Sporadic Hemiplegic Migraine: A Challenging Diagnosis

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Migraine affects about 12% of the population in Western countries. Familial hemiplegic migraine (FHM) is the only variety of migraine characterized by an autosomal dominant pattern of inheritance and transient hemiparesis followed by migraine headache. Some sporadic cases of hemiplegic migraine with cerebellar signs have also been reported. Sporadic hemiplegic migraine (SHM) has clinical symptoms identical to FHM and distinct from migraine with aura.

CASE REPORT

A 20-year-old right-handed gentleman presented with a 2-day history of left-sided weakness (evolving to maximum deficit over half an hour), followed by severe unilateral headache, vomiting and altered level of consciousness. Weakness was preceded by numbness of the left half of the body, and blurring of vision. There was no history of fever, neckache, sphincter dysfunction, abnormal movement, or head trauma. Over the last 8 years, the patient had experienced several similar episodes of less severe intensity, none lasting more than 4 hours. Frequency was variable ranging from 2 per month to once every 3-4 months. A brain CT scan and cerebrospinal fluid examination done in 1993 were normal. Subsequent brain CTs repeated CT at variable intervals were also normal. Past medical, surgical and family history were unremarkable. He was unmarried, a non-smoker and did not consume alcohol. He had quit schooling after the 10th grade because of his illness. Medication use included multiple analgesics, including intravenous injections for headache.

On examination at the time of admission, he was opening eyes on painful stimuli, localizing with the right hand, and had no verbal response. Power was 0/5 in the left half of the body with extensor plantar response on the left. He had a left facial nerve palsy of upper motor neuron type. He was moving the right side spontaneously, and there were no signs of meningeal irritation. Fundi were normal. Sensory, cerebellar, and gait examination were deferred. Routine blood tests including complete blood count, urea,
Creatinine, electrolytes, and random sugar were within normal limits. Anti-epileptic drug levels were in the subtherapeutic range. Magnetic resonance imaging of the brain (including diffusion-weighted studies) was normal.

EEG showed severe right hemispheric dysfunction. The patient's family did not consent to CSF analysis. There was no significant clinical improvement after 3 days of hospitalization. An initial diagnosis of epilepsy (made in the past at another facility) was questioned. On the basis of a detailed history, exclusion of metabolic abnormalities, and normal neuroimaging, a diagnosis of sporadic hemiplegic migraine was tentatively made. He was started on NSAIDs, flunarizine and valproic acid. Strength on the affected side gradually improved to 3/5, he became fully conscious and oriented, and he was discharged on above medications. After 3 days, he was again hospitalized with severe headache, vomiting, imbalance, and drowsiness. He was given steroids for status migrainosus and intravenous verapamil (5 mg over 5 minutes), followed by oral verapamil (120 mg twice a day). Within 48 hours his power returned to normal and the headache subsided completely. Repeat EEG also showed improvement. A final diagnosis of SHM was reached.

**DISCUSSION**

Migraine is characterized by episodes of headache that are often throbbing, frequently unilateral, and may be severe. In migraine without aura (previously known as common migraine), attacks are usually associated with nausea, vomiting, and sensitivity to light, sound or movement. A combination of features is required for the...
diagnosis, though not all features are present in every attack or every patient. Any severe and recurrent headache is most likely to be a form of migraine and to be responsive to anti-migranous therapy. In 15% of patients, migraine attacks are usually preceded or accompanied by transient focal neurological symptoms; such patients have migraine with aura (previously known as classic migraine).  

Hemiplegic migraine was initially described in 1910 as a type of migraine consisting of recurrent headache associated with transient hemiparesis. Familial hemiplegic migraine (FHM) is a rare autosomal dominant form of migraine with aura in which some degree of hemiparesis is present during attacks. In FHM, the aura typically lasts longer than in migraine with aura and usually comprises visual, sensory, aphasic and motor symptoms. However, patients with similar clinical symptoms but without other affected family members have been reported, and the form sporadic hemiplegic migraine has been used. Based on the literature, two subforms of FHM families exist - pure FHM in 80% and FHM families with cerebellar symptoms in 20%. Mutations in the gene CACNA1A, which encodes a neuronal calcium channel, are present in 50% families with hemiplegic migraine including those with cerebellar signs. Eight mutations in CACNA1A have been identified in 18 families affected by hemiplegic migraine and two patients with SHM. Genetic studies have revealed de novo mutations or incomplete penetrance of CACNA1A in patients with SHM, thereby suggesting that SHM may be part of the spectrum of FHM syndromes.

FHM is a disorder with an interesting and complex symptomatology. Motor aura symptoms are obligatory. Furthermore, 69% have concurrence of basilar migraine, with basilar-type symptoms during attacks. During episodes, 95% patients always experience a headache, 4% experience headache in some attacks, and 1% never experience headache. Other associated symptoms include cerebellar ataxia associated with progressive cerebellar atrophy and epilepsy. FHM has a younger age of onset compared with migraine with aura.  

SHM has clinical symptoms identical to FHM. In SHM, presence of motor aura symptoms is essential. Other common aura symptoms are sensory (98%), visual (91%), and some form of aphasia (81%). Up to 72% of patients with SHM have concurrence of basilar migraine. Headache almost always occurs in close temporal relation to the aura, and SHM has an age aof onset younger than 45 years. The most differential diagnoses of SHM typically includes epilepsy (post-ictal weakness following seizure, or Todd's phenomenon), transient ischemic attack or stroke, metabolic abnormalities associated with focal deficits (hypercapnia, hypoglycemia, hyponatremia, hypocalcemia, hepatic failure and renal failure), meningitis or encephalitis, carotid dissection, anti-phospholipid antibody syndrome, SLE, and ornithine transcarbamylase deficiency.

Inherited disorders associated with migraine headache that may include hemiparesis include cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL); mitochondrial myopathy, encephalopathy, lactic acidosis and stroke like episodes (MELAS); hereditary hemorrhagic telangiectesia (HHT); hereditary cerebral amyloid angiopathy; familial cerebral cavernous malformation; and benign familial infantile convulsions.  

Current therapeutic recommendations are based on isolated case reports and include propranolol, phenytoin, flunarizine, and verapamil. Successful responses of FHM to intravenous verapamil have been reported. Verapamil, administered either orally or intravenously, is also effective for SHM. Our patient had no further episodes after starting oral verapamil 240 mg daily, and was symptom-free at one-year follow up. SHM is a relatively rare disorder which should be considered in patients presenting with episodes of reversible neurological deficits with headaches; a therapeutic trial of calcium channel blocker (verapamil) may be administered.

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