EM CASE OF THE WEEK

BROWARD HEALTH

Author: Srividya Kakulavarapu I **Editor:** Matthew Holme, MD

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Hemiplegic Migraine

A 49-year-old African American female with a past medical history of hypothyroidism, asthma, and hypertension presents to BHMC ED with a chief complaint of an occipital headache. She experiences these episodes intermittently. She states that previous episodes have been associated with a variety of symptoms including numbness of the cheek, chest, and upper extremity, chest tightness, photophobia, phonophobia, nausea, vomiting, dizziness, loss of balance, visual changes, shortness of breath, and aphasia. Patient is afebrile and vitals are currently within normal limits. Physical examination demonstrates diminished sensation to the left side of the face that extends down the left upper extremity to the left digits. There is also decreased strength in flexion and extension of the left arm. The remainder of her neurological exam is normal. Which of the following is the most appropriate initial diagnostic test for this patient's condition?

A. PA and Lateral Chest X-Ray

B. CT Head without Contrast

C. CT Angiography

D. Ultrasound of the Carotid Arteries

E. No diagnostic testing necessary

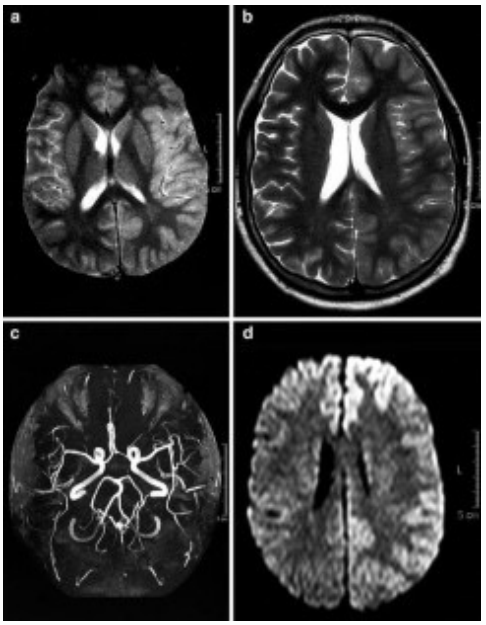


Figure 1: Butteriss DJ, Ramesh V, Birchall D. Serial MRI in a case of familial hemiplegic migraine. Neuroradiology. 2003;45(5):300-303. doi:10.1007/s00234-003-0979-z.

The above are MRI findings in a patient with familial hemiplegic migraine (FHM) with repeated episodes of hemiparesis.

FHM is caused by a penetrant autosomal dominant genetic mutation.

Department of Emergency Medicine
1600 S. Andrews Avenue
Fort Lauderdale, FL 33316

BROWARD HEALTH MEDICAL CENTER
Emergency Department

The correct answer to the question above is B. To rule out hemorrhagic intracranial process, the best initial choice is head CT scan without contrast.

The primary feature that separates hemiplegic migraine from other types of migraine with aura is the presence of motor weakness as a manifestation of aura in at least some attacks. Migraine auras most often manifest as visual disturbances but can also involve sensory, verbal, and rarely motor disturbances. Thus, hemiplegic migraine is a unique subtype of migraine with aura. Hemiplegic migraine may be familial or sporadic. Patients who are the first member of their family to have hemiplegic migraine are classified as having sporadic hemiplegic migraine. It is a rare disorder with an estimated prevalence of 0.01%. The familial and sporadic versions occur with equal prevalence and the average age of onset is 12-17 years (range 1-51 years). Like other forms of migraine, females have a higher prevalence with ratios ranging from 2.5:1 to 4.3:1.

Discussion

Hemiplegic migraine is characterized by migraine attacks that include motor weakness during the aura phase. Attacks may variously include severe headache, scintillating scotoma, visual field defect, numbness, paresthesia, unilateral weakness, aphasia, fever, lethargy, coma, and seizures. The symptoms can last for hours to days, or rarely, weeks, but must resolve completely. The clinical symptoms of sporadic hemiplegic migraine are indistinguishable from those of familial hemiplegic migraine. Chronic cerebellar ataxia and epilepsy can occur in patients with hemiplegic migraine. Studies have shown a significant association with epilepsy.

