Hemiplegic Migraines; A Barrage on the Mind

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Hemiplegic Migraines; A Barrage on the Mind

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Abstract
Hemiplegic migraines (HM) are a rare type of migraine with an aura phase that includes muscle weakness. This form of migraine can either occur sporadically or can be seen in patients with familial hemiplegic migraine which is an autosomal dominant subtype that runs in families. Migraines are classically accompanied by phonophobia, photophobia, neurological symptoms, nausea or vomiting. The subcategory of hemiplegic migraine involves a transient wave of motor weakness and muscle spasms secondary to decreased threshold of excitation in the neurologic system. The etiology of Hemiplegic Migraines is either idiopathic condition or genetic mutation in the CACNA1A, ATP1A2 and SCN1A encoding proteins involved in ion transport. Our case outlines a 72 year old Caucasian female with past medical history of transient ischemic attack (TIA) and paroxysmal atrial fibrillation, presenting to the emergency department with sudden onset of headache, slurred speech and numbness and tingling in the left hand and leg. Tissue plasminogen activor t-PA) was administered within the 45 minute window, and a computed tomography angiography(CTA) of the head and neck was negative. Diagnosis of Hemiplegic Migraine was suggestive given the neurologic signs and symptoms with negative neurologic imaging. The expedited movement of neuronal depolarization down the easily excitable neurons causes a high amplitude wave of energy down the muscle fibers. Other hemiparesis syndrome differentials include MELAS, hereditary hemorrhagic telangiectasia, amyloidosis, and familial infantile convulsions. This case highlights the rarity of hemiplegic migraines and although genetic testing was not performed in our patient, it has been shown that some patients have a genetic predisposition. Strong clinical skills are needed to match the concurrent neurologic symptoms with migraines.

Keywords
Hemiplegic Migraines, Headache

Conflict of Interest Statement
The authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest

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Abstract

Hemiplegic migraine (HM) are a rare type of migraine with an aura phase that includes muscle weakness. This form of migraine can either occur sporadically or be seen in patients with familial hemiplegic migraine which is an autosomal dominant subtype that runs in families. Migraines are classically accompanied by phonophobia, photophobia, neurological symptoms, nausea or vomiting. The subcategory of hemiplegic migraine involves a transient wave of motor weakness and muscle spasms secondary to decreased threshold of excitation in the neurologic system. The etiology of Hemiplegic Migraines is either idiopathic condition or genetic mutation in the CACNA1A, ATP1A2 and SCN1A encoding proteins involved in ion transport. Our case outlines a 72 year old Caucasian female with past medical history of transient ischemic attack (TIA) and paroxysmal atrial fibrillation, presenting to the emergency department with sudden onset of headache, slurred speech and numbness and tingling in the left hand and leg. Tissue plasminogen activator t-PA) was administered within the 45 min window, and a computed tomography angiography (CTA) of the head and neck was negative. Diagnosis of Hemiplegic Migraine was suggestive given the neurologic signs and symptoms with negative neurologic imaging. The expedited movement of neuronal depolarization down the easily excitable neurons causes a high amplitude wave of energy down the muscle fibers. Other hemiparesis syndrome differentials include MELAS, hereditary hemorrhagic telangiectasia, amyloidosis, and familial infantile convulsions. This case highlights the rarity of hemiplegic migraines and although genetic testing was not performed in our patient, it has been shown that some patients have a genetic predisposition. Strong clinical skills are needed to match the concurrent neurologic symptoms with migraines.

Keywords: Hemiplegic migraines, Headache

1. Introduction

Hemiplegic migraines (HM) are a rare type of migraine with an aura phase that includes muscle weakness. This form of migraine can either occur sporadically or can be seen in patients with familial hemiplegic migraine, a familial autosomal dominant subtype. While migraines are common disorders affecting approximately 15%–20% of the population, hemiplegic migraines are rare occurrences with a prevalence of 0.01%. The motor auras seen in these migraines are thought to be caused by a self-propagating wave of neuronal and glial depolarization resulting in cortical depression. Studies have shown that these attacks are more common in women with an average age of onset of 12 and 17 years old.

2. Case presentation

A 72 year old Caucasian female with past medical history (PMH) of multiple episodes of TIA, paroxysmal atrial fibrillation, orthostatic hypotension, and hypertension presented to the ED with sudden onset of a headache without radiation or laterality, numbness and tingling in the fingers of her left hand, weakness in the left leg, left sided facial droop, and slurred speech. On admission vitals were: Temperature 97.3 F, HR 55 beats per minute, RR: 12 per minute, BP: 151/54, Oxygen Saturation 100% on room air. Her National Institute of Health (NIH) stroke scale score was 8.

On physical examination, muscle strength was 4+ in the left upper and lower extremities. Left facial droop was confirmed. Cranial nerve VII deficits were
noted and the patient could not smile, frown, or puff out her cheeks. Forehead was spared and facial sensation was intact. The Biceps, brachioradialis, triceps, patellar, and Achilles reflexes were tested and were globally 1+ on the left side and 2+ on the right side. CT head showed no acute intracranial abnormalities. CTA head revealed no sign of dural sinus thrombosis or significant intracranial venous or arterial abnormalities. CTA neck showed minimal left greater than right atherosclerotic plaque formation in the bilateral carotid bulbs, without hemodynamically significant stenosis. Posterior venous abnormalities, while possible, were considered unlikely given her symptoms. As the patient arrived within the tPA window, and no obvious bleed was seen on the CTA head, alteplase was administered in the Emergency Department. However, this intervention resulted in only minimal improvement.

