Persistent focal enhancement of the cisternal segment of oculomotor nerve in ophthalmoplegic migraine

Ihtesham A. Qureshi, MD; Gustavo J. Rodriguez, MD; Salvador Cruz-Flores, MD; Alberto Maud, MD

Ophthalmoplegic migraine (OM) is a rare form of migraine characterized by recurrent episodes of headache with ophthalmoplegia related to paresis of cranial nerves III, IV, or VI.¹ Adult onset of ophthalmoplegic migraine is rare as patients usually have a history of typical migraine headaches early in life in addition to a positive family history.² OM is a diagnosis of exclusion requiring appropriate laboratory test and neuroimaging studies to ensure an accurate diagnosis.³ In most cases of OM, there is a transient enhancement of the apparent origin of the oculomotor nerve.^{1,4} The recent revision of the International Headache Classification has reclassified OM from a subtype of migraine to the category of neuralgia.⁵ OM is rare, with an estimated annual incidence of 0.7 per million.⁶

We describe a patient presenting with clinical features consistent with adult-onset OM and persistent focal enhancement of the oculomotor nerve over time. Although described previously in children, such presentation in the form of persistent enhancement of the cisternal segment of oculomotor nerve with ophthalmoplegic migraine is rare in adults.

Case report

A 53-year-old right-handed woman presented with pulsatile hemicranial pain around the right V1 distribution along with nausea, vomiting, sonophobia, and photophobia. She had a history of migraine attacks since childhood, but the attacks became more severe around 3–6 months prior to presentation; at the same time, she started to notice diplopia in horizontal gaze and ptosis in the right eye (figure 1). Severe attacks with diplopia and ptosis were as frequent as every 3 weeks. There is no known relevant family history. She had a history of autoimmune hepatitis and breast cancer in remission. The patient had no evidence of recurrence of breast cancer over the time she was followed. At the time of change of character of her migraines, verapamil 120 mg was added to her regimen. This has improved her condition, as both the frequency and attacks were decreased. She had attained menopause before the beginning of OM, so no hormonal relationship has been established. Her vital signs were normal and her general physical examination was unremarkable. Her neurologic examination revealed a paralysis of the right superior, medial, and inferior rectus muscles with almost complete ptosis. Pupils were equal and reacted to light and accommodation. She was treated with IV hydration, antiemetics, anti-inflammatories, and serotonin receptor agonist medication. Brain MRI with gadolinium demonstrated focal enhancement of the cisternal segment of the right oculomotor nerve (figure 2). Because of her history of breast cancer, lumbar puncture was performed to investigate meningeal neoplastic infiltration. CSF analysis including flow cytometry and cytologic analysis was negative. The patient was discharged on calcium channel

Practical Implications

Contrast enhancement of the cisternal segment of oculomotor nerve on MRI in patients with ophthalmoplegic migraine could persist over time in spite of the complete remission of the ocular symptoms.

Neurology Department, Texas Tech University of Health Sciences Center, El Paso.

Funding information and disclosures are provided at the end of the article. Full disclosure form information provided by the authors is available with the **full text of this article at Neurology.org/cp.**

Correspondence to: drqureshi786@hotmail.com

Figure 1 Patient photographs

The top 4 images reveal paralysis of the right superior, medial, and inferior rectus muscles with almost complete ptosis with preservation of pupil; the bottom 4 images reveal restoration of normal function of oculomotor nerve.

blocker for secondary prevention of her vascular headache attacks. During the next 6 months, a total of 3 CSF analyses at different times failed to reveal neoplastic meningeal infiltration. Brain MRI with gadolinium was repeated at 6 and 12 months and 2, 3, and 4 years since first diagnosis revealed persistent enhancement of the right oculomotor nerve at the cisternal segment. The first MRI was done 48 hours after the attack. Subsequent follow-up MRIs were performed while the patient was not actively having an attack. In between the attacks, her oculomotor function in the right eye was normal (figure 1).

