“Ophthalmoplegic Migraine” With Reversible MRI Enhancement of the Cisternal Sixth Cranial Nerve

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Abstract: A 45-year-old woman reported multiple episodes of reversible left eye pain and diplopia stretching over 12 years. Ophthalmic examinations had repeatedly disclosed a left sixth cranial nerve palsy. Postcontrast brain MRI performed 3 weeks after clinical onset of the most recent episode demonstrated enhancement of the cisternal segment of the left sixth cranial nerve. Five months later, when symptoms and signs had largely abated, postcontrast brain MRI was normal. The clinical diagnosis satisfies the criteria for “ophthalmoplegic migraine.” Although reversible cisternal enhancement of the third cranial nerve has been often described in this condition, this is the first report of cisternal enhancement of the sixth cranial nerve.

A 45-year-old woman had episodic severe left eye pain and double vision for 12 years. The episodes occurred 2–3 times per year and symptoms usually lasted about 4 weeks. Each episode started abruptly with steady left periocular pain (rated 8 of 10 in severity) that often awakened her from sleep. She reported photophobia and phonophobia but no obvious pupil size changes, tearing, or nasal discharge. The pain usually abated a few hours after she received parenteral ketorolac, meperidine, or corticosteroids. After receiving treatment, she usually slept but always awoke to binocular diplopia that was horizontal and worse at distance and on left gaze. The pain never lasted for more than 2 or 3 days, but the diplopia usually lasted for at least 4 weeks.

Propranolol, topiramate, and indomethacin did not prevent the attacks. Abortive agents, including nonprescription medications and triptans, neither helped the headaches nor prevented the diplopia. She had a history of depression, hypertension, two cesarean deliveries, an appendectomy, and a fractured elbow. Her medications were valsartan, metoprolol, alprazolam, a daily vitamin, and topiramate (50 mg daily).

At the time of our first examination, the patient reported a typical episode starting 3 months earlier, with diplopia that was initially present in the primary gaze position and gradually improved so that it was present on left gaze. Visual acuity was 20/20 in each eye at distance and near viewing. Her eyes were aligned in primary position but she had only 75% abduction of the left eye. The right pupil was fractionally larger than the left, but both reacted briskly to light and near stimuli. The palpebral fissures were equal in height. The ophthalmologic and neurologic examinations were otherwise normal. When examined 2 months later, 5 months after the onset of the current episode, she had diplopia only on extreme left gaze and 90% abduction of the left eye.

Brain and orbit MRI, which was performed 3 weeks after the onset of the current episode, demonstrated enhancement of the left sixth cranial nerve in the pre-pontine cistern (Fig. 1A–C). Brain MRA showed no abnormality. A second brain MRI (Fig. 1D), performed 5 months after the commencement of the current episode, demonstrated no sixth cranial nerve enhancement.

“Ophthalmoplegic migraine” (OM), a rare enigmatic disorder with an annual incidence of about 1 per million (1), was reclassified by the International Classification of Headache Disorders, 2nd ed. (ICHD-III) (2) as a recurrent neuralgia under the category “cranial neuralgias and central causes of facial pain.” The onset of OM is usually, but not always, in the first decade. Involvement of the third cranial nerve is more common than involvement of the fourth or sixth cranial nerves, and simultaneous impairment of more than one ocular motor nerve is exceptional (1).

Many patients with OM clinically involving the third cranial nerve demonstrate enhancement or thickening, or both, of the cisternal segment of the third cranial nerve (3,4) that is usually reversible within 7–9 weeks (3) (Fig. 2). Isolated enhancement of the fourth cranial nerve has been reported once in this condition (5) (Fig. 3). There has also been one previous report of reversible enhancement of the intraparenchymal portion of the sixth cranial nerve in OM (Fig. 4) (6), but no previous report of enhancement of the sixth cranial nerve in the pre-pontine cistern.
cisternal portion of the sixth cranial nerve such as we are describing here.

MRI enhancement of the cranial nerves occurs with infectious, inflammatory, demyelinating, and neoplastic disorders. Enhancement of the cisternal portion of the sixth cranial nerve is reported with trauma, ischemia, venous congestion associated with a medullary venous malformation, leukemia, chemical meningitis, the Fisher variant of acute inflammatory demyelinating polyneuropathy (AIDP), polyneuritis cranialis (probably a variant of AIDP), multiple sclerosis, diabetes mellitus, meningoencephalitis, autoimmune disorders such as the Tolosa-Hunt syndrome (7), Lyme disease (8), and vincristine therapy (9). This is the first report of enhancement of the cisternal portion of the sixth cranial nerve in OM.

REFERENCES