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Recurrent Painful Cranial Neuropathy in a Child involving Multiple Cranial Nerves

Dinesh Lulla
Detroit Medical Center - Pediatric Neurology
3950 Beaubien Street
Detroit
Michigan
48201
United States
T: 3138329620 F: 3137453012
Email: dinesh.lulla5@gmail.com

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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" (**Figure 1 A,B and C**). Magnetic Resonance Imaging (MRI) brain revealed post contrast enhancement of the cisternal segments of the 3rd and 4th cranial nerves (**Figure 2 A and B**).

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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, followed by the abducens nerve.[1] The trochlear nerve is believed to be involved in 8% of individuals with recurrent painful cranial neuropathy.[2] Clinical or radiographic involvement of 3rd and 4th 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 to 1mm. Special sequences such as 3 dimensional (3D) T2 weighted post contrast imaging on a 3 Tesla MRI scanners with thin slices are often required.

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 visualization of the trochlear nerve in a patient with recurrent painful cranial neuropathy.

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

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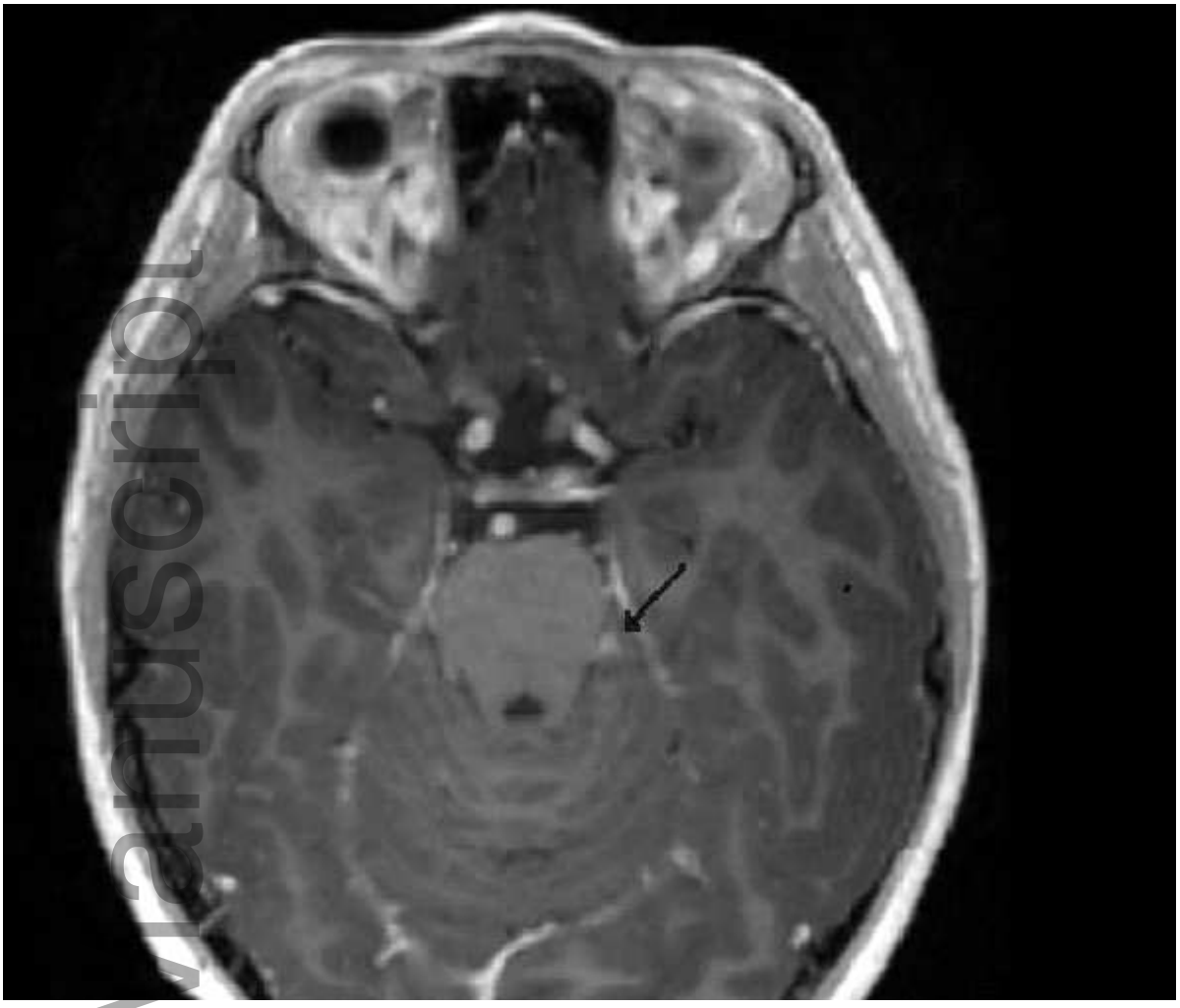