Evaluation of patients with headache and neurological symptoms where the diagnosis of hemiplegic migraine is considered includes the application of established diagnostic criteria and clinical investigations to exclude entities in the differential diagnosis.

A diagnosis of hemiplegic migraine is made if the following are satisfied:

1. The patient's symptoms fulfill the International Classification of Headache Disorders diagnostic criteria (see below).
2. Testing to exclude other considered causes within the differential diagnosis is necessary for patients in whom the diagnosis of hemiplegic migraine is not established.

A thorough family history is essential to distinguish familial from sporadic hemiplegic migraine.

International Classification of Headache Disorders 3

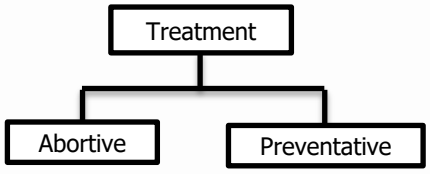
1.2.3 Hemiplegic migraine

A. Attacks fulfilling criteria for 1. Migraine with aura and criterion B below

B. Aura consisting of both of the following:
   1. fully reversible motor weakness
   2. fully reversible visual, sensory and/or speech/language symptoms

Treatment

The management of hemiplegic migraine involves pharmacologic treatment with agents typically used to abort or prevent migraine that do not potentiate the risk of cerebral vasoconstriction. Patients with severe neurologic impairment may require hospitalization and additional measures.



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Treatment (cont.)

First-line options
- Verapamil for patients with frequent or prolonged aura
- Flunarizine, topiramate, or amitriptyline for patients with pain predominant headaches
- Acetazolamide for patients with familial hemiplegic migraine

Alternative options
- Lamotrigine for patients with persistent aura symptoms
- Ketamine, naloxone, and onabotulinmtoxinA

Acute management of severe attack
- Parenteral methylprednisolone

In nearly all patients with hemiplegic migraine, the aura symptoms eventually resolve completely though they may be prolonged. In rare cases, hemiplegic migraine leads to permanent neurologic deficits, cerebral infarction, cognitive decline, or death. Most such patients have phenotypes characterized by early onset of hemiplegic migraine with severe attacks, recurrent coma, or seizures. In many patients, the frequency of attacks decreases after age 50, and hemiplegic attacks can evolve into more typical migraine attacks without hemiparesis.

Take Home Points

- **Hemiplegic migraine may be familial or sporadic.**
- **Hemiplegic migraine is characterized by complex migraine auras that include motor weakness and at least one other feature such as paresthesia, aphasia, or brainstem impairment. Unilateral or bilateral headache commonly accompanies the aura. Severe cases may also include fever, lethargy, coma, and seizures. The symptoms can last for hours to days, sometimes weeks. Most resolve completely.**
- **The diagnosis of hemiplegic migraine is made when symptoms fulfill the diagnostic criteria, including at least two episodes of migraine with reversible complex aura consisting of motor weakness and one or more additional feature (visual, sensory, aphasic, or brainstem). It is a diagnosis of exclusion.**
- **Treatment options are similar to other forms of migraine treatment and include verapamil, flunarizine, topiramate, amitriptyline, or acetazolamide.**

About the Author

This month's case was written by Srividya Kakulavarapu. Srividya was a 4th year medical student from Nova Southeastern University Dr. Kiran C. Patel College of Osteopathic Medicine. She did her emergency medicine rotation at BHMC in April 2022. Srividya is currently an intern in Internal Medicine at the University of Miami / JFK Medical Center in Palm Beach.

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UpToDate: Migraine – Pathophysiology, Epidemiology, Clinical Manifestation, Diagnosis, Management, Prognosis

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