Oculomotor Nerve Enhancement in a Patient with Headache

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Abstract—Recurrent painful ophthalmoplegic neuropathy is a rare condition that is characterized by recurrent unilateral headache with diplopia. Commonly, MRI will show contrast enhancement of the involved cranial nerve(s) (III, IV and/or VI). Condition is very responsive to steroids. We are reporting an 18 year old male patient who presented with 3 episodes of left periorbital headache with diplopia secondary to left oculomotor palsy which enhanced on MRI. Diplopia completely resolved on receiving steroids.

Keywords — Diplopia, headache, ophthalmoplegic migraine, recurrent painful ophthalmoplegic neuropathy.

I. INTRODUCTION

Recurrent painful ophthalmoplegic neuropathy, previously known as “ophthalmoplegic migraine”, is a rare condition that usually manifests with recurrent unilateral headache with diplopia. Diplopia is secondary to ipsilateral palsy of one or more of the cranial nerves controlling extraocular muscles (III, IV, VI). Typically, MRI will show contrast enhancement of the involved cranial nerve(s). Condition responds well to steroids.

II. METHODS

A healthy 18 year old male presented to our ER with headache, diplopia and left eye ptosis. Headache started 3 days earlier, localized to the left frontotemporal and retro-orbital areas, and described as throbbing with photophobia and phonophobia. Headache preceded diplopia and ptosis by one day. Although headache eventually subsided, persistent diplopia caused the patient to seek medical attention.

He had two similar episodes of headache and diplopia within the past three years. Previous evaluations included normal non-contrast CT head, CT angiogram, orbit MRI with/without contrast (limited study due to orthodontic braces), and lumbar puncture. He was previously treated with oral steroids (Medrol Pack), with complete symptom resolution in few days. He neither had a known family history of headache syndromes nor ocular disorders.

Initial neurological examination showed right pupil of 3 mm and left pupil of 5 mm with no reactivity to light on the left side. Left eye's adduction and elevation were restricted. Patient had 2 mm ptosis of the left eye. Rest of neurological exam was otherwise normal. MRI brain with contrast revealed focal nodular thickening and enhancement of the left oculomotor nerve as it exits the midbrain (Fig.1). MRA head was unremarkable. He received 500 mg methylprednisolone intravenously followed by oral steroid taper, with normalization of ocular function in 1 day.

III. DISCUSSION

Recurrent painful ophthalmoplegic neuropathy is a rare condition. It was previously known as ophthalmoplegic migraine. The typical clinical presentation includes migraine-like headache accompanied by longer-lasting diplopia. If the oculomotor nerve is involved, pupillary abnormalities and ptosis are common. There is usually a 1-4 day lag between headache onset and diplopia. Diplopia and ocular findings usually outlast the headache too. This disorder most commonly occurs in children, but adult onset cases have been reported.

Both clinical observations and gadolinium enhancement of involved nerves on MRI have allowed reclassification of this disorder from migraine variant to the category of painful cranial neuropathies¹,²,³.

Transient enhancement of the cisternal segment of the oculomotor nerve is seen on MRI in 76% of cases. Recurrent demyelinating cranial neuropathy has gained favor within recent years as an explanation for this common finding². Multiple observations showed benefit of steroids in treating acute exacerbations. However, due to the rarity of this condition there are no published treatment trials.

According to The International Classification of Headache Disorders ICHD (3rd edition)¹, this condition has the following diagnostic criteria:
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A. At least two attacks fulfilling criterion B.
B. Unilateral headache accompanied by ipsilateral paresis of one, two or all three oculomotor nerves.
C. Exclusion of orbital, parasellar, or posterior fossa lesions by appropriate imaging.
D. Not better described by another ICHD-3 diagnosis.

Our patient fulfilled all criteria for ophthalmoplegic neuropathy. His brain MRI documented common findings of thickening and gadolinium enhancement of the affected oculomotor nerve. He had complete resolution of the ocular symptoms following corticosteroid treatment, reportedly of therapeutic benefit in some patients.\(^1,2\).

The Differential diagnoses of oculomotor nerve palsy include:

a) Posterior communicating artery aneurysm.
b) Diabetic third nerve palsy.
c) Cavernous sinus thrombosis or Tolosa Hunt syndrome.
d) Stroke or demyelination in the midbrain.
e) Compression of the oculomotor nerve by a mass lesion or uncal herniation.
f) Migraine

It’s of extreme importance that we rule out these causes which can be dangerous in certain situations. In our patient, these causes were ruled out on the MRI brain with/without

Figure 1. T1-weighted Magnetic Resonance Imaging without (A, C) and with (B, D) contrast demonstrated enhancement of the cisternal segment of the left CN III.
contrast and MRA brain. Besides, our patient did not have diabetes and his glucose levels were always normal.

One interesting differential diagnosis is Tolosa Hunt syndrome. There are some case reports in the literature of Tolosa Hunt syndrome with extra-cavernous sinus granulomatous tissue involving cranial nerves (like II, III, VI, VII, etc.) in addition to the evidence of disease inside the cavernous sinus. Our patient had no evidence of granulomatous inflammation in the cavernous sinus, superior orbital fissure or orbit on the MRI, which takes away an essential part of the criteria for diagnosis of Tolosa Hunt syndrome according to ICHD-3rd edition. In our patient, inflammation was exclusively involving the oculomotor nerve with focal nodular thickening of the nerve as it exits the midbrain (the cisternal segment). Another feature that makes Tolosa Hunt unlikely in our patient is that cranial nerve deficits in Tolosa Hunt usually resolve over 1-8 weeks, and our patient’s ocular symptoms and signs resolved in 1 day after he received IV steroids.

While response to steroids is observed in both migraine variant and painful ophthalmoplegic neuropathy, focal enhancement of the cranial nerve is the key differentiating characteristic of the latter.

Overall prognosis of recurrent painful ophthalmoplegic neuropathy is good, and symptoms usually resolve after a few days or weeks. However, there are reports of persistent deficits following multiple ophthalmoplegic episodes.

**Questions (please choose one single answer):**

1) Recurrent painful ophthalmoplegic neuropathy (previously known as ophthalmoplegic migraine) can present with unilateral headache and palsy of which cranial nerves?
   - A) III
   - B) IV
   - C) VI
   - D) Any of the above cranial nerves or a combination of them.

2) Regarding clinical presentation, which symptoms/signs resolve first:
   - A) Ocular symptoms/signs resolve before resolution of headache.
   - B) Both ocular symptoms/signs and headache resolve around the same time.
   - C) Ocular symptoms/signs persist for a brief period after the resolution of headache.

3) Most common finding on MRI in this condition is?
   - A) Meningeal enhancement.
   - B) A lesion involving the nucleus/nuclei of the cranial nerve (s) involved.
   - C) Enhancement of the involved cranial nerve (s).
   - D) Aberrant vessel compressing the cranial nerve (s) involved.

4) Best treatment for this condition is:
   - A) Lamotrigine
   - B) Steroids
   - C) Indomethacin
   - D) Carbamazepine

**Correct answers:** 1: D; 2: C; 3: C; 4: B

**REFERENCES**


