ANNALS of Clinical and Translational Neurology
Open Access
AMERICAN NEUROLOGICAL ASSOCIATION

INTERACTN CASE

The case of a 29-year-old woman with headaches and diplopia

Shubadra Priyadarshini

Northwestern Memorial Hospital, Chicago, Illinois

interACTN Case #28: Available: http://interactn.org/2020/11/09/case-28-case-of-a-29-year-old-woman-with-headaches-and-diplopia/

Summary of case

A 29-year-old woman with a history of shingles presented with a 3-month history of headaches and diplopia. She was in her usual state of health until 3 months prior to presentation, when she first developed severe headaches. Headaches were localized to the left periorbital area and left temple. They were described as "stabbing" in nature. Initially, she was having daily headaches, which would wake her up from sleep.

She denies any visual aura, photophobia, phonophobia, nausea, or emesis. She denies eye tearing, rhinorrhea, or conjunctivitis. She also denies fevers/chills and neck pain/stiffness. Headaches are not associated with changes in position.

About 2–3 weeks after onset of headaches, the patient developed binocular diplopia. In particular, she noted the diplopia was worse with leftward gaze and with objects greater than 30–60 cm away. Her boss also noticed her left eyelid was drooping around this time, which resolved by the time of presentation.

On exam, her left eye was unable to fully abduct with far-left gaze. On downward gaze, her left eye had a subtle hypertropia. Her left eye ptosis had resolved. Otherwise, the rest of her neurologic exam was intact. Her pupils were 4 → 2 mm bilaterally. Visual fields were intact to finger counting. On fundoscopic exam she had sharp disc margins OU. There was no ptosis bilaterally. Extraocular movements were intact in the right eye.

Her magnetic resonance imaging (MRI) brain showed T2 signal hyperintensity in the left cavernous sinus, with ill-defined, asymmetric enhancement with extension of mild enhancement (Figure 1). Vessel and sinus imaging was without any evidence of thrombosis. Her cerebrospinal fluid (CSF) studies were bland (white blood cell 2 and 1, protein 26, glucose 61, all infectious, cytology, and inflammatory markers negative). All serum studies were negative or within normal limits as well, including an autoimmune panel, Lyme disease, human immunodeficiency virus, and vasculitis labs. Her symptoms improved after a course of steroids.

Diagnosis: Tolosa–Hunt syndrome.

Take-home points:

A. The cavernous sinus contains several key anatomic structures, including cranial nerves III, IV, VI, and the opthalmic and maxillary branches of V. Cranial nerve VI runs medially, inferior to the cavernous portion of the internal carotid artery.
B. The differential for cavernous sinus lesions include solid neoplasms (meningiomas being most common), systemic

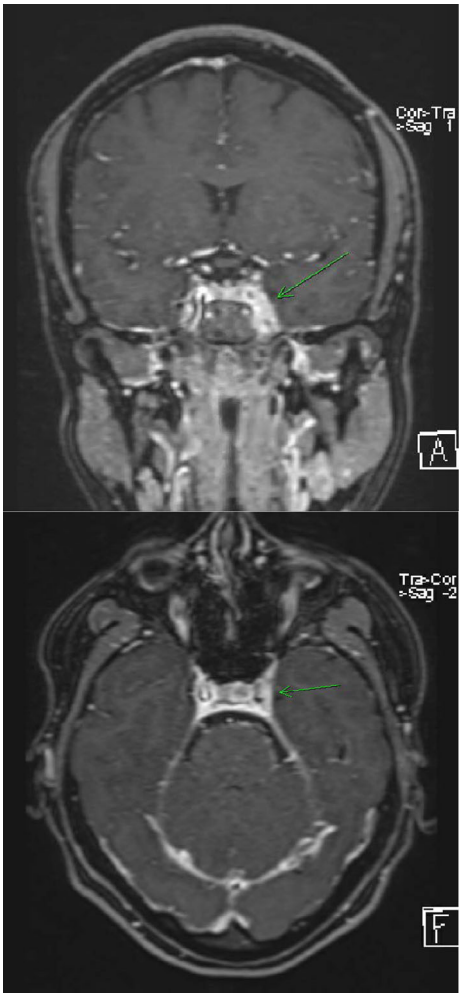

Figure 1. Coronal postcontrast (top) and axial postcontrast (bottom) MRI brain demonstrating asymmetric enhancement and enlargement of the left cavernous sinus

malignancies such as lymphoma, fungal and bacterial infections, and granulomatous disease. Among the granulomatous disease, the differential includes sarcoidosis, granulomatosis with polyangiitis, and Tolosa–Hunt.[1]

C. Tolosa–Hunt syndrome is an idiopathic, granulomatous inflammation of the cavernous sinus. Patients most typically present with unilateral, periorbital headaches, with cranial nerve palsies, most commonly oculomotor > abducens > trochlear and ophthalmic branch of the trigeminal nerve.[2]

D. The diagnosis of Tolosa–Hunt can be challenging, as painful ophthalmoplegia and the MRI findings of cavernous sinus lesions are present in numerous conditions. Therefore, it is important to undergo broad serum and CSF work-up, as well as follow patients routinely with serial imaging and potential repeat CSF.[2,3]

E. Treatment for Tolosa–Hunt is steroid regimen, but half of patients will have a remitting-relapsing course[2,3]

References

1. Kline LB, Hoyt WF. The Tolosa-Hunt syndrome. J Neurol Neurosurg Psychiatry 2001;71(5):577–582. https://doi.org/10.1136/jnnp.71.5.577. PMID: 11606665; PMCID: PMC1737614.
2. Zhang X, Zhou Z, Steiner TJ, et al. Validation of ICHD-3 beta diagnostic criteria for 13.7 Tolosa-Hunt syndrome: Analysis of 77 cases of painful ophthalmoplegia. Cephalalgia 2014;34(8):624–632. Epub 2014 Jan 29. Erratum in: Cephalalgia. 2015 Mar;35(3):285. PMID: 24477599.
3. Amrutkar C, Burton EV. Tolosa-Hunt syndrome. [Updated 2020 Aug 10]. In: StatPearls [Internet]. Treasure Island, FL: StatPearls Publishing; 2020. Available from: https://www.ncbi.nlm.nih.gov/books/NBK459225/

