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Hemiplegic migraine in An Adolescent Lebanese Patient.

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Abstract

We present the case of a 12-year-old female Lebanese patient who initially presented to the emergency room for sudden onset of right sided weakness and dysarthria; All the investigations turned out to be negative. A thorough history revealed a diagnosis of hemiplegic migraine, and she was treated appropriately with an excellent outcome.

Keywords: *Migraine, hemiplegic migraine, familial hemiplegic migraine, calcium channel blockers.*

Introduction

Migraine is a common disorder, affecting women more than men and it affects 15% to 20% of the population, whereas hemiplegic migraine is reported as a very rare prevalence. Hemiplegic migraine is a rare subtype of migraine with aura and is distinct because of the presence of motor weakness as an aura manifestation. Many trigger factors can play a role in inducing hemiplegic migraine A hemiplegic migraine can run in family (familial hemiplegic migraine) or happen sporadically (sporadic hemiplegic migraine). The diagnosis requires a good history, and the prognosis is usually very promising.

Case Report

In this study, we present the case of a 12-year-old girl patient who initially presented to the emergency room (ER) for sudden onset of right sided weakness, and dysarthria and severe headache.

The patient is a full-term infant born to non-consanguineous parents. She was delivered at the hospital via spontaneous vaginal delivery with a birth weight of 3500 g and head circumference of 35 cm. There were no complications during pregnancy or delivery. Her developmental history was negative: all the milestones were normal for age. Family history is positive for migraine in her mother and her aunt requiring them frequent hospitalisations.

She was hospitalized before, at the age of 7 years, because of sudden onset of weakness on the right side and severe headache, and this was the first time she had such a weakness; She was hospitalised for 5 days, with negative brain imaging, and was diagnosed as migraine, and then after 2 weeks, she was completely fine, and her weakness resolved completely, but unfortunately discharged on no treatment.

As interval history, between 7 and 12 years, she complained monthly of a headache, of 2-5 hours duration, once or twice a week, associated with photophobia, phonophobia and numbness in her extremities, without any weakness.

Upon presentation to the ER, her physical exam showed that she is pain with light avoidance, cooperative, dysarthric, right sided motor power 2-3/5 and DTR +2; On the left side, she had normal motor power of 5 /5 and DTR +2; she had proper sensation and proprioception on both sides.

Urgent CT brain was done in the ER to rule out any haemorrhagic stroke or arteriovenous malformation (AVM) and turned out to be negative.

Magnetic resonance imaging (MRI) brain, and MRI brain stroke protocol, with magnetic resonance venography (MRV) and angiography (MRA) were done to R/O ischemic stroke, or arterial or venous malformation, and all were negative.

All routine blood tests including, CBCD, electrolytes, LFT's, glucose, calcium, phosphorus, magnesium, were all negative.

The diagnosis of hemiplegic migraine was considered.

As management, she received Paracetamol IV, and NSAID to decrease her headache; but with no significant result; then we started her on Verapamil, and we noted a significant improvement of her headache, and within 24 hours of starting Verapamil, she was discharged home, with resuming of normal motor function.

Unfortunately, genetic testing was not performed because of the hight cost.

Discussion

While migraine is considered as a common disorder, as it affects 15% to 20% of the population, hemiplegic migraine shows lower prevalence of 0.01%. Moreover, a Danish study indicated that the prevalence of sporadic hemiplegic migraine is 0.002% and that of familial hemiplegic migraine is 0.003%. Women are 3 times more likely to be affected and the average age of onset is 12 to 17 years.[1][2][3][4][5]

Many trigger factors can play a role in inducing hemiplegic migraine, like sleep deprivation or excessive sleep, exertion, head trauma, and conventional angiography.[6] Hemiplegic migraine is a rare subtype of migraine with aura and is distinct because of the presence of motor weakness as an

aura manifestation. A hemiplegic migraine can run in family (familial hemiplegic migraine) or happen sporadically (sporadic hemiplegic migraine).[1]

Familial Hemiplegic Migraine (FHM) FHM is an autosomal-dominant subtype of hemiplegic migraine that runs in the family.

The diagnostic criteria for familial hemiplegic migraine requires that at least one first or second-degree relative has had attacks fulfilling the diagnostic criteria for hemiplegic migraine.

The International Classification of Headache Disorders-3 classifies FHM into 4 subtypes based on the genetic associated mutation.

