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# Diagnosis of familial hemiplegic migraine in an intubated pregnant patient with Glasgow coma score of 3

Mehmet Kandilcik[a,*], Hafize Oksuz[b]

[a] *Pazarcık State Hospital, Department of Anesthesiology and Reanimation, Kahramanmaras, Türkiye*
[b] *Sutcu Imam University, Faculty of Medicine, Department of Anesthesiology and Reanimation, Kahramanmaraş, Türkiye*

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**Abstract**

Familial hemiplegic migraine (FHM) is a rare disease that manifests as hemiparesis and is characterized by attacks of marked motor aura, often accompanied by seizures, nystagmus, ataxia, and altered consciousness. Patients with a diagnosis of FHM may also have coma and subsequent respiratory failure requiring intubation. In this case report; FHM diagnosis process of the patient who was admitted to the reanimation intensive care unit after the cesarean section is described. In this process, the importance of detailed anamnesis obtained from the patient's relatives during the reanimation intensive care unit period in the diagnosis of FHM was emphasized, as the diagnostic imaging and laboratory tests performed on the patient were normal.



## Introduction

Hemiplegic migraine (HM) is a rare form of migraine with aura, characterized by transient motor weakness or hemiparesis (motor aura). HM is also associated with other non-motor aura symptoms (visual, sensory) and other symptoms that typically accompany migraine, such as nausea, vomiting, photophobia [1]. Motor aura represents the differentiating feature of HM compared to other forms of migraine with aura [2]. HM is classified as sporadic hemiplegic migraine (SHM) and familial hemiplegic migraine (FHM). FHM has an autosomal dominant inheritance and is rare [1].

## Case Report

A 26-year-old patient, 36 weeks pregnant, underwent emergency cesarean section under general anesthesia. Postoperatively, she was transferred in the anesthesia reanimation intensive care unit as intubated. It was learned that the patient was hospitalized in neurology intensive care unit due to seizure before cesarean section. In the neurology intensive care unit, for diagnostic purposes, MR imaging and lumbar puncture were performed, laboratory markers for viral infection were studied, and all these tests were found to be normal. Computed tomography (CT) was performed on the first day of admission to the anesthesia reanimation intensive care unit after cesarean section. CT report: bilateral basal ganglia, thalamus, internal and external capsule, both centrum semiovale are normal; ventricles are normal sized; there are no findings in favor of intracranial, intraaxial, extraaxial hemorrhage; skull is intact and there are no findings in favor of fractures in the calvarial bones. Thereupon, the patient's relatives were contacted to obtain detailed anamnesis for differential diagnoses. In the detailed anamnesis, it was reported that nausea, visual impairment and movement weakness occurred in attacks at the age of 2-3, aura symptoms lasted 2-3 hours, and muscle weakness lasted for at least half an hour and at most a week.

It was documented that she had a seizure at the age of 7 and her brain imaging and EEG results were normal. It was learned that she had attacks every 2 months during pregnancy, that she had speech impairment and muscle weakness on one side of her body, and that this situation

*Corresponding author:
*Email address:* dr.mehmetkandilcik@gmail.com (Mehmet Kandilcik)

**Table 1.** Updated ICHD Diagnostic Criteria for Familial Hemiplegic Migraine.

| ICHD III (published 2013)10 |
|---|
| A. At least 2 attacks fulfilling criteria B and C |
| B. Aura consisting of both of the following: |
| 1. Fully reversible motor weakness |
| 2. Fully reversible visual, sensory, and/or speech/language symptoms |
| C. At least 2 of the following 4 characteristics: |
| 1. At least 1 aura symptom spreads gradually over ≥5 min and/or 2 or more symptoms occur in succession |
| 2. Each individual non-motor aura symptom lasts 5–60 min, and motor symptoms last <72 h |
| 3. At least 1 aura symptom is unilateral |
| 4. The aura is accompanied, or followed within 60 min, by headache |
| D. Not better accounted for by another ICHD-3 diagnosis; and transient ischemic attack and stroke have been excluded |
| E. At least 1 first- or second-degree relative has had attacks fulfilling criteria for A–D |

Abbreviations: ICHD: international classification of headache disorders.

lasted for about 3 hours and disappeared. It was told that in the 36$^{th}$ week of her pregnancy, she went to the emergency clinic when the same attack was accompanied by a seizure, and was admitted to the neurology intensive care unit with a preliminary diagnosis of acute stroke. The patient's relatives reported that these attacks