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Case Report

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Neuroimaging in Hemiplegic Migraine - Case Report and Literature Review

Omar Islam[1*] and Zara Khan[2]

[1]Omar Islam, MD FRCPC, DABR. Associate Professor, Department of Diagnostic Radiology, Queen's University, Kingston, Canada

[2]Zara Khan, Faculty of Health Sciences, Queen's University, Kingston, Canada

***Corresponding author**
Omar Islam, MD FRCPC, DABR. Associate Professor, Department of Diagnostic Radiology, Queen's University, Kingston, Canada.

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Case Report

Presenting Illness

A 29-year-old man presented to the emergency department with sudden-onset aphasia and left-arm weakness. The patient's initial appearance was that of a healthy, well-developed male. He had not had any fevers, chills, cough, or nausea.

There was no history of prior medical conditions, prescription drugs, or history of smoking or illicit drug use. He had no remarkable social history, such as lifestyle behaviors, personal habits, or environmental factors that may influence his health, including occupation, social support, and living conditions. He worked in construction as a heavy-equipment operator.

Physical Examination and Workup

Vital signs were within normal range, including heart rate, respiratory rate, temperature, and blood pressure. Physical examination revealed a patient who was stable and in no obvious distress. It was determined that the patient was right-handed. He had a grade of 2/5 motor weakness in his left arm. His left leg had normal motor function. The patient had both receptive and expressive aphasia. The remainder of the physical examination was normal. Cranial nerve examination was normal.

Initial laboratory investigations were all normal, including CBC, electrolytes, and renal function. Chest radiography performed in the emergency department also was normal.

Given the stroke-like symptoms, a CT of the head with CT angiogram of the neck and head was performed, which was normal. See representative image from CT head (Figure 1).

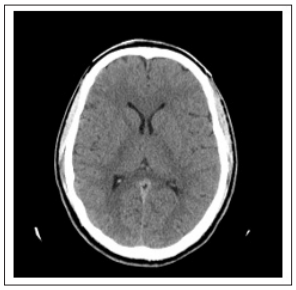

Figure 1: Axial Non-Contrast CT Head Performed on Patient at Initial Presentation to the Emergency Department

There were no signs of an acute cortical infarction or intracranial hemorrhage. The major vessels of the neck and circle of Willis were normal, with no carotid or vertebrobasilar stenosis or dissection, no large-vessel occlusive thrombus, and no CT angiography signs of vasculitis.

Given the clinical history and negative CT results, the patient underwent an urgent MRI of the brain. Diffusion-weighted imaging of the brain was normal. This excluded an acute infarction (Figure 2).

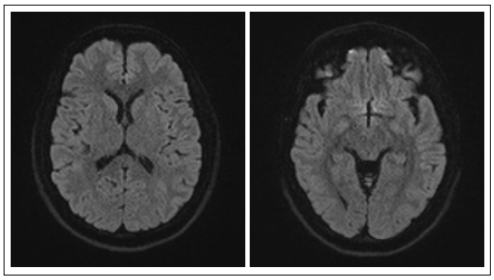

Figure 2: Axial Diffusion-Weighted MRI of the Patient's Brain

MRI findings were not normal. Susceptibility-weighted imaging revealed dark signal (susceptibility artifact) within multiple cortical veins in the left cerebral hemisphere (Figure 3).

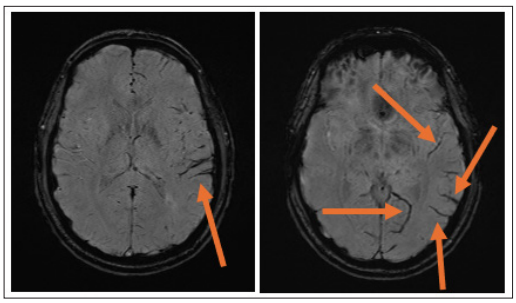

Figure 3: Axial Susceptibility-Weighted MRI of the Patient's Brain

There were no signs of subarachnoid hemorrhage, either on CT or MRI. The dark signal on susceptibility-weighted imaging was confined to the cortical veins, and not the cerebrospinal fluid within the subarachnoid spaces [1].

A diagnosis of hemiplegic migraine was made based on the characteristic imaging appearance.

Clinical Course and Prognosis

The patient in this case was managed conservatively, and his symptoms resolved within 24 hours without the need for aggressive intervention.

Discussion

Epidemiology

Hemiplegic migraine is a subtype of migraine with aura [2]. It is a reversible neurologic condition with no underlying vascular occlusion [2,3].

It has an estimated prevalence of approximately 0.01%. Hemiplegic migraine is more commonly observed in females than in males, with an approximate 3:1 ratio [2]. Hemiplegic migraine can be classified into familial and sporadic categories. Familial hemiplegic migraine follows an autosomal dominant inheritance pattern [4]. Sporadic hemiplegic migraine occurs without a family history. Genetic mutations in CACNA1A, ATP1A2, and SCN1A have been associated with familial cases.[3] Many sporadic cases have no identifiable genetic markers [4]. Owing to its rarity, hemiplegic migraine is often misdiagnosed as other neurologic conditions [4].

