Case Report

More than meets the eye: Point-of-care ultrasound diagnosis of acute optic neuritis in the emergency department

Noa P. Yee *,1, Saman Kashani, MD MSc 2, Thomas Mailhot, MD 2, Talib Omer, MD 3,4

Department of Emergency Medicine, Los Angeles County + University of Southern California Medical Center, Los Angeles, CA, United States of America

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A B S T R A C T

Optic neuritis (ON) is an inflammatory condition that causes demyelination and thickening of the optic nerve leading to acute/subacute vision loss. It is frequently associated with other conditions like multiple sclerosis, but is often misdiagnosed, which can lead to a suboptimal prognosis. Ultrasound is rarely utilized to help make this diagnosis, even though it can easily detect a thickened retrobulbar optic nerve sheath diameter. We describe four cases in which ultrasonographic measurement of the optic nerve sheath diameter aided in the diagnosis of ON.

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1. Introduction

It is estimated that 1–6% of patient visits to the emergency department (ED) cite ocular disturbances as the chief complaint [1]. Point-of-care ultrasonography (POCUS) has been used to detect ocular pathologies such as retinal detachment, increased intracranial pressure, and trauma to the eye [2-44,9]. However, another ocular pathology, optic neuritis (ON), is frequently overlooked. ON is an acute inflammatory demyelinating disorder of the optic nerve that, if left unidentified and untreated can lead to neuronal degeneration or axonal damage [10]. Typical symptoms are unilateral, subacute, and painful vision loss [11]. Pain is often worsened with extra-ocular movement and the examiner may detect an afferent pupillary defect [12]. Etiology of ON can be from inflammation, toxins, infections, or idiopathic [11].

ON is more commonly seen in women and in patients between the age of 20–40 years old [13,14]. In the United States, the incidence is reported to be approximately 6.4 per 100,000 [15]. Moreover, it has been shown to be associated with demyelinating disorders such as neuromyelitis optica and multiple sclerosis (MS) [16]. Approximately 50% of patients with MS develop ON at some point during their disease course and a small study found ultrasound measurement of optic nerve sheath diameter (ONSD) to be promising in the diagnosis of optic neuritis [17].

It has been shown that ON is linked to increased ONSD [18-24]. POCUS use by ED physicians for the evaluation of ONSD was shown to be efficacious and accurate when compared to CT and MRI, with a sensitivity as high as 100% and a specificity of 95% [19,25,26].

We present a case series of four patients who present to the ED with visual complaints. This case series is from a large, urban, high-volume ED in Los Angeles between December 2015 and May 2017. Initial patient exam and ultrasound was performed by ED residents and reviewed by dedicated US fellowship-trained ED faculty. POCUS ultrasound examinations were performed using a high frequency (13–6 MHz) linear array transducer on an M-Turbo ultrasound machine (SonoSite™, Bothell, WA). A transparent dressing (Tegaderm™) was used to avoid gel contact with the patient’s eyes. A generous amount of sterile gel was applied to minimize pressure of the probe on the globe. ONSD was measured 3 mm behind the posterior scleral surface, perpendicular to the optic nerve sheath. In order to measure the optic nerve sheath on-axis, the patients were instructed to look straight ahead and the ultrasound image view was verified to include the lens or iris; this was performed to prevent the possibility of measuring the optic nerve at an angle off midline, which may underestimate the ONSD – also known as the cylinder tangent effect [27,28]. ONSD measurements >5 mm or in patients with unilateral symptoms, a difference of at least 0.3 mm were considered abnormal [7]. Of note, no cases of ON were omitted that had a ONSD of <5 mm.

1.1. Case #1

A 35-year-old man with no past medical history, presented to the ED complaining of atraumatic right-sided vision loss and 4/10 right-eye pain with eye movement for 12 days. He described his vision loss as beginning with right-eye blurry vision which gradually worsened over the
next week until his vision went “completely black.” The patient denied symptoms in his left eye. He denied any concerning social or family history and was not taking any medications.