DISCUSSION

The International Headache Society's Revised International Classification of Headache Disorders 2004 established criteria for OM, which include (1) at least 2 attacks of migraine-like headache accompanied or followed within 4 days of onset by paresis of one or more of III, IV, and VI cranial nerves; and (2) exclusion of parasellar, orbital fissure, and posterior fossa lesions by appropriate investigators.⁷

Other diagnoses to consider include multiple sclerosis, carcinomatous meningitis, Miller Fisher variant of Guillain-Barré syndrome, diabetes mellitus, Tolosa-Hunt syndrome (THS),

Figure 2 Brain MRI

Brain MRI with gadolinium shows focal enhancement of the cisternal segment of the right oculomotor nerve persistently visible in all years.

and aneurysm. Perhaps the most important consideration is THS, an idiopathic sterile inflammation of the orbital apex in adults. THS commonly presents with painful orbitopathy with ptosis, diplopia due to ocular cranial nerve involvement, variable pupillary changes, and preserved visual acuity. MRI can depict enhancement of the orbital apex at the level of the superior orbital fissure and it can occasionally extend to the anterior aspect of the cavernous sinus. The inflammation and ocular symptoms typically respond to steroids as well as the abnormal enhancement on the neuroimaging studies. Most importantly, OM attacks are paroxysmal in nature in addition to a lack of response to steroids and the complete resolution of the ocular findings after the headache attack resolves as compared to THS.²

Several case reports with OM as a diagnosis have shown postenhancement MRI that reveals reversible thickening and enhancement of cranial nerve III.^{4,5} Our patient showed persistent enhancement of the cisternal segment of oculomotor nerve that was never resolved after years of follow-up.

REFERENCES

1. Bharucha DX, Campbell TB, Valencia I, Hardison HH, Kothare SV. MRI findings in pediatric ophthalmoplegic migraine: a case report and literature review. *Pediatr Neurol* 2007;37:59–63.
2. Stommel EW, Ward TN, Harris RD. MRI findings in a case of ophthalmoplegic migraine. *Headache* 1993;33:234–237.
3. Walsh FB, Hoyt WF. *Clinical Neuro-ophthalmology*. Vol. 2, 3rd ed. Baltimore: Williams & Wilkins; 1969:1975.
4. O'Hara MA, Anderson RT, Brown D. Magnetic resonance imaging in ophthalmoplegic migraine of children. *J AAPOS* 2001;5:307–310.
5. Levin M, Ward TN. Ophthalmoplegic migraine. *Curr Pain Headache Rep* 2004;8:306–309.
6. Hansen SL, Borelli-Møller L, Strange P, Nielsen BM, Olesen J. Ophthalmoplegic migraine: diagnostic criteria, incidence of hospitalization and possible etiology. *Acta Neurol Scand* 1990;81:54–60.
7. Classification Committee of the International Headache Society. International Classification of Headache Disorders II. *Cephalgia* 2004;24(suppl 1):1–160.

Received May 17, 2016. Accepted in final form August 4, 2016.

AUTHOR CONTRIBUTIONS

I. Qureshi was involved in manuscript writing. G. Rodriguez was involved in critical revision of manuscript. S. Cruz-Flores was involved in critical revision of manuscript. A. Maud was involved in manuscript writing.

STUDY FUNDING

No targeted funding reported.

DISCLOSURES

I. Qureshi reports no disclosures. G. Rodriguez serves as an Associate Editor for *World Journal of Radiology*, *Journal of Neurology & Stroke*, and *Symbiosis Journal of Neurology*. S. Cruz-Flores has received funding for travel from AHA and AAN; serves as a consultant for Lilly, Sunovion, Quintiles, and Duke University; and receives research support from Astra Zeneca. A. Maud reports no disclosures. Full disclosure form information provided by the authors is available with the **full text of this article at Neurology.org/cp**.

Practical Implications

Neurology® Clinical Practice is committed to providing clinical insights that are helpful to neurologists in everyday practice. Each Full Case will include a “Practical Implications” statement, a pearl of wisdom for the practicing clinician.