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Case Report

Hemiplegic Migraine - Rare Cause of Hemiparesis in a 28-year-old Filipino

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Abstract

Background: Hemiplegic migraine is a very rare subtype of migraine with reported prevalence of 5 in 100,000 population. It exists in two forms - familial and sporadic, depending on the presence or absence of a family history. Most of the reported cases of this rare entity are from Europe with very few from other parts of the world.

Objectives: To report the case of an Asian female diagnosed with sporadic hemiplegic migraine and highlight the difficult aspects of acute treatment and prophylactic choice.

Methods: Case Report.

Results: We report the case of a 28-year old Filipino with 5-year history of migraine without aura who presented at the emergency room with her first hemiplegic attack associated with other aura symptoms, which included visual, somatosensory and brainstem auras. Brain Imaging and other tests excluded other potential causes of similar presentation. Her headaches did not resolve with regular intravenous acute treatment (IV paracetamol, ketoprofen and fluids) and her weakness was yet to resolve completely at day 6 after onset. She also required a special choice of prophylactic treatment to control subsequent headache attacks.

Conclusion: Sporadic hemiplegic migraine is very rare and has not been well-reported from non-European countries. This case report describes an Asian patient with many unique features and management challenges.

Keywords: *Migraine; Aura; Hemiplegic; Sporadic; Headache*

Introduction

Hemiplegic Migraine is a rare subtype of Migraine with aura in which attacks are associated with transient motor weakness or hemiparesis [1]. Recurrent motor weakness associated with migraine was first described in 1910 [2]. This Migraine variety is very uncommon, with an estimated prevalence of 5 in 100,000 population [3]. The condition exists in 2 forms - familial and sporadic, both of which share similar features except for the presence of a positive family history in at least one first-degree or second-degree relative in the familial form [1]. As at 2011, only about 100 - 200 families affected by familial hemiplegic migraine and about 200 patients affected by sporadic hemiplegic migraine had been published [4]. The attacks typically manifest with motor symptoms and are always accompanied by other aura forms (visual, sensory, aphasic) [5]. Onset age is broad, and reports have been made of onset as early as one year of age [5]. Associated additional neurological manifestations have been described in some cases including cognitive delay, seizures, cerebellar ataxia, and even loss of consciousness [5]. Three genes have been implicated in familial hemiplegic migraine, namely CACNA1A, ATP1A2 and SCN1A, all of which are dominantly inherited [5]. Though mutations in these genes can be found in some sporadic cases also, it is more likely

when age at onset of hemiplegic attacks is below 16 years and when extra neurologic symptoms are present [5]. Up to 25% of the familial forms and majority of sporadic cases do not have associated mutations in any of these three genes [4]. Diagnosis of hemiplegic migraine depends upon careful history-taking and reasonable exclusion of other potential causes of similar presentation [4].

Not very many cases of this condition have been published so far, and much of what are contained in the literature are from Western Europe [5,6] with little pockets of case reports from other parts of the world. Owolabi published the first case report from Nigeria in 2013 [7] and we found no prior case reports from United Arab Emirates during our literature search.

Case Report

Here we report the case of a 28-year-old Filipino lady who presented at our emergency room on 01 July 2019 with headache and right-sided weakness of 11 hours duration. Her symptoms had started in the morning with numbness on the right side of the face which had later spread to involve the right upper limb and later the right lower limb over several minutes. Within one hour from onset of numbness, she started to experience left sided throbbing headache which gradually increased in intensity to 10 out of 10 over the next several hours. Within 3 hours, she developed right sided facial weakness with gradual progression of weakness to involve the right upper and later the right lower limb. During evaluation at the emergency room 11 hours later she admitted having experienced photopsia earlier on in the attack which had resolved at the time of evaluation.

She had been diagnosed with migraine 5 years earlier and had suffered many attacks of migraine without aura. She had never experienced any typical migraine aura before during any of her previous attacks; hence she was terrified by this attack. Her past medical history was positive for complete thoracic transverse myelitis 2 years earlier at the age of 26 years from which she completely recovered without any sequelae. She was born to a monogamous family of unrelated Filipino parents and has 4 siblings. One of the siblings also has history of migraine without aura. No history of migraine or any neurological disease in any of the parents or grand-parents.

She is married and has 2 children but there is no history of migraine or alternating hemiplegia in any of her children. As at the time of presentation she was on combined oral contraceptive pills (COCPs) for contraception.

On neurological examination, she was conscious and oriented, had no aphasia but had mild photophobia. No meningeal signs and optic fundi were normal by direct ophthalmoscopy. There was obvious right facial paresis of the upper motor neuron variety, right hemiparesis grade 1 out of 5 with brisk reflexes and equivocal plantar response. There was also hemi-hypoesthesia involving the right half of the face and right sided limbs; light touch perception was rated 30 - 40% compared to the left side.

Brain Magnetic Resonance Imaging (MRI) with contrast and Magnetic Resonance Venography (MRV) were unremarkable as were blood tests for inflammatory markers. She was diagnosed with sporadic hemiplegic migraine as she lacked any family history to qualify for familial hemiplegic migraine. Intravenous ketoprofen and paracetamol were administered initially together with continuous Intravenous fluids at 100 milliliter/hour.

She continued to have moderate to severe headaches for 36 hours in spite of regular Intravenous paracetamol 1g every 6 hours and Intravenous Ketoprofen 50 mg every 12 hours, with only short-lived reduction in pain score immediately after administration of these medications. After 36 hours of unrelenting headache, we gave a trial of Intravenous dexamethasone 16 mg administered in 100 milliliters of saline over 20 minutes which did not lead to complete resolution of headache after 2 hours. Intravenous Magnesium sulfate was subsequently administered at a dose of 1g in 250 ml of saline over 30 minutes which led to significant reduction in pain score to 2 out of 10. Oral Naproxen was subsequently commenced in addition to amitriptyline 12.5 mg at night for prophylaxis. She reported an episode of tinnitus the following morning.