- The most common type, FHM1, accounts for around 50% and is associated with mutations in the CACNA1A gene located on chromosome 19p13.
- FHM2 results of mutations in the ATP1A2 gene on chromosome 1q23, and it accounts for less than 25%.
- FHM 3 results of mutations in the SCN1A gene on chromosome 2q24.
- FHM4 is diagnosed if unknown genetic mutation.

Sporadic Hemiplegic Migraine (SHM) occurs only in person without any family history of hemiplegic migraine. [1][2][3][4]

Typically, migraine attacks start in the first or second decade of life, with the frequency of attacks decreasing with age. Unilateral motor weakness as aura manifestation in at least a few of the attacks is the typical feature of hemiplegic migraine. The weakness frequently starts in ascendant way; it starts with the hand and gradually spreads up to the arm and face. Weakness is rarely bilateral, but most often is unilateral and can alternate from one side to the other.

Aura has many forms: motor weakness being the most frequent, visual disturbances like visual field defects and scotoma are well noted, and numbness, paresthesia, ataxia, lethargy, and fever are also reported. Most cases with HM have associated headache. Headache usually occurs during the aura but can occur after the aura symptoms. The symptoms usually occur over 20 minutes to half an hour, but rarely, aura symptoms and motor weakness can develop acutely and can mimic a stroke.

The symptoms' duration can last for few hours to days and rarely can last up to 4 weeks. The symptoms resolve completely in most of the cases. The severe form of HM attacks can be associated with encephalopathy or coma. Whenever we have hemiplegia and impaired consciousness, these symptoms can last for many days to months before they resolve completely. Permanent brain injury, infarction,

cerebral atrophy, cognitive decline, and death might be rarely a result of severe attacks. Some patients with FHM, specifically with FHM2 were found to have more seizures independent of hemiplegic migraine.

The diagnosis of HM is clinical and can be challenging sometimes. A large list of differential diagnosis like transient ischemic attack, central nervous system infection (encephalitis, meningitis), brain tumors, seizures with postictal paralysis, metabolic disease (MELAS, homocystinuria and ornithine transcarbamylase deficiency), syndrome of stroke-like migraine attacks after radiation therapy. [1][2][3][4][7]. We should rule out other common pathologies that can cause headache with neurological deficit.

A detailed history is crucial for the diagnosis of HM. The key point for HM diagnosis is episodic, reversible, unilateral weakness as aura, plus at least one other kind of aura. Motor and sensory symptoms are usually more prominent in the upper extremities than lower.

Unilateral hyperreflexia with positive Babinski sign is usually seen on physical exam during acute attack. Exam between the attacks is typically normal.

As for prognosis, most patients with HM, aura symptoms resolve completely despite that sometimes they may be prolonged. Very rarely, hemiplegic migraine leads to permanent neurological deficits, cerebral infarctions, cognitive decline, or death. [8]

Treatment of HM includes avoidance of triggers if possible and involves pharmacological treatment with abortive and preventive medications like the drugs used in treating migraine with aura except for agents that may exacerbate ischemia. Severe attacks may need hospitalization. Acute episodes are managed with NSAIDs and antiemetics. Some studies support the use of intranasal ketamine at the onset of attack.

Verapamil has been used as a prophylactic and abortive agent for hemiplegic migraine with good results especially in FMH 1. Other drugs that have been used for prophylactic treatment include flunarizine, ketamine, lamotrigine, and naloxone. Acetazolamide may be effective as preventive medication. Initial therapy with verapamil, flunarizine or acetazolamide is recommended for patients with a hemiplegic migraine for preventive treatment. Patients failed on previous medications should try lamotrigine, topiramate, valproic acid, and amitriptyline. Ergotamines and triptans are usually contraindicated in a hemiplegic migraine because of concern for potential cerebral vasoconstriction.[9]10][11]

Conclusion

Hemiplegic migraine affects patients' well-being and can lead, in some cases, to dangerous outcomes. Therefore, establishing a correct diagnosis by avoiding useless investigations is very important as it provides proper treatments, and prevent attacks. In this case report, the Lebanese adolescent showed a direct improvement when the adequate treatment was administered. However, a genetic testing can be useful as it may identify the genetic variant associated to the phenotype and may provide further confirmation of the accuracy of the treatment.

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