Typical and Atypical Clinical Presentations

Hemiplegic migraine presents with transient unilateral motor weakness or hemiparesis [5]. This is often accompanied by additional aura symptoms such as visual disturbances, sensory abnormalities, and speech impairment [2,5]. The weakness typically lasts from minutes to hours but may persist for several days in some cases. Visual disturbances, including scintillating scotomas and hemianopia, are common [5]. Sensory symptoms such as paresthesia and numbness frequently occur. Speech impairments such as dysphasia or aphasia may be present. This is particularly true if the dominant hemisphere is affected [6].

Atypical presentations of hemiplegic migraine may include persistent neurologic deficits, episodic confusion, prolonged aura lasting for weeks, and altered levels of consciousness. In severe cases, symptoms may mimic a stroke [5]. This can lead to unnecessary thrombolytic treatment or invasive diagnostic testing. Pediatric patients may present with symptoms suggestive of stroke or seizure disorder, complicating the initial diagnosis [2].

Useful and Required Diagnostic Studies

Neuroimaging is critical in evaluating patients with suspected hemiplegic migraine to rule out other serious neurologic conditions such as ischemic stroke, intracranial hemorrhage, or space-occupying lesions [7].

MRI is the preferred imaging modality, as diffusion-weighted imaging is typically normal, which helps to exclude acute ischemic stroke [7]. Fluid-attenuated inversion recovery (FLAIR) sequences may occasionally reveal transient cortical edema [8]. Susceptibility-weighted imaging often shows dark signal artifacts in cortical veins, indicative of transient vascular involvement, which is a characteristic finding in hemiplegic migraine [9]. This suggests transient venous congestion or altered hemodynamics, which may contribute to the pathophysiology of the condition [10]. The absence of infarction on diffusion-weighted imaging and the lack of parenchymal abnormalities on FLAIR and T2-weighted sequences further support the diagnosis of hemiplegic migraine rather than an acute ischemic event.

Electroencephalography may be considered in patients with seizure-like activity. Findings may reveal diffuse slowing over the affected hemisphere, but these findings are nonspecific [8].

CT is often performed acutely to exclude hemorrhage, but it lacks the sensitivity to detect the subtle changes characteristic of hemiplegic migraine [11].

Carotid Doppler ultrasonography would not be indicated in patients presenting with acute stroke-like symptoms, unless the patient has a contraindication for CT angiography, in which case magnetic resonance angiography would also be a viable alternative.

Genetic testing is recommended in cases in which familial hemiplegic migraine is suspected, as mutations in CACNA1A, ATP1A2, and SCN1A have been identified in affected individuals [4].

Management

Management of hemiplegic migraine includes both acute symptom relief and preventive strategies aimed at reducing attack frequency and severity. Acute treatment options include NSAIDs and acetaminophen as first-line options for pain relief [5]. Calcium channel blockers such as verapamil are sometimes used off-label for aborting attacks, while intravenous verapamil has shown efficacy in stopping acute attacks in select cases [12]. Ketamine, an N-methyl-D-aspartate receptor antagonist, has been reported to alleviate aura symptoms in certain cases, although its use remains off-label [4].

Preventive treatment strategies focus on reducing attack frequency and severity. Calcium channel blockers such as verapamil are commonly prescribed for this purpose [5]. Antiepileptic drugs, including lamotrigine, sodium valproate, and topiramate, may help prevent recurrent attacks in patients with frequent or severe symptoms, and calcitonin gene-related peptide (CGRP) targeting therapy has also recently been introduced for this purpose [13,14]. Beta-blockers such as propranolol remain controversial but may be beneficial in some individuals. Acetazolamide, a carbonic anhydrase inhibitor, has been effective in some patients for reducing the severity and frequency of attacks [4].

Thrombolytic therapy is not indicated because hemiplegic migraine is a reversible neurologic condition with no underlying vascular occlusion [2,3]. Hospitalization is not typically required unless the symptoms are prolonged, severe, or raise concerns for another serious neurologic condition such as epilepsy or persistent encephalopathy [4].

A lumbar puncture is typically unnecessary, especially if there is no clinical suspicion of meningitis, encephalitis, or subarachnoid hemorrhage [15].

While hydration, sleep regulation, and avoidance of triggers are essential in migraine management, patients with hemiplegic migraine may require pharmacologic intervention to prevent recurrent attacks, and a structured pharmacologic approach is needed [16].

If attacks become more frequent or significantly affect the patient's quality of life, long-term preventive therapy should be tailored accordingly [5]. Patients should also be educated about recognizing early warning signs and seeking medical attention if symptoms deviate from their usual pattern.

Treatment should focus on migraine prevention and symptomatic relief, and stroke risk is managed through standard cardiovascular risk factor modification rather than anticoagulation.

The patient in this case was managed conservatively, with symptoms resolving within 24 hours without the need for aggressive intervention. This aligns with the expected clinical course of hemiplegic migraine, where neurological deficits typically resolve completely between attacks.

