CASE REPORT

Ophthalmoplegic migraine with trigeminal nerve involvement

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SUMMARY

We report a 25-year-old man with a history of uncontrolled migrainous headaches who developed third nerve palsy and sensory loss over V1 distribution of trigeminal nerve, during an attack of severe migraine. Gadolinium-enhanced MRI of the brain and cavernous sinus was normal and did not disclose nerve enhancement. CT angiogram was also normal. The patient recovered uneventfully in 2 weeks on oral steroids. The commonest cranial nerve implicated in ophthalmoplegic migraine is the oculomotor nerve. Involvement of the fifth nerve has never been reported.

BACKGROUND

In ophthalmoplegic migraine (OM) third cranial nerve is commonly involved with pupillary dilation but in adult cases pupillary involvement is uncommon. Sixth and fourth cranial nerve can be involved rarely, but other cranial nerve involvement has not been reported. We report a case of OM of adult onset with pupillary involvement which is rare but associated fifth nerve involvement (V1 distribution) in our case further adds to its rarity.

CASE PRESENTATION

A 25-year-old man with a history of left temporal migrainous headaches of 5 years duration presented with acute episode of headache associated with drooping of the left eye lid for 3 days. The headache was severe in intensity, predominantly left periorbital and temporally pulsating and associated with nausea, photophobia and phonophobia. The frequency of his past migrainous headaches was 6–8 episodes per year. The patient used to take non-steroidal anti-inflammatory drugs and antiemetics during attacks. The patient denied taking any prophylaxis. The migrainous headaches had increased in frequency over the last 6 weeks and were occurring daily for the last 3–4 days before developing ptosis. He had a history of similar ptosis of the left eye lid associated with similar migrainous headaches 2 years ago, which had resolved in less than 2 weeks. Details of treatment are unknown.

Examination of the patient during the present episode disclosed left incomplete third nerve palsy with involvement of the pupil. In addition, the patient also had sensory loss over V1 distribution on the left side including the left forehead, eye lid and medial aspect of the nose. Rest of the complete general physical and neurological examination was within normal limits.

INVESTIGATIONS

All relevant investigations including complete blood haemogram, biochemistry, thyroid profile and collagen profile including lupus erythematosus (LE), antinuclear antibodies (ANA) antineutrophil cytoplasmic-antibody (ANCA) were within normal limits. His cerebrospinal fluid (CSF) study was also normal. Relevant normal radiology included a normal chest x-ray and ultrasound abdomen. Contrast MRI of the brain and cavernous sinus was within normal limits (figure 1) and did not show any nerve enhancement. CT angiogram did not disclose any aneurysm.

DIFFERENTIAL DIAGNOSIS

- ▶ Tolosa-Hunt syndrome
- ▶ Cavernous sinus thrombosis
- ▶ Posterior communicating artery aneurysm

TREATMENT

In view of the above history, findings and a normal MRI, a diagnosis of adult onset ophthalmoplegic migraine (OM) was considered. He was treated with analgesics, short course steroids and propranolol.

OUTCOME AND FOLLOW-UP

The patient recovered completely in 10 days and was discharged on migraine prophylaxis. In 6 month follow-up, patient's headaches were well controlled on propranolol and amitriptyline without further episodes of OM.

DISCUSSION

OM was first described by Charcot in 1890.^{1 2} It was considered a variant of migraine in the past. However, more recently, it has been reclassified as a recurrent inflammatory demyelinating neuropathy

Figure 1 MRI (gadolinium-enhanced) brain and cavernous sinus within normal limits.

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of idiopathic or postviral origin because of the presence of reversible enhancement of the third cranial nerve.^{2 3} Based on a survey of the literature on OM in the postimaging era, enhancement on magnetic resonance is not a sine qua non for diagnosis of OM.¹ Recent case reports testify to the lack of enhancement of the involved cranial nerves, thus challenging the viral hypothesis.^{2 4-6} Moreover, OM often occurs during a severe attack of migraine. This prompts suspicion of involvement of the migrainous process in pathogenesis of OM.² Lal proposed a reversible breakdown in blood nerve barrier because of neurogenic inflammation of vasa nervosa, a hypothesis supported by Ambrosetto.⁷ We report a similar case that developed two attacks of ophthalmoplegia associated with severe migraine, the last attack involving the fifth nerve along with the third nerve.

