Middle Cerebral Artery Occlusion with Migraine

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SUMMARY A seven-year-old boy with migraine and a family history of hemiplegic migraine is described who, during an exacerbation, developed dysphasia and right hemiparesis. A CT scan showed a hypodense left cerebral lesion. Angiography revealed occlusion of the left middle cerebral artery at its origin. This represents the youngest case of stroke with migraine and, to our knowledge, is the first case report of angiogram-documented middle cerebral occlusion associated with migraine.

The headaches would usually be relieved by sleep but occasionally remitted spontaneously. One week prior to admission, the patient's headaches became more severe, prolonged and unresponsive to sleep or analgesics.

Four days prior to admission, he became increasingly reticent and lethargic and intermittently appeared confused and disoriented. For two days prior to admission, he had some right facial drooping, slurring of speech and dragging of the right foot while walking, worse at the end of the day. He was seen by a pediatrician and transferred to the Dartmouth-Hitchcock Neurology Service.

The child was the product of a normal pregnancy and delivery, had normal developmental milestones and was doing well in school. There was no history of severe illness, fever, head trauma, syncope, seizures or congenital heart disease.

The family history was remarkable for migraine headache. The child's mother complained of monthly episodes of common migraine. His father suffered severe and frequent complicated migraine, usually associated with profound photophobia, nausea, vomiting, pallor and often numbness and paresthias of the right side of the face and body. The maternal uncle was also reported to have frequent throbbing headaches often associated with numbness of the left arm and, at times, complete right arm paralysis. The child's maternal grandmother had a long history of complicated migraine associated with right-sided hemihypesthesia, hemiplegia and aphasia.

Physical examination showed no abnormalities. The patient was afebrile. He was in the 40th percentile for height and 50th percentile for weight. There was no rash or abnormal pigmentation. He was normocephalic and there was no evidence of bruits, trauma, sinusitis, pharyngitis or otitis. The fundoscopic exam was normal. The neck was supple and normal to palpation. The cardiopulmonary exam was normal.

On neurologic exam, the child responded promptly to questioning but spoke with a hesitant telegraphic speech in 3-4 word sentences. Cranial nerve exam showed full peripheral fields to confrontation and normal extraocular and pupillary function. There was slight right lower facial weakness. The motor exam showed mild right hemiparesis, arm weaker than leg, with a marked pronator drift of the right arm, drooping

TRANSIENT NEUROLOGICAL DEFICITS sometimes accompany migraine headache. Hemiplegia, hemihypesthesia, aphasia, ophthalmoplegia, and visual field deficits during migraine may mimic transient ischemic attacks and are manifestations of intracerebral vasospasm which, when prolonged, can result in seizure, infarction, or death. CT scans of the brain have occasionally corroborated clinical evidence for focal infarction and edema occurring during migraine attacks. Regional cerebral blood flow measurements have demonstrated focally-impaired hemispheric perfusion during migraine. Nevertheless, despite convincing evidence for neurovascular instability, permanent neurological deficits are rarely suffered by migraineurs. In general, CT scans of the brain and cerebrovascular angiography performed in cases of complicated migraine show no abnormalities. While vasospasm has been visualized occasionally in migraine patients, complete occlusion of intracerebral vasculature has been demonstrated in only a small number of cases.

In the following case, we report clinical and radiological evidence for complete occlusion of the left middle cerebral artery in a child who suffered from recurrent severe migraine headaches. This case corroborates evidence for vasospasm-induced intracerebral thrombosis and infarction which may occur in migraine. It underscores the importance as well as the difficulty of early diagnosis of this process in very young patients.

Case Report

The patient is a seven-year-old right-handed boy admitted to the Dartmouth-Hitchcock Medical Center for evaluation of mental status changes, speech difficulty and right hand clumsiness.

The child was described as having been colicky from birth with frequent episodes of prolonged irritability, abdominal cramps and daily vomiting up to the age of three to four years. During the year prior to admission, he began suffering from two to three episodes per week of severe headache, usually lasting one to two hours and often associated with profound pallor, anorexia and lethargy. Typically, they began behind the left eye and progressed to a bifrontal distribution. He was unable to describe the character of his pain.

The child was seen by a pediatrician and transferred to the Dartmouth-Hitchcock Neurology Service. For two days prior to admission, he had some right facial drooping, slurring of speech and dragging of the right foot while walking, worse at the end of the day. He was seen by a pediatrician and transferred to the Dartmouth-Hitchcock Neurology Service.

Four days prior to admission, he became increasingly reticent and lethargic and intermittently appeared confused and disoriented. Four days prior to admission, he became increasingly clumsy with his right hand and appeared unsteady on his feet. On the day of admission, he had some right facial drooping, slurring of speech and dragging of the right foot while walking, worse at the end of the day. He was seen by a pediatrician and transferred to the Dartmouth-Hitchcock Neurology Service.

The child was the product of a normal pregnancy and delivery, had normal developmental milestones and was doing well in school. There was no history of severe illness, fever, head trauma, syncope, seizures or congenital heart disease.

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Physical examination showed no abnormalities. The patient was afebrile. He was in the 40th percentile for height and 50th percentile for weight. There was no rash or abnormal pigmentation. He was normocephalic and there was no evidence of bruits, trauma, sinusitis, pharyngitis or otitis. The fundoscopic exam was normal. The neck was supple and normal to palpation without adenopathy. Carotid pulsations were strong and there was no cervical bruits. The cardiopulmonary, abdominal and musculoskeletal exams were normal.

