Ophthalmoplegic migraine in a child, an accelerated clinical and radiologic response to steroid therapy

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Abstract

Ophthalmoplegic migraine is characterized by recurrent attacks of migraine-like headache with paresis of ocular cranial nerves. To date, the exact etiology of ophthalmoplegic migraine remains unknown. We report a 9-year-old girl with typical clinical features of ophthalmoplegic migraine. She presented to us shortly after onset of her fifth episode. The initial episodes of ophthalmoplegia used to last for about 2-3 months with gradual and complete recovery. Brain MRI with contrast study revealed a thickened, enhancing right oculomotor nerve in the cisternal segment during the acute phase of ophthalmoplegia. She was treated with steroid for two weeks as well as with divalproex sodium for prophylaxis of migraine. There was complete recovery of ophthalmoplegia after four weeks of treatment with complete resolution of third nerve enhancement on repeat imaging. There were no further episodes of ophthalmoplegia within a follow up period of one year. Steroid therapy may hasten the recovery of ophthalmoplegia and prophylactic treatment of migraine may reduce the episodes and severity of ophthalmoplegic migraine.

INTRODUCTION

Ophthalmoplegic migraine is a rare disorder characterized by childhood onset and ophthalmoplegia following migrainous headaches. According to the International Headache Society (ICHDII) criteria, it is diagnosed when at least two attacks of headache with migrainous characteristics occur and are associated with or followed within 4 days of onset by paresis of one or more of the third, fourth or sixth cranial nerves and in the absence of any demonstrable intracranial lesion other than MRI changes within the affected nerve.1 The third cranial nerve is commonly involved in recurrent attacks and the involvement of the sixth and fourth nerves is uncommon. It was first recognized by Charcot in 1890.2 Ophthalmoplegic migraine is an uncommon presentation with an annual incidence of 0.7 per million.3

We report in this communication a patient with typical clinical features of ophthalmoplegic migraine who had reversible MRI contrast enhancement of third cranial nerve. There was faster recovery of her ophthalmoplegic attack with steroid therapy and a decrease in frequency as well as severity of migraine attacks with divalproex sodium treatment.

CASE REPORT

A 9 year-old female had recurrent attacks of headache for the last three years. The pain was right hemicranial, largely retroorbital throbbing/pulsatile and moderate to severe in intensity. It was associated with photophobia, phonophobia and vomiting. After the initial attack she had similar attacks at the interval of 15 days to one month lasting for 12-24 hours.

Five attacks of her headache were followed after 2-3 days by drooping of right eyelid with difficulty in moving the eyeball and diplopia. She presented to us shortly after onset of her fifth episode. The initial episodes of ophthalmoplegia used to last about 2-3 months and were followed by gradual and complete recovery.

She had no associated weakness of the extremities, face, numbness, abnormal body movement, protrusion or reddening of eye on the affected side or pain during eye movement. There was no history of fever, loss of consciousness, seizure like activity, joint pain, photosensitivity or rash.

Her general examination was normal and there were no focal neurological signs except ptosis and adduction paralysis of the right eye with dilated and non-reactive pupil (Figure 1).
The laboratory investigations included complete blood count, erythrocyte sedimentation rate, routine chemistries, liver and renal function test and cerebrospinal fluid study were normal. Brain MRI and MR angiography were within normal limits. Brain MRI with contrast study revealed a thickened, enhancing right oculomotor nerve in the cisternal segment (Figure 2a and 2b). Repeat MRI study after 3 months showed resolution of oculomotor nerve enhancement.

She was treated with prednisolone 1mg/ kg body weight for two weeks and divalproex sodium 250mg per day. There was complete recovery of ophthalmoplegia after four weeks and the headache frequency and severity decreased. There was no recent episode of ophthalmoplegia within a follow up period of one year.

DISCUSSION

The pathophysiology of ophthalmoplegic migraine remains obscure. This condition was included as a migraine variant in the first Headache Classification of the International
Headache Society in 1988. Based on postcontrast enhancement seen on MRI in some patients, ophthalmoplegic migraine is considered as recurrent inflammatory demyelinating illness of idiopathic or post-viral origin\textsuperscript{1,4,5}; therefore, it was moved out of the “migraine” group and repositioned as a “neuralgia” in the revised 2004 classification. Activation of the trigeminovascular system during an attack of migraine releases neuropeptides near the vessel wall. This causes a sterile inflammation of the wall of vasa nervorum leading to a breach in the blood nerve barrier, which is formed by the endothelium of the vasa nervorum. This leads to nerve edema and injury. Imaging may or may not show enhancement. Once the attack of ophthalmoplegic migraine subsides, the decrease in neurogenic inflammation of the vessel wall leads to restoration of the blood nerve barrier and decrease in nerve edema and enhancement. Thus, this hypothesis accounts for all facets of ophthalmoplegic migraine including antecedent severe migraine headaches and nerve enhancement. Migraine is considered secondary to irritation of fibers of the fifth nerve which accompany the inflamed third nerve.\textsuperscript{5,6}

In fact, a number of sinister conditions including vascular aneurysms, carotid dissection, carotid-cavernous fistula, infarcts due to vasculitis and neoplasm’s like primary intracranial tumors and metastatic disease can lead to a similar presentation. Other differentials that could be considered are orbital pseudo tumor, sarcoidosis, Tolosa-Hunt syndrome, and mycobacterial infection.\textsuperscript{7} Our patient’s laboratory examination was within normal limits, thus excluding the other causes. Furthermore none of these conditions resolve spontaneously.

Optimal treatment of ophthalmoplegic migraine is still unclear. Corticosteroids have been used with mixed results and the dose or the duration of treatment is not well established. For now, early high dose steroids are recommended to rapidly resolve an acute episode.\textsuperscript{5,9} Our patient’s ophthalmoplegia also resolved within one month with steroid therapy, while previously used to take two to three months for resolution of symptoms. Ideally, prophylactic therapy would prevent the occurrence of repeated episodes and prevent the development of permanent eye muscle palsies, but the report suggests that therapy has met with only limited success.\textsuperscript{10} Migraine prophylactic medications such as beta-blockers and calcium channel blockers have been proposed as treatment for ophthalmoplegic migraine.\textsuperscript{11} Our patient was started on valproex sodium prophylaxis. Divalproex sodium reduces the firing of serotonin-containing neurons in the dorsal raphe nucleus and reduces activation and sensitization of trigeminal nerve. There was a decrease in frequency as well as severity of migraine attacks with prophylactic treatment. In conclusion, our patient suggests that uncontrolled migraine may be the cause of ophthalmoplegic migraine and prophylaxis treatment of migraine may reduce the frequency of ophthalmoplegic attacks.

REFERENCES