On arrival to the ED, the patient was alert and oriented and in no acute distress. His vital signs were within normal limits with the exception of slight tachycardia. Physical examination was unremarkable with the exception of the ophthalmologic exam.

The ocular examination revealed pupils that were 7 mm, the right eye was sluggishly reactive to light with a positive afferent pupillary defect, and the left eye was briskly reactive to light. Extraocular movements were intact bilaterally. Visual acuity showed no light perception in the right eye and 20/20 in the left. POCUS revealed an enlarged ONSD of 6.7 mm with no evidence of vitreous hemorrhage or retinal detachment. The POCUS findings in conjunction with the physical exam were concerning for acute ON.

Ophthalmology and neurology were both consulted from the ED and it was agreed that the most likely diagnosis was ON. An MRI of the brain showed an asymmetric, diffuse enhancement of the post-chiasmatic right optic nerve (cisternal and intracanalicular segments) with associated T2 FLAIR hyperintensity, without mass-like expansion. Fluid distention of the right optic nerve sheath was also identified as well as mild flattening of the posterior aspect of the right globe with suggestion of papilledema. A lumbar puncture and basic laboratory tests were unremarkable. The patient was hospitalized and started on a course of intravenous (IV) methylprednisolone with a good response. The patient’s vision in his right eye improved to detecting hand motion during his hospital stay. The patient’s final diagnosis was acute idiopathic right-sided ON, and he was discharged with MS clinic follow up.

1.2. Case #2

A 48-year-old man with no past medical history, presented to the ED complaining of acute onset bilateral vision loss for approximately 19 h. He described his vision as “black or looking through smoke,” associated with a mild frontal headache and worsened with eye movement. He had no prior history of visual complaints, and the patient denied any concerning social or family history and was not taking any medications.

On arrival to the ED, the patient was alert and oriented and his vital signs were within normal limits. The oculocer examination revealed atraumatic pupils that were equal, round and reactive with normal extraocular movements bilaterally. Visual acuity was limited to hand motion at 1 ft bilaterally. In addition, a left-sided relative afferent pupillary defect was noted. Intraocular pressures were within normal limits bilaterally. POCUS revealed a dilated ONSD of 7.8 mm on the right and a dilated ONSD of 7.5 mm on the left. No evidence of retinal detachment was detected bilaterally. Mild vitreous opacities were detected in the right eye, but not in the left eye.

Ophthalmology and neurology were both consulted from the ED and the diagnosis of ON was again suspected. Ophthalmology performed a dilated eye exam which supported the diagnosis of ON. The patient was admitted to the neurology service for additional workup of possible MS and for treatment of ON. An MRI of the brain showed mild increased T2 signal within the bilateral intraorbital optic nerves with associated marked enhancement and mild enlargement. Mild stranding and enhancement were seen within the bilateral intracanal fat adjacent to the optic nerves. These findings were most compatible with ON. A lumbar puncture and basic laboratory tests were unremarkable. IV methylprednisolone was started with minimal improvement in symptoms. Due to the refractory nature of his ON, the patient was then started on plasma exchange. He experienced improvement in his right eye vision, but had worsening and persistent visual loss in his left eye. No other focal neurologic deficits were noted. His final diagnosis was acute idiopathic bilateral ON. The patient was discharged in stable condition and referred for follow up at a neurology clinic.

1.3. Case #3

A 34-year-old female with no past medical history, presented to the ED complaining of acute left-sided blurry vision and mild associated photophobia for three days. The patient stated that her symptoms began three days prior with an increasing headache and bilateral eye pain associated with increased blurry vision in both eyes. The blurry vision was worse in her left eye than her right eye. She described her eye pain as bilateral retro-orbital pain that was sharp in nature, 7/10 in severity and radiated to the occiput. She denied any history of trauma, double vision, fever, or viral infections. The patient denied any concerning social or family history and was not taking any medications.