On neurologic exam, the child responded promptly to questioning but spoke with a hesitant telegraphic speech in 3-4 word sentences. Cranial nerve exam showed full peripheral fields to confrontation and normal extraocular and pupillary function. There was slight right lower facial weakness. The motor exam showed mild right hemiparesis, arm weaker than leg, with a marked pronator drift of the right arm, drooping

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of the right shoulder and slight dragging of the right foot on rapid walking. There was a right hemisensory deficit, right-sided hyperreflexia with a right Babinski response.

The laboratory revealed normal electrolytes, arterial blood gases, urinalysis, glucose, PT/PTT, BUN, creatinine, and complete blood count with differential. The ESR was 17 with a negative ANA, LE prep and nonreactive RPR. The lumbar puncture showed an opening pressure of 170 mm Hg with a normal spinal fluid exam. Urine nitroprusside reaction and cystine screen was negative. Serum cholesterol and triglyceride levels were 188 and 189 respectively. The EKG, chest x-ray and echocardiogram were normal.

A CT scan of the head was performed with and without contrast enhancement. An area of decreased density was identified in the left frontoparietal region with slight compression of the left lateral ventricles compatible with ischemia or infarction of the left cerebral hemisphere (fig. 1). EEG showed a 2–4 Hz slow wave focus over the left hemisphere. On the third hospital day, cerebral angiography was performed which showed a complete occlusion of the left middle cerebral artery at its origin (fig. 2). Collateral flow from the anterior cerebral was seen to fill most of the branches of the middle cerebral artery except for the posterior division. There was no suggestion of arteritis.

The child suffered severe headache, nausea and vomiting despite gradual neurologic improvement for three days following the arteriogram. On the sixth hospital day, a repeat CT scan showed hypodensity of the left fronto-parietal region without evidence for edema or mass effect. The patient was begun on propranolol 1
mg/kg tid for headache prophylaxis. After discharge, the child was followed closely as an out-patient and improved steadily. Six months after hospitalization he is free from recurrent headache and demonstrates only a barely detectable right-sided hemiparesis and no detectable physical or mental handicaps.

A follow-up flow study brain scan performed one year later showed normal flow in the middle cerebral artery distribution bilaterally.

Discussion

Migraine headache is frequently difficult to diagnose and commonly overlooked in the pediatric population.

It is well established, however, that migraine may start at a very young age and may present with very different symptoms from the usual adult forms.

Migraine in children has been defined in varying ways. The criteria for diagnosis are, as defined by Vahlquist and revised by Prensky, that headache must be recurrent, separated by symptom-free intervals, and accompanied by at least three of the following six symptoms: abdominal pain, nausea and vomiting; localized unilateral headache; a throbbing pulse-like quality to the pain; complete relief after a period of sleep; an aura which may be visual, sensory or motor; and a family history of migraine. However, because young children may be unable to describe the character and location of their pain, the diagnosis of migraine may be especially difficult. Motion sickness, cyclic vomiting, recurrent abdominal pain, acute confusional states and autonomic symptoms of pallor and sweating are accepted migraine equivalents in children.

Hemiplegic migraine, an uncommon variant of complicated migraine, frequently begins in the pediatric age group and is often familial. Such neurologic symptoms are uniformly transient however, and with rare exception resolve over minutes to days without permanent neurological sequelae.

Patients who suffer migraine may have both a neurovascular instability and a hypercoagulable state which predisposes them to stroke. Migraine-associated cerebral infarction, however, is an uncommon and poorly understood phenomenon. Prolonged vasospasm of the intracranial arteries during migraine is an accepted underlying mechanism causing irreversible neurologic deficits and death. The family history as well as the clinical presentation indicate that this patient was suffering from migraine. It is probable that he had middle cerebral artery vasospasm, thrombosis and infarction as a result of this disorder.

Branch occlusion of intracranial vessels has rarely been documented with migraine. Complete obstruction of the posterior cerebral artery has been demonstrated angiographically in only three case reports of stroke with migraine and occlusion of the anterior cerebral artery documented only once. In Fisher's study of 120 patients with late-life migraine accompaniments and, in Pearce's report of 40 patients with complicated migraine, cerebral angiography was unre-markable in every case where it was performed. Similarly, in Boisen's report of 7 cases and Connor's series of 15 strokes with migraine, angiography, when employed, was normal in every case but one. While migraine-associated fronto-temporal infarction has been documented on CT, to our knowledge occlusion of the middle cerebral artery with this disorder has never before been demonstrated.

Stroke in children is not uncommon. Approximately 5% of patients with stroke admitted to hospitals are below age 20. These children often have predisposing factors such as cyanotic heart disease, homocystinuria, severe pharyngitis or head and neck trauma. Twenty to thirty percent who become hemiplegic have no predisposing factors, a completely negative metabolic workup and a normal cerebral arteriogram. Basal vasculature occlusion without associated telangiectasia is a common finding in idiopathic acute childhood hemiplegia and among these, middle cerebral occlusion is present 40% of the time. The incidence of migraine and the contribution its associated neurovascular instability and hypercoagulable state may play in this population has not been adequately addressed. Migraine-associated cerebral infarction may be a more common entity than is currently recognized, particularly in the pediatric population. It must enter the differential in all cases of acute childhood hemiplegia.

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