OM is a rare disorder characterised by recurrent ophthalmoplegia and childhood onset.^{2 3 5 6} The third nerve is the commonest cranial nerve involved.^{2 8 9} Gadolinium-enhanced MRI may or may not disclose enhancement of the involved nerves.^{2 4 5} Lal *et al*^{2 10} described a new variant of OM characterised by single attacks of sixth nerve palsy in adults. In the series of 62 patients, 14 patients had two or more attacks fulfilling International Headache Society (IHS) criteria. None of them had enhancement of the nerves. An overwhelming majority had increased severity of migrainous headaches before developing ophthalmoplegia during a severe episode of headache.^{2 10} However, no patient had involvement of the fifth nerve as was seen in our case.

Since all the episodes of migraine were not associated with ophthalmoplegia in our patient and many episodes of ophthalmoplegia with headaches may not be of migraine type headaches as reported by Gelfand *et al*,¹¹ it is possible that some patients can have migraine as a coincidence that present with ophthalmoplegia and that it might not necessarily have a causal relationship to migraine, but that migraine might rather be a risk factor to develop ophthalmoplegic cranial neuropathy. Our patient had all the features of OM including a history of migrainous headaches, increase in severity of migrainous headaches before developing ophthalmoplegia during a severe attack. He also had one episode of ophthalmoplegia (involving left third nerve) during an attack of migraine 2 years ago (total 2 episodes), hence fulfilling IHS criteria³ for OM.

Owing to the presence of reversible nerve enhancement, OM is currently considered to be a demyelinating neuropathy of viral or idiopathic aetiology.^{2 3} Gelfand *et al*¹¹ in their systemic review found that in up to one-third of patients, the headache was not migrainous or associated with migrainous symptoms. In three-quarters of the cases involving the third nerve, there was focal nerve thickening and contrast enhancement on MRI and proposed that OM probably was not a variant of migraine but rather a recurrent cranial neuralgia or ophthalmoplegic cranial neuropathy.

However, the uniform absence of a viral prodrome, normal CSF and absence of nerve enhancement points go against this hypothesis.^{2 4-6} 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 who were diagnosed with OM, there was a suggestion that this could be an inflammatory/demyelinating disorder; therefore, it was moved out of the 'migraine' group and repositioned as 'neuralgia' in the revised 2004 classification.¹

However, most of the patients of OM are associated with severe migraine. This phenomenon suggests that migraine may play a role in its pathogenesis.² Attacks of migraine are

associated with release of neuropeptides in the wall of intracranial vessels, including vasa nervosa. These neuropeptides lead to sterile inflammation of the vessel wall of the vasa nervosa. This causes damage to the blood nerve barrier which is formed by the endothelial cells of the vasa nervosa, producing nerve oedema and dysfunction. Reversal of this process leads to recovery of nerve function.² Thus, OM may be related to an attack of migraine irrespective of presence or absence of enhancement and we hypothesise that probably the same mechanism is responsible for trigeminal nerve involvement in our patient.

To conclude, OM is uncommon and occurs during childhood and may or may not be associated with nerve enhancement. However, the adult variant has fewer attacks and may not show enhancement of the involved cranial nerves. As our case demonstrates, fifth nerve involvement can occur in OM.

Learning points

- ▶ Ophthalmoplegic migraine (OM) is a condition which begins almost always in childhood and adult cases are rare though OM can occur for the first time in adult life.
- ▶ Oedematous wall of cavernous part of internal carotid artery compressing the third nerve leading to oculomotor palsy has been postulated. Depending on the presence or absence of pupillary involvement, a compressive or ischaemic aetiology is suspected.
- ▶ Though postviral inflammatory demyelinating hypothesis has been postulated, absence of viral prodrome, absence of nerve enhancement in many cases and normal cerebrospinal fluid in all the patients points away from this hypothesis.
- ▶ Though the commonest cranial nerve implicated in OM is the oculomotor nerve one should thoroughly examine the patient for other cranial nerve involvement including fifth nerve.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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