Rare disease

Adult-onset migraine-related ophthalmoplegia and omolateral fetal-type posterior cerebral artery

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Summary

A 33-year-old woman with a long history of typical migraine without aura developed a pupillary-involving right third nerve palsy, after a typical migraine attack. The right pupil was 5 mm and showed delayed direct and consensual photomotor responses; the left pupil was 3 mm and reactive. Pupillary reaction to convergence was slow on the right eye. Ptosis, impaired elevation of the eye and weakened adduction were noted in the right eye. CT scan of the brain showed no abnormalities, whereas a CT digital cerebral angiography revealed a fetal-type right posterior cerebral artery (PCA). MRI disclosed thickening and contrast-enhancement of the cisternal portion of the right oculomotor nerve. A lumbar puncture, performed 5 days after the onset of ocular symptoms, yielded acellular cerebrospinal fluid (CSF) with normal protein and glucose levels. Ptosis and diplopia recovered within a week, whereas blurred vision, anisocoria and accommodation deficit subsided after 10 weeks.

BACKGROUND

Ophthalmoplegic migraine (OM) is a rare childhood-onset condition characterised by recurrent attacks of oculomotor nerve paresis accompanying or following headache with migrainous characteristics. Based on MRI findings of focal swelling and gadolinium enhancement of the cisternal portion of the affected oculomotor nerve, OM is currently considered an inflammatory demyelinating condition, either idiopathic or post viral and is classified under cranial neuralgias.^{1–3} There are reports of adult-onset, single ophthalmoplegic attacks in subjects with an antecedent history of migraine and these forms are difficult to classify.⁴

CASE PRESENTATION

A 33-year-old Caucasian woman was admitted to the hospital because of blurred vision in the right eye, ptosis of the right upper eyelid and vertical diplopia. Two days before the onset of ocular disturbances, the patient presented a right migraine attack with superimposed orbital pain, which subsided over the following hours after treatment with paracetamol 1000 mg. Medical history was unremarkable, with the exception of the diagnosis of migraine without aura (MoA), in accordance with criteria of the international classification of headache disorders (ICHD-II).¹ The patient had a positive family history of migraine and presented monthly headache attacks since the age of 16 years. On examination, the right pupil was 5 mm and showed a delayed response of direct and consensual photomotor reflexes; the left pupil was 3 mm and reactive. Pupillary reaction to convergence was slow on the right eye and induced an increase in anisocoria difference. Ptosis, impaired full elevation of the eye and weakened adduction were noted in the right eye in the absence of frank ocular deviation. Fundus oculi, visual fields and the remainder of the neurologic examination were unremarkable. Routine blood chemical values and complete blood count were not contributory.

INVESTIGATIONS

A CT scan of the cranium revealed no abnormalities, whereas a CT digital cerebral angiography revealed a fetal-type right posterior cerebral artery (PCA). Brain MRI showed thickening and postcontrast enhancement of the cisternal portion of the right third cranial nerve, whereas MR angiography confirmed the circle of Willis vascular anomaly and excluded neurovascular conflicts (figure 1). A lumbar puncture performed 5 days after the onset of ocular symptoms yielded acellular cerebrospinal fluid (CSF) with normal protein content.

DIFFERENTIAL DIAGNOSIS

Intracranial circulation disorders (ie, posterior communicating artery aneurism), infectious disease (ie, neurosyphilis, neuroborreliosis, tuberculosis, HIV), diabetes, inflammatory disease (ie, Miller-Fischer syndrome, chronic inflammatory demyelinating polyneuropathy (CIDP)), tumors, Tolosa-Hunt syndrome.

OUTCOME AND FOLLOW-UP

The clinical course was thereafter characterised by spontaneous and progressive improvement with full recovery of ptosis and vertical diplopia within a week, but only slight amelioration of blurred vision, anisocoria and accommodation deficit. The foregoing disturbances subsided 10 weeks after the onset. Repeat brain MRIs showed persistent thickening and reduced postcontrast enhancement of the oculomotor nerve at 3 months from the onset, whereas only a slight thickening was observed after 6 months (figure 1).

DISCUSSION

The patient here described had a 17-year history of MoA fulfilling ICHD-II criteria and, after an otherwise habitual migraine attack, developed a pupillary-involving

Figure 1 Axial T1-weighted MRI images enhanced by gadolinium at the level of the pituitary stalk: at presentation (a), the cisternal portion of the right third cranial nerve is thickened and enhanced by gadolinium; (b) slight thickening but no enhancement of the oculomotor nerve was found 6 months later; (c) MRI-angiography shows that the right posterior cerebral artery arises from the internal carotid artery.

oculomotor paresis associated with nerve thickening and Gd-enhancement at MRI, changes typically observed in OM.² The current line of thinking holds that oculomotor nerve alterations in OM reflect an underlying inflammatory demyelinating illness, that is, a limited form of CIDP or Guillain-Barré syndrome (GBS), and that migrainous pain is secondary to irritation of trigeminal fibers accompanying the oculomotor nerve.³ However, such an explanation remains unverified and is in contrast with the universal detection of normal CSF protein in investigated cases. Moreover, in CIDP and GBS with oculomotor palsy the cisternal portion of the oculomotor nerve is spared and MRI alterations are confined to the intracavernous segment of the nerve. Adult-onset migraine-related ophthalmoplegia is usually characterised by a typical history of migraine and single ophthalmoplegic attacks,⁴ and as such, it cannot be simply ascribed to cranial CIDP or GBS, solely based on MRI findings of thickening and Gd-enhancement of the first portion of the oculomotor nerve.² Alternative pathogenic hypotheses claim a major role of the trigeminovascular system in inducing chemical inflammation with breakdown of the blood-nerve barrier. The oculomotor root exit zone is particularly vulnerable since is supplied by many perforating branches arising from the proximal PCA, a vascular region with a high

trigeminovascular receptor density. Hence, activation of the trigeminovascular system may be ensued by focal vasogenic edema and nerve dysfunction, in keeping with MRI and clinical findings. In the case index, the presence of an ipsilateral fetal-type PCA may further explain the occurrence of migraine and ophthalmoplegia, since this vascular abnormality may contribute to migraine development and altered vasoreactivity.⁵ Taken together, this report further suggests that adult-onset migraine with ophthalmoplegia is not a demyelinating disorder.

Learning points

- ▶ Consider the adult variant of OM as cause of oculomotor palsy.
- ▶ Consider Tolosa-Hunt syndrome, circle of Willis disorders and anomalies, diabetic. ophthalmoparesis, infectious disorders, polyneuritis cranialis and lymphomas in the differential diagnosis of OM.
- ▶ Brain MRI studies are important in documenting reversible enhancement of the third nerve and ruling out parasellar, cavernous sinuses or orbital fissures pathologies.

Competing interests None.

Patient consent Obtained.

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