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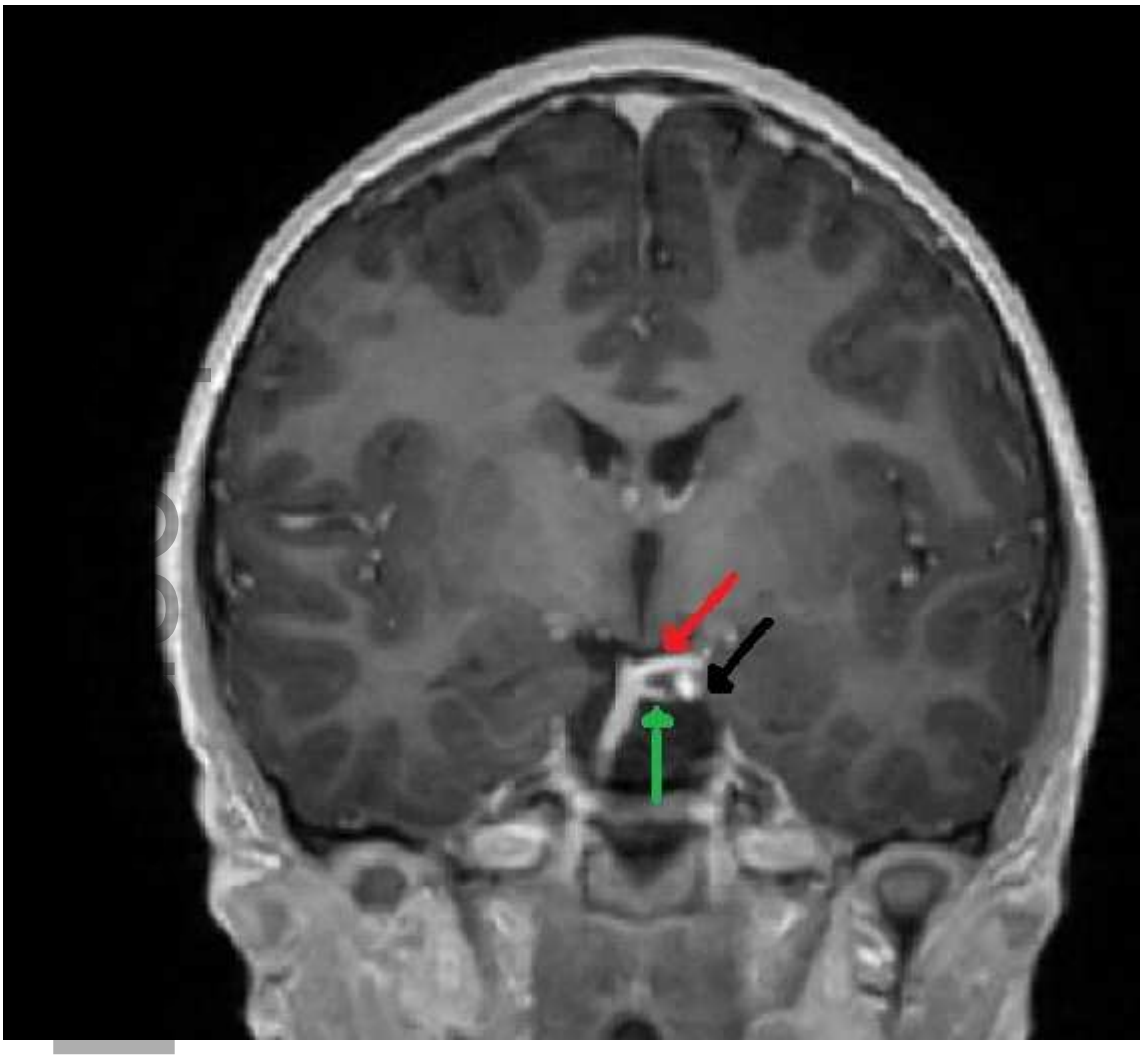

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