Companion Case

A companion case illustrates the typical MRI findings in hemiplegic migraine. This is a case of an 11-year-old female who presented to the emergency department with acute onset of right-sided hemiplegia and facial droop. The clinical picture was that of an acute stroke or migraine.

An urgent MRI was performed which revealed no acute infarction on diffusion-weighted imaging (Figure 4). The FLAIR sequence was normal with no parenchymal or leptomeningeal signal abnormality (Figure 5). The T2 sequence was also normal with no anatomical abnormality (Figure 5). However, the maximum-intensity projection (MIP) from the susceptibility-weighted sequence revealed characteristic dark signal (susceptibility artifact) within multiple cortical veins in the left cerebral hemisphere (Figure 6).

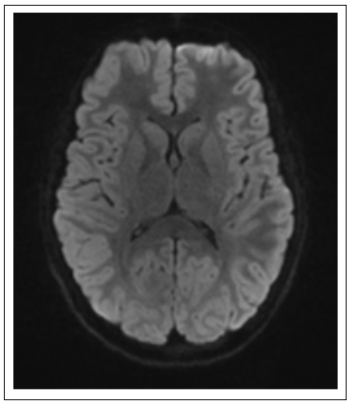

Figure 4: Axial Diffusion-Weighted Imaging

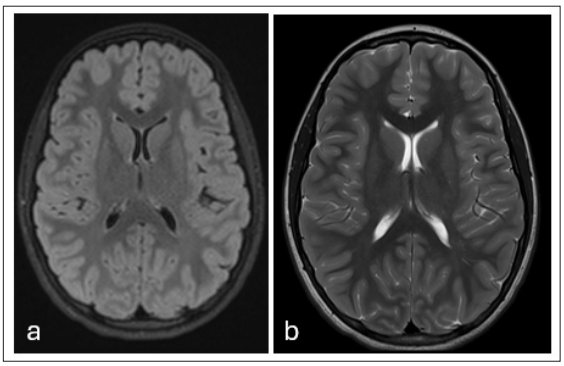

Figure 5: (a) Axial FLAIR and (b) Axial T2-Weighted Sequences

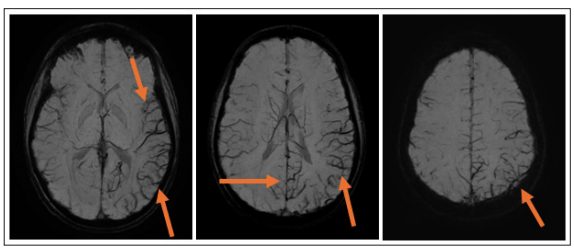

Figure 6: Axial Susceptibility-Weighted Imaging

These imaging findings are consistent with those observed in hemiplegic migraine, where susceptibility-weighted imaging (SWI) reveals prominent cortical vein susceptibility artifacts [9]. This suggests transient venous congestion or altered hemodynamics, which may contribute to the pathophysiology of the condition [12]. The absence of infarction on diffusion-weighted imaging (DWI) and the lack of parenchymal abnormalities on fluid-attenuated inversion recovery (FLAIR) and T2-weighted sequences further support the diagnosis of hemiplegic migraine rather than an acute ischemic event.

In the case of the 11-year-old female, right-sided hemiplegia and facial droop were the predominant manifestations. While these symptoms can mimic a cerebrovascular event, their transient nature, and the absence of infarction on imaging support the diagnosis of hemiplegic migraine.

Prognosis

The overall prognosis for patients with hemiplegic migraine is that most fully recover between attacks but may develop mild residual neurologic symptoms in severe cases [5]. Long-term management should include trigger identification, lifestyle modifications, and preventive medications for high-risk patients [5]. While hemiplegic migraine can significantly affect a person's quality of life, recognizing and diagnosing the condition early and instituting appropriate treatment help minimize disability and improve long-term outcomes.

Most patients experience complete resolution of neurologic symptoms between episodes. However, in some patients, particularly those with frequent or prolonged attacks, persistent neurologic deficits (e.g., mild motor weakness, ataxia, cognitive dysfunction) may develop over time [5]. Dissimilar to progressive neurologic disorders, hemiplegic migraine does not typically worsen with each episode, and long-term outcomes are generally positive with appropriate management [5].

While prolonged aura or recurrent episodes may contribute to residual symptoms, hemiplegic migraine is not a degenerative condition [7]. Most patients experience recovery between attacks, and preventive treatment can help reduce the frequency and severity of symptoms [7]. Although some pediatric patients experience a reduction in attack frequency with age, many individuals continue to experience episodes into adulthood [17]. Some studies have suggested a slightly higher risk for stroke in individuals with familial hemiplegic migraine; however, routine anticoagulation is not recommended [18].

Conclusion

Hemiplegic migraine has a characteristic imaging appearance on MRI. Recognition of these findings, knowledge of the pathophysiology, and understanding of the management are important in the diagnosis and treatment of patients presenting with Hemiplegic Migraine.

Disclosure

Figures are first author's own.

Queen's University Health Sciences ethics approval received (TRAQ #: 6043375).

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