

Magnetic Resonance Findings in Recurrent Painful Ophthalmoplegic Neuropathy with Reversible Enhancement of Oculomotor Nerve

¹Ali Sami Kivrak, ¹Alaaddin Nayman, ¹Yahya Paksoy, ²Serefnur Ozturk

ABSTRACT

Recurrent painful ophthalmoplegic neuropathy is a rare disorder characterized by repeated attacks of paresis of one or more ocular cranial nerves, with ipsilateral headache. We describe a 24-year-old man with recurrent painful ophthalmoplegic neuropathy and oculomotor nerve palsy, in whom clinical features and magnetic resonance imaging findings on pre and post treatment period.

Key words: Ophthalmoplegic migraine, recurrent painful ophthalmoplegic neuropathy, MRI, oculomotor nerve

Geçici Okulomotor Sinir Kontrastlanması ile Birlikte Tekrarlayıcı Ağrılı Oftalmoplejik Nöropatide Manyetik Rezonans Bulguları

ÖZET

Tekrarlayan ağrılı oftalmoplejik nöropati ipsilateral baş ağrısı ile birlikte bir veya daha fazla oküler kranial sinirlerinin tekrarlanan parezi atakları ile karakterize nadir bir hastalıktır. Biz tekrarlayan ağrılı oftalmoplejik nöropati ve okulomotor sinir felci olan 24 yaşındaki bir erkek hastanın tedavi öncesi ve sonrası MRI/klinik bulgularını tanımladık.

Anahtar kelimeler: Oftalmoplejik migren, tekrarlayan ağrılı oftalmoplejik nöropati, MRG, okulomotor sinir

INTRODUCTION

Ophthalmoplegic migraine, according to the new classification, renamed as recurrent painful ophthalmoplegic neuropathy, is a rare disorder characterized by the association of recurrent attacks of migraine headache and involvement of cranial nerves III, IV, or VI. The oculomotor nerve is the most commonly affected. The disorder usually presents in children, but may also effect young adults. The episodes of ophthalmoplegia may persist for several hours to several weeks, months or in rare cases may be constant (1-7). Because of the self-limiting and reversible nature of this disease, the pathophysiology of this condition is still unclear although there is current evidence to suggest that it is a demyelinating process (1,2,5).

Previous reports show that ophthalmoplegic migraine may or may not be associated with changes seen on magnetic resonance imaging (MRI) (1-6). Contrast-enhanced MRI performed during symptomatic and postsymptomatic periods in patients with ophthalmoplegic migraine and knowledge of imaging findings may hold great value in providing prompt differential diagnosis.

Clinical features in a patient with recurrent painful ophthalmoplegic neuropathy and the findings of the MRI findings which were obtained pre and post treatment period are discussed.

¹Selcuk University School of Medicine, Department of Radiology, Konya, Turkey-

²Selcuk University School of Medicine, Department of Neurology, Konya, Turkey

Correspondence: Ali Sami Kivrak,
Department of Radiology, Selcuk University School of Medicine,
Alaeddin Keykubat Campus, 42075, Konya, TURKEY.
Ph: +90 332 2415000 Fax: +90 332 241 21 84 e-mail: alisamikivrak@hotmail.com

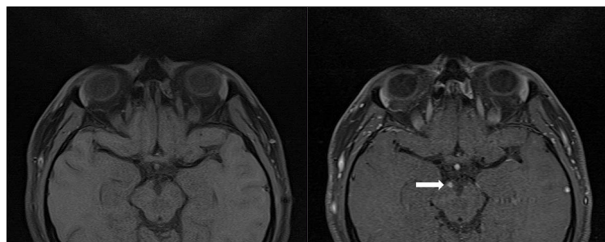


Figure 1. Axial pre-contrast T1 weighted MR image (a) not showing oculomotor nerve involvement, but axial post-contrast T1 weighted MR image (b) showing thickening and enhancement of the cisternal portion of the left oculomotor nerve (arrow).

CASE

A 24-year-old man with a long history of migraine presented with 7-day history of acute-onset headache on the right supraorbital side associated with anisocoria and diplopia. The findings of oculomotor paresis occurred within 2 days after the onset of headache without photophobia and vomiting. Since five years old, he had encountered one or two attacks of migraine with and/or without aura monthly. There was no prior episode of headache associated with oculomotor nerve palsy in history of the patient. Physical examination revealed a complete palsy of the right oculomotor nerve, with restriction of downward gaze and adduction of the right eye. Ptosis was not present. The right pupil was dilated and nonreactive. The optic disc was normal.

