Residents and Fellows: Teaching Images in Headache

Recurrent Painful Cranial Neuropathy in a Child Involving Multiple Cranial Nerves

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A 9-year-old boy with a history suggestive of "ophthalmoplegic" migraine presented with an episode of left frontal headache and diplopia. On examination, in addition to oculomotor weakness in the form of ptosis and pupillary dilation, he had symptoms of superior oblique palsy with difficulty looking "down and in" (Fig. 1A–C). Magnetic resonance imaging (MRI) of the brain revealed post-contrast enhancement of the cisternal segments of the third and fourth cranial nerves (Fig. 2A,B).

Ophthalmoplegic migraine is reclassified in the International Classification of Headache Diseases – 3rd edition as recurrent painful cranial neuropathy – reflective of the putative underlying pathology. The diagnostic criteria require that the child experience paresis of one or more the ocular motor nerves in association with unilateral headache. In clinical practice, the oculomotor nerve is most commonly affected,

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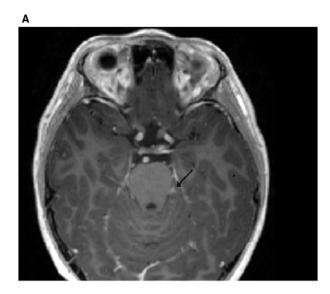




Fig. 1.—(A) Left superior oblique palsy with difficulty looking down and in. (B) Left superior oblique palsy with difficulty looking down and to the right. (C) Left oculomotor nerve weakness as noted by left-sided ptosis. [Color figure can be viewed at wileyonlinelibrary.com]

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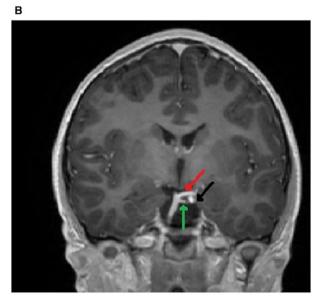


Fig. 2.—(A) Axial T1 post-contrast image shows enhancement of the left fourth cranial nerve in the perimesencephalic cistern. (B) Coronal T1 post-contrast image shows enhancement of the cisternal portion of the left third cranial nerve (black arrow) between the superior cerebellar artery (red arrow) and the posterior cerebral artery (green arrow). [Color figure can be viewed at wileyonlinelibrary.com]

followed by the abducens nerve. The trochlear nerve is believed to be involved in 8% of the individuals with recurrent painful cranial neuropathy. Clinical or radiographic involvement of the third and fourth cranial nerves is rarely if ever reported in the literature.

Imaging of the trochlear nerve is often challenging as it is the cranial nerve with the least number of axons and a diameter of 0.7-1 mm. Special sequences such as 3-dimensional (3D) T2-weighted post-contrast imaging on 3 Tesla MRI scanners with thin slices are often required.³

The most common causes for post-contrast enhancement of the trochlear nerve in the cisternal space include trauma, trochlear nerve sheath tumors including schwannomas, meningiomas, neurofibromas, meningitis, or following lumbar puncture. To our knowledge, this is the only report of the visualization of the trochlear nerve in a patient with recurrent painful cranial neuropathy.

Our patient was admitted to the hospital and received 5 days of pulsed intravenous steroids. A follow-up visit in the neurology clinic 2 weeks after discharge showed a significant improvement in the movements of the left eye.

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