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Correspondence and Clinical Notes

Clinical Notes

Ophthalmoplegic Migraine and Infundibular Dilatation of a Cerebral Artery

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Ophthalmoplegic migraine (OM) is a childhood disorder of uncertain etiology manifesting recurrent unilateral headache associated with a transitory oculomotor (usually IIIrd nerve) palsy. Recent publications emphasize the finding on MRI of contrast enhancement in the IIIrd nerve suggesting that OM may be a recurrent inflammatory neuropathy.

We report the case of a 7-year-old boy with typical symptoms of this disorder. Angio MR and Angio CT revealed the presence of an infundibular dilatation of a perforating branch of the posterior cerebral artery adjacent to the symptomatic IIIrd nerve. We speculate that this and perhaps other cases of OM may have a different pathophysiology related to compression of the IIIrd nerve by an adjacent vascular structure that could activate the trigeminovascular system and produce migrainous pain.

Key words: ophthalmic migraine, headache, cerebral artery

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A 7-year-old boy presented following several episodes of IIIrd nerve palsy that had first begun when he was 9 months old. The patient seemed distressed, probably in pain. The ophthalmoplegia persisted initially for 2-5 days. He had 4 recurrences up to the age of 3.

By this time the patient was able to report frontotemporal and orbital pain, always on the right side. The pain is moderate in intensity and constant and usually lasts all day. Analgesics provide partial relief and allow him to fall asleep.

He has nausea and occasionally vomits during the first few days of a episode. Photophobia and phonophobia are also present. Usually at the onset of pain, a IIIrd nerve palsy becomes apparent, with ptosis, external ophthalmoplegia, and mydriasis.

The pain subsides after 3-7 days; the ophthalmoplegia resolves within 1-4 weeks. The symptom-free interval varies from weeks to months.

Regarding family history, it should be noted that the child's mother has infrequent migraine without aura.

On neurologic examination during one episode of ophthalmoplegic migraine (OM), there is paralysis of all the muscles innervated by the oculomotor nerve, including a palpebral ptosis. A slight but unequivocal mydriasis is also present. The VIth nerve is not affected and sensation over the face and skull is preserved. No other abnormalities were seen. General examination was normal. Blood pressure was also normal.

The child had a clear response to steroids (oral prednisone, 1 mg/kg/day). This treatment was used twice and the pain subsided much earlier (within 24-48 hours).

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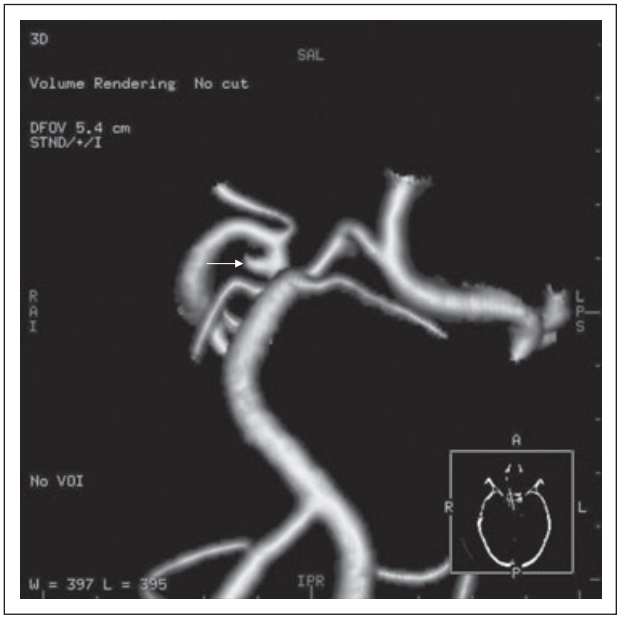

Figure.—Angio CT showing the infundibular dilatation of a perforating branch of the right posterior cerebral artery (arrow).

Recovery from the ophthalmoplegia was not as evident as was recovery from pain during the first trial with steroids but we had the impression in the subsequent course of prednisone that there was a global benefit. This second time the treatment was also started earlier (during the first day of pain).

We also instituted migraine prophylaxis with flunarizine and the number of headache (and ophthalmoplegic) episodes decreased.