On arrival to the ED, the patient was alert and oriented and vital signs were within normal limits. ED physical exam revealed a headache exacerbated by ocular movement, and the rest of the physical exam was unremarkable. POCUS was remarkable for increased optic nerve diameter bilaterally at 5.5 mm and 5.6 mm without evidence of retinal detachment or vitreous hemorrhage. Neurology and ophthalmology consults were obtained from the ED.

On detailed ophthalmic exam, visual acuity on the right was 20/80, and with pinhole 20/40. Visual acuity on the left side was 20/150, and with pinhole 20/80. Visual fields were intact, but blurry bilaterally. Fundoscopic exam revealed mild bilateral papilledema, bilateral swollen peripapillary nerve fibers and blurred disc margins. The remainder of the neurologic exam was unremarkable. An MRI brain was completed in the ED and was concerning for enhancement of bilateral optic nerves as well as T2/FLAIR hyperintensities throughout the supratentorial white matter. In conjunction with the physical exam, these findings were concerning for MS and the patient was admitted to the hospital. During her admission, diagnostic tests including lumbar puncture, CSF studies, and MRI of the cervical spine were all unremarkable. The patient was started on IV methylprednisolone with good response. The patient’s final diagnosis was acute bilateral ON secondary to MS, and she was discharged with outpatient neurology and ophthalmology follow-up.

1.4. Case #4

A 28-year-old female with a past medical history of MS and previously-resolved ON, presented to the ED complaining of left eye pain and blurry vision for 4 days. She reported intermittent left-sided vision loss in her peripheral visual field during the past 4 days without double vision, numbness, tingling, or weakness. She also reported that her visual fields were getting smaller, particularly her left temporal field. She also endorsed blurriness in her central vision, associated retrobulbar pain and pain with extraocular movements. No other concerning findings were noted.

On physical examination, the patient was alert and oriented. Vital signs were within normal limits. Visual acuity on the right side was 20/40, and with pinhole 20/25. Visual acuity on the left was 20/150, with pinhole 20/30. The neurologic examination was otherwise unremarkable.

POCUS was remarkable for an increased ONSD on the left at 5.5 mm and on the right at 5.6 mm, with no evidence of retinal detachment or vitreous hemorrhage. Subsequent imaging studies of the brain were consistent with these findings. Her MRI brain detected multiple (>30) periventricular and subcortical areas of T2 prolongation compatible with multiple sclerosis. Orbital MRI detected a subtle hyperintensity on coronal STIR images within the posterior intraorbital portion of the left optic nerve and optic chiasm volume loss of the right optic nerve, both consistent with ON.

Ophthalmology and neurology were consulted from the ED, and a detailed fundoscopic exam was performed, which was remarkable for left eye disc pallor and blurred disc margins. The remainder of the neurologic exam did not demonstrate any further focal lesions. The patient’s final diagnosis was acute ON related to MS, and treatment for a recurrent
MS flare was initiated while in the ED. The patient was directly transferred to an acute rehabilitation facility for in-patient IV methylprednisolone and given a referral an ophthalmology clinic. See Table 1 below for a brief summary of the cases covered with respective POCUS images and measurements obtained.

2. Discussion

Point of care ultrasound is an emerging diagnostic modality in the ED for the evaluation of ocular pathology. The eye, like other fluid-filled anatomy, is well-suited for US visualization. Furthermore, the versatility, accessibility, and absence of side effects make POCUS an attractive tool for emergency physicians. Fundoscopic examination is useful to diagnose various conditions resulting in vision loss, however this exam may be difficult to perform due to poor patient compliance or non-dilated pupils. Moreover, specialized equipment that may improve the ability to perform fundoscopy (e.g., Panoptic ophthalmoscopes) is not always available in the ED, and ophthalmologic consultation is similarly limited. In this case series, POCUS showed value in identifying an inflammatory pathology thus initiating the management and specialty care of a time-sensitive diagnosis.