She was discharged from the hospital after 60 hours because the medical insurance no longer approved her hospital stay, and at that time limb weakness had improved to grade 3 out of 5 while facial weakness had improved to slight asymmetry noticeable only when asked to perform voluntary facial movements. Sensation in the face and limbs had also improved to 85%. She was advised to stop COCPs.

During clinic follow up 7 days after onset of symptoms, facial paresis had resolved completely but there was still mild hemiparesis grade 4 out of 5 on the right side. She reported further short episodes of headaches rated 5 - 8 out of 10 in the intervening period, and acetazolamide 250 mg bid was added to amitriptyline for prophylaxis.

Discussion

This case highlights several important facts about sporadic hemiplegic migraine.

It is a rare condition that always requires exclusion of alternative conditions [4]. In this case, we excluded inflammatory brain lesions and venous sinus thrombosis by MRI brain with contrast and MRV. Systemic inflammatory markers were also checked and found to be normal. It has been emphasized that typical hemiplegic migraine attacks would always be associated with simultaneous occurrence of at least 2 forms of aura [8]. Our patient had in addition to hemiparesis, visual aura, sensory aura and brainstem aura in the form of tinnitus occurring during the same attack. Our patient being female agrees with the literature which consistently documents higher prevalence in females [3-5].

One of the interesting features in our patient is the fact that all her previous attacks in the last 5 years had been of non-hemiplegic type and she only had her first hemiplegic attack at age of 28 years. This agrees with findings from other studies of co-existence of non-hemiplegic migraine forms in patients with hemiplegic migraine, and in fact some patients may have only one hemiplegic attack in their entire lifetime. [5]. Many other authors have described the occurrence of hemiplegic attacks in the setting of previous migraine with aura but our patient's previous attacks have been migraines without aura. It has indeed been hypothesized that hemiplegic migraine might exist on the same continuum as other forms of migraine with aura and that hemiplegia occurs in the rare instance when cortical spreading depression is strong enough to cross the central sulcus and reach the motor area [4]. Such individuals will therefore have non-hemiplegic attacks most other times when the cortical spreading depression is not strong enough to reach the motor area. However, the fact that all of our patient's previous attacks had been without aura suggests that hemiplegic migraine could also co-exist with common migraine, though this is yet to be reported by other authors.

In our case, weakness began to gradually improve after 24 hours but did not resolve completely until after 6 days. In fact, an author has suggested a range from one day to four weeks [4]. The current International Headache Classification states that motor auras generally lasts less than 72 hours [1] but some experts based on clinical experience have suggested re-introduction of 'migraine with prolonged aura' to incorporate prolonged attacks [4]. Our case supports this view. In Owolabi's report from Nigeria, weakness persisted for over 12 days [7].

In terms of treatment, there we could not find specific recommendations on acute treatment or prophylaxis in hemiplegic migraine. Some authors recommend using the same acute treatment as for other migraine phenotypes except that caution must be exercised with the use of triptans and ergotamine which are feared to have the propensity to cause ischemic brain injury in the setting of hemiplegic migraine [4]. Intravenous ketoprofen and paracetamol which constitute our hospital's usual protocol for acute migraine attack did not lead to significant relief in this case, and we tried Intravenous dexamethasone and magnesium infusion based on anecdotal reports of their efficacy [9,10]. Also, acetazolamide 250 mg bid was included in her prophylactic regimen based on reports of positive response to this medication [11]. Interestingly, her headaches have since been under control since starting acetazolamide.

Most of the cases published so far of hemiplegic migraine have been cases of European origin. Hence we hope to contribute to existing literature by reporting this case occurring in a patient of Asian origin and also to share the unique features of our case including our choice of acute treatment and prophylaxis bearing in mind the fact that there are no standard guidelines on how to best manage this rare but challenging migraine subtype.

Another interesting phenomenon that we observed in our patient was the sequence of improvement of weakness and sensation which tended to occur first in the face followed by the upper limb and thereafter the lower limb. This followed the exact order in which the symptoms had appeared. We did not observe this phenomenon in the literature.

Limitations

Our patient was not screened for genetic mutations in the three recognized genes known to be associated with hemiplegic migraine. Though we cannot rule out completely the fact that she might harbor one of these mutations, it is well documented that genetic screening is most likely to yield positive results in those with hemiplegic attack onset before age 16 years and in those with other associated neurological manifestations such as ataxia. Our patient did not have any of these features and so we did not consider genetic screening imperative.

Conclusion

We have described a very rare form of migraine - sporadic hemiplegic migraine-which occurs in about 5 out of 100,000 population. Most previous reports have been from Europe but here we have described an Asian patient who presented with weakness lasting for over 6 days and whose weakness evolved in a rather unique sequence and recovered after a similar sequence. We have also highlighted the challenging aspects of her acute treatment and prophylactic choices for which there are no clear recommendations in literature due to the rarity of the disorder. Finally, our case supports earlier reports of prolonged motor aura in hemiplegic migraine, much longer than the 72-hour duration specified in the international classification of headache disorders.

Approvals and Patient Consent

The study design was pre-approved by the Research and Ethics Committee of Belhoul Speciality Hospital Dubai, and written informed consent was obtained from the patient. The written consent gave authorization to disclose any potentially recognizable information in the report.

Disclosures

None.

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