degree of underestimation of the ONSD for the study patient's nonaffected left eye. However, this did not affect our diagnosis of AON. In this case, a stark incongruity between the affected and nonaffected optic nerves was more valuable in narrowing the diagnosis. From the ED perspective, it is worthy to consider if the overall morphology of the optic nerve and the OND measurement may be more useful than the ONSD measurement. In a 2015 systematic review, Lochner et al. observed that increased OND is a highly reliable feature of AON, is consistent with optic nerve edema and the presence of an early optic nerve lesion, is equivalent to Gd-DTPA leakage identifiable on MRI, and is detectable via TOS in approximately 80% of patients with AON (21). Lochner et al. further noted that the majority of the studies included in their analysis did not distinguish clearly between OND and ONSD, and that "when the sheath was included, most of the increase was attributable to this and not to the optic nerve itself" (21).

Based on the available data, Lochner et al. could not propose "a clear cut-off value" for OND between affected eyes to either contralateral eyes or to controls (21). The authors nonetheless suggested a relationship between the "thickness of the nerve and the clinical features of AON" such that a higher OND may be associated with more severe visual loss and lower likelihood of full recovery (21). This is not a new suggestion. Even in 1995, Dees et al. observed, "Echographically normal optic nerves were associated with limited visual loss (better than 6/60) and recovery to 6/9 or better in all cases, whereas echographically swollen optic nerves were associated with visual loss to less than 6/60 in 55% of cases" (5). All of this suggests that an expanded ED case series with a larger sample of patients, multiple sonographers, and clear parameters may offer more information regarding ONSD, OND, or assessment for optic nerve congruity as descriptive statistics in ED presentations of AON.

Alternative diagnoses including elevated intracranial pressure, acute angle closure glaucoma, migraine, stroke, and brain mass were considered during the evaluation of the study patient. These diagnoses were viewed as less likely than AON given the patient's known history of RRMS, the physical examination findings, and the transorbital ultrasound images obtained at bedside. Giant cell arteritis was considered and deemed unlikely in light of normal erythrocyte sedimentation rate and C-reactive protein levels during recent RRMS exacerbations in the study patient. As such, inflammatory markers and temporal artery ultrasound were not ordered.

WHY SHOULD AN EMERGENCY PHYSICIAN BE AWARE OF THIS?

Overall, this case report has clinical implications with respect to the diagnosis of AON in ED settings. TOS

may serve as an economical, timely, frequently available, noninvasive, and low-risk diagnostic modality for suspected AON. This may, in turn, lead to provision of the treatment of choice, improved patient outcomes, and potentially expedited workup for MS in previously undiagnosed cases. Given an absence of reliable diagnostic clinical criteria for AON, the challenges associated with traditional clinical assessment, and certain limitations inherent to brain MRI, bedside ultrasound may be worthy of further investigation in urgent or emergent presentations of this condition.

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