Patient stated that she was admitted for a similar event in 2017, for which she also received IV tPA. That event was not associated with headaches. Echocardiogram performed then and at the time of this admission showed normal cavity size, atrial septa, wall thickness, diastolic parameters, and left ventricular ejection fraction with no regional wall motion abnormalities (Table 1).

The patient was admitted for cerebral infarction due to unspecified occlusion or stenosis of the basilar arteries, and also diagnosed with hemiplegic migraine given neurologic signs and symptoms with negative neuro imaging findings. Following admission she was started on Topiramate 25 mg po BID for one week and then increased to 50 mg po BID with resolution of migrainous symptoms.

### 3. Discussion

Migraines are classically accompanied by phonophobia, photophobia, neurological symptoms, nausea or vomiting. The subcategory of hemiplegic migraine (HM) involves a transient wave of motor weakness and muscle spasms secondary to decreased threshold of excitation in the neurologic system. Hemiplegic migraine is a rare and complex disease, characterized by migraine with a reversible motor aura. Motor symptoms usually last for hours to days with a spectrum of visual, sensory, or possibly speech impairments. The etiology of this disorder is either idiopathic or genetic mutation in the CACNA1A, ATP1A2 and SCN1A encoding proteins involved in ion transport. The expedited movement of neuronal depolarization down the easily excitable neurons causes a high amplitude wave of energy down the muscle fibers. In the genetic form, exacerbations start in the first to second decade of life.

Differentiation of classical migraine from HM is the type of aura. A migraine aura usually presents with visual symptoms, however motor symptoms are uncommon. In HM, the aura is characterized by the presence of motor weakness, often accompanied by impairments in vision, speech, and sensation. Our patient presented with recurrent headaches associated with aphasia, numbness, and weakness, thus consistent with the presentation of HM auras.

Neuroimaging, cerebrospinal fluid analysis, and electroencephalography are useful but the diagnosis is clinical with the aid of genetic confirmation. If undiagnosed, HM may be mistaken for epilepsy due to its tendency to recur along with the fact that auras seen in HM present similar to postictal (Todd) paralysis. The aura in hemiplegic migraines can include visual symptoms which are almost never seen in epilepsy, allowing for some differentiation when present. The duration of symptoms seen in HM is approximately 60 min, much longer than is typically seen in epilepsy. Other hemiparesis syndrome differentials need to be excluded as well. These include mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes (MELAS Syndrome), hereditary hemorrhagic telangiectasia (HHT), amyloidosis, and familial infantile convulsions. MELAS syndrome usually presents in individuals between the ages of 2–15 years with definitive diagnosis obtained via genetic testing or muscle biopsy showing many ragged red fibers. HHT usually presents with signs of bleeding including nose bleeds, telangiectasias on the lips and oral mucosa, and

### Table 1. Labs at the time of this admission.

| CBC: | White Count: 8.5 (ref: 4–10 x 10³ uL) |
|      | Hemoglobin: 12.6 (ref:11.6–15.2 g/dL) |
|      | MCV: 90.9 (ref: 80–97 FL)            |
|      | Platelets: 254,000 (ref: 150,000–450,000) |
| CMP: | Sodium: 142 (ref: 136–145 mmol/L)    |
|      | Potassium: 3.8 (ref: 3.5–5.1 mmol/L)  |
|      | Chloride: 108 (ref: 98–107 mmol/L)   |
|      | Bicarbonate: 26 (ref: 21–31 mmol/L)  |
|      | BUN: 17 (ref: 7–25 mg/dL)            |
|      | Creatinine: 0.7 (ref: 0.6–1.2 mg/dL) |
| Lipid Profile: | Cholesterol: 141 (ref: <200 mg/dL) |
|      | Triglycerides: 203 (ref: 0–150 mg/dL) |
|      | HDL: 35 (ref: >39.9 mg/dL)           |
|      | LDL (calculated): 65 (ref: <100 mg/dL) |
| APTT: | 35.8 (ref: 22.9–36.5 s)              |
| PT:  | 11.8 (ref: 11.0–14.6 s)              |
| INR: | 0.9 (ref: 0.8–1.2)                   |
| HbA1c: | 5.8 (ref: 4.0–6.0%)                  |
hemorrhages in the gastrointestinal system, or brain. Amyloidosis can present as a headache, loss of consciousness and vomiting; long term complications of this disease also includes dementia. Lastly, familial infantile convulsions usually presents with convulsions between 3 and 8 months of life with clusters of brief seizures lasting only 3–5 min over a few days.

A complication of HM is permanent brain damage and atrophy as well as cognitive deterioration in a stepwise developmental pattern. Neuroimaging can reveal small white matter lesions or demyelination.

4. Conclusion

This case highlights the rarity of hemiplegic migraines and the genetic predisposition in these patients. Strong clinical skills are needed to match the concurrent neurologic symptoms with migraines. This patient received tPA for a presumed stroke—twice. One way to differentiate between hemiplegic migraines and stroke is to note the onset of symptoms. Rapid onset of symptoms suggests a stroke. However, if symptom onset is slower it is more likely to be a hemiplegic migraine. Deducing when the symptoms first started can also differentiate HM from a stroke. Hemiplegic migraines usually present in the first few decades of life whereas a stroke is more likely to occur in individuals over 50 years of age. Lifestyle habits should also be evaluated as poor dietary habits, low exercise, alcohol and smoking can increase the risk of stroke.

Conflict of interest

The authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest.

References