Brain MRI and MR angiography performed on the day of admitted to the hospital. Non-contrast and contrast-enhanced MRI examinations were performed with a 1.5-T unit (Excelart, Toshiba, Tokyo, Japan) using conventional sequences in addition to the three-dimensional fast asymmetric spin-echo (3D-FASE) sequences and 3D constructive interference in the steady-state (3D-CISS) sequences, which are the slice thickness was 1.0 mm. Brain MRI revealed a focal thickened cisternal portion of the right oculomotor cranial nerve at the root exit zone that enhanced intensely after contrast medium. Enhancement of the cavernous sinus or the adjacent dura was not seen. MR angiography helped to exclude any intracranial vascular malformation. He was treated with methylprednisolone 1000 mg/d IV for 3 days. On follow-up examination within 2 week of symptom on-

set, the patient's symptoms have almost completely resolved. Control MRI performed at 4 months after the first MRI study demonstrated almost complete resolution of oculomotor nerve thickening and enhancement.

DISCUSSION

Recurrent painful ophthalmoplegic neuropathy (Ophthalmoplegic migraine) is a rare episodic childhood condition in which a unilateral oculomotor palsy is preceded by headache. In the International Classification of Headache Disorders (3rd edition), recurrent painful ophthalmoplegic neuropathy is described as at least 2 attacks characterized by a "migraine-like" headache without aura followed within 4 days by paresis cranial nerves III, IV, and VI, including ophthalmoparesis, ptosis, or mydriasis (7). This clinical condition should not be attributed to another disorder such as tumor, infection, and aneurysm. Thrombosis must have been excluded by appropriate imaging (1,5). Recurrent painful ophthalmoplegic neuropathy is a diagnosis of exclusion. Patients with symptoms of oculomotor nerve palsy frequently examined with noninvasive imaging tests such as computed tomography, MRI, or MR angiography studies to eliminate these possibilities (1,2). Recurrent painful ophthalmoplegic neuropathy is a self-limited condition with a low frequency of recurrence and in the majority of patients with resolution without permanent sequelae (6).

Recurrent painful ophthalmoplegic neuropathy is frequently associated with changes revealed by enhanced MRI, most commonly with reversible enhancement and thickening of cisternal portion of the oculomotor nerve. Other conditions that may lead to solitary oculomotor nerve thickening and enhancement include neoplastic (e.g., schwannomas, hemangiomas, lymphoma), inflammatory and infiltrative conditions (e.g., sinusitis, basilar meningitis) and trauma (1,2,6). MRI indicates characteristic, reversible focal thickening and enhancement of the cisternal tract at the root exit zone of the third cranial nerve, which is usually the nerve most frequently affected (3). The inclusion of MRI findings of enhancement of an edematous third cranial nerve at the root exit zone in patients with recurrent, painful ophthalmoplegia and recurrent headache was suggested as a supportive diagnostic criterion in recurrent painful ophthalmoplegic neuropathy (3). If MR images shows enhancement of the cisternal segment of the oculomotor

nerve, further imaging may not be necessary (2).

Effective treatment methods for recurrent painful ophthalmoplegic neuropathy are not clear in literature. Nevertheless, oral steroids may be of possible benefit in treating acute exacerbations based on available observational data. Because recurrent painful ophthalmoplegic neuropathy is not a variant of migraine, migraine-specific acute and preventive therapies probably do not have a role in treatment (5). Contrast-enhanced MRI is a useful adjunct in the diagnosis and monitoring response to treatment of recurrent painful ophthalmoplegic neuropathy with oculomotor nerve involvement.

REFERENCES

1. Bharucha DX, Campbell TB, Valencia I, et al. MRI findings in pediatric ophthalmoplegic migraine: a case report and literature review. *Pediatr Neurol* 2007; 37:59-63.
2. Mark AS, Casselman J, Brown D, et al. Ophthalmoplegic migraine: reversible enhancement and thickening of the cisternal segment of the oculomotor nerve on contrast-enhanced MR images. *AJNR Am J Neuroradiol* 1998; 19:1887-91.
3. Miglio L, Feraco P, Tani G, et al. Computed tomography and magnetic resonance imaging findings in ophthalmoplegic migraine. *Pediatr Neurol* 2010; 42:434-6.
4. Lal V, Sahota P, Singh P, et al. Ophthalmoplegia with migraine in adults: is it ophthalmoplegic migraine? *Headache*. 2009; 49:838-50.
5. Gelfand AA, Gelfand JM, Prabakhar P, et al. Ophthalmoplegic "migraine" or recurrent ophthalmoplegic cranial neuropathy: new cases and a systematic review. *J Child Neurol* 2012; 27:759-66.
6. Ramelli GP, Vella S, Lövblad K, et al. Swelling of the third nerve in a child with transient oculomotor paresis: a possible cause of ophthalmoplegic migraine. *Neuropediatrics* 2000; 31:145-7
7. Headache Classification Committee of the International Headache Society (IHS). *The International Classification of Headache Disorders*, 3rd edition (beta version). *Cephalalgia* 2013; 33:629-808.