Our patient had at first one normal, non-enhanced, interictal MRI.

We repeated the MRI twice, respectively, during the 5th and during the 2nd day of pain in 2 different episodes of OM.

The patient was not under prednisone treatment at the time of the second and third MRI studies. In both episodes, the patient had a right-sided IIIrd nerve palsy. No contrast enhancement was seen in the IIIrd nerve; we have found, however, an infundibular dilatation of a perforating branch of the posterior cerebral artery emerging just above the superior cerebellar artery, adjacent to the affected IIIrd nerve. We subsequently performed CT angiography to delineate the artery lumen and studied with T2 weighted imaging the cisternal space adjacent to the IIIrd nerve to confirm that the dilated vessel was in contact with the oculomotor nerve at its exit from the midbrain (Fig.).

Ophthalmoplegic migraine has been considered a recurrent demyelinating neuropathy and not a variant of migraine based on the finding on MRI with gadolinium of an enlarged and enhancing IIIrd nerve, ipsilateral to the symptoms and signs.[1-11]

Although the reported cases of OM with IIIrd nerve enhancement provide a frame for understanding its pathophysiology, the question of what causes this inflammation remains unanswered. Bharucha et al[9] reviewed the reported 52 cases of OM. Five cases had a normal MRI. In 44 patients reported in this study, MRI was done initially during one episode of OM and was repeated 1-8 months later. The IIIrd nerve enhancement was still present in 36.

Our patient's second and third MRIs were obtained with a 2-month interval and did not show the enhancing nerve lesion. The patient was in both instances symptomatic with headache and a IIIrd nerve palsy.

We found, however, an arterial infundibular dilatation in contact with the IIIrd nerve and confirmed this by CT angiography, which has the advantage of representing the true intraluminal arterial morphology.

Infundibular dilatation has been reported most frequently at the origin of the posterior communicating artery from the internal carotid. Its pathologic significance is uncertain. Some believe that there is at this point some defect in the arterial wall structure that may lead later to aneurysm formation.[12]

We speculate that the contact between the IIIrd nerve and the dilated intracranial artery could episodically produce a migrainous syndrome sustained by activation of the trigeminovascular system as well as compression and paralysis of the IIIrd nerve.

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Hemicrania Continua Evolving From Cluster Headache Responsive to Valproic Acid

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Hemicrania continua (HC) is a rare type of primary headache characterized by a prompt and enduring response to indomethacin. We describe a patient who suffered from cluster headache evolving into ipsilateral HC, who does not tolerate a long-term indomethacin therapy. The case was complex in terms of diagnosis, associated comorbidity, and choice of treatment; after several trials with different therapeutic regimens, we started the patient on a therapy with valproic acid and obtained an improvement of her HC.

Key words: hemicrania continua, cluster headache, valproic acid, indomethacin

INTRODUCTION

Hemicrania continua (HC) is a rare type of primary headache characterized by a complete response to therapeutic doses of indomethacin. It was first described in 1984 by Sjaastad and Spiering.[1] Clinically, it is characterized by moderate pain, continuous and unilateral without side-shift, but with severe exacerbations accompanied by ipsilateral autonomic phenomena, such as miosis, ptosis, lacrimation, and nasal congestion.[2]

Pathogenetically, an overlapping of the circuits involved in HC and cluster headache (CH) could be hypothesized. Using positron emission tomography, Matharu et al[3] demonstrated that there was a significant activation of the posterior hypothalamus contralaterally to pain, an activation of the rostral dorsal pons ipsilaterally, as well as an activation of some subcortical formations. Centonze et al[4] describe a case of CH evolving into HC, in which the pain side remained unchanged.

We report here what, to our knowledge, is the first case ever observed of CH evolving into HC responsive to valproic acid (VPA).

CASE REPORT

In December 2005, a 51-year-old woman, without previous history of headache, sought treatment at the Parma Headache Centre for a headache that had begun in June 2005. The headache was characterized by unbearable, excruciating pain in the right temporal-orbital region radiating ipsilaterally into the nasal wing, ramus of the mandible, and

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