ON is usually diagnosed based on the patient’s clinical profile and exam with subsequent confirmation via MRI. Accurate diagnosis is critical in starting appropriate and prompt ophthalmologic and neurologic evaluation and management. Often, the treatment includes intravenous steroids with the goal of preserving long term visual function. While ultrasound can adequately assess for papilledema, the meta-analysis of Lochner et al. showed that sonographic papilledema in ON was only seen in 6–43% of patients with ON, so it is not very sensitive, as confirmed by our case series [18]. This may be due to the fact that in the acute setting papilledema may take some time to develop clinically. Optic disc elevation measurements are not part of our institutions sonographic optic nerve assessment protocol. If it is measured, a measurement of the disc height (from the dome of the papilla to the fundus) of >0.6 mm is considered positive [29].

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Disc swelling was also found to be related with the thickness of the optic nerve, since ONSD was increased in all patients with papillitis, compared to only 15/16 of those without in one study and ONSD values were higher in patients with papillitis than in those without (5.4 SD ± 0.5 versus 4.4 SD ± 0.5 mm) in another study [30,31].

Literature concerning ONSD measurements is typically discussed with concern for elevated intracranial pressures, but there are only a few reports describing the utility of ED ultrasound use in the diagnosis of ON. This case series confirms the findings of other similar case reports that ONSDs increased in ON and POCUS is a viable imaging modality in measuring ONSD [21,22]. By providing detailed information on presentation, diagnosis and outcomes of these patients, we attempted to further delineate the role of POCUS in the timely diagnosis of these patients. Characteristic clinical symptoms noted in the introduction are important, but nonspecific for the disease process and of limited diagnostic value in this case series [24]. In each of these cases, POCUS increased the providers index of suspicion for ON and led to a more timely diagnosis, underscoring the potential value of POCUS in this clinical setting.

It appears that there could be value in measuring the difference between affected eye and non-affected eye in a patient with unilateral symptoms given that there is some variance between healthy individuals. Dees et al. and Lochner et al. infer that “Nerve swelling is recorded if the ONSD is at least 0.3 mm larger than that of the contralateral unaffected nerve [32,33].” Moreover, there is the possibility that a relationship exists between the thickness of the nerve and the clinical features of the acute optic neuritis. That is, the greater the ONSD is, the more severe the visual acuity loss and the lower the probability of full recovery may be. Though this case series does not serve to confirm or contest such an association, it merits further investigation.

Lastly, it is imperative to acknowledge the limitations of POCUS in the emergency setting including the following factors: provider training, equipment availability, and inaccurate measurements. In addition this case series from a single center where ultrasound is not done on.

Table 1

| Case series summary - Ultrasound measurement of Optic Nerve Sheath Diameter (ONSD). |
|------------------|------------------|------------------|------------------|------------------|
| Age (years) | 35 | 48 | 34 | 28 |
| Sex | Male | Male | Female | Female |
| OD Ultrasound Image | ONSD = 6.7mm | ONSD = 8.6mm | ONSD = 5.5mm | ONSD = 5.6mm |
| OS Ultrasound Image | ONSD = 3.4mm | ONSD = 7.6mm | ONSD = 5.8mm | ONSD = 5.5mm |
| Intraocular Pressure | OD = 14mm Hg OS = 17mm Hg | OD = 17mm Hg OS = 14mm Hg | OD = 16mm Hg OS = 14mm Hg | OD = 12mm Hg OS = 11mm Hg |
such patients; as such, we do not know if there are any ON cases where POCUS would have been effective.

3. Conclusion

Point-of-care ultrasound is an accessible and effective means of visualizing the eye. In this case series, ED use of POCUS consistently demonstrated utility in measuring the ONSD, and it aided in expediting the diagnosis of acute ON in all cases. Moreover, because of the absence of radiation and rapidity of use, ultrasonography has considerable advantages over conventional imaging modalities. Employing POCUS in the setting of a patient with ON can streamline the ED physician’s decisions leading to ED neurology and ophthalmology consultations, and thus, guides patient care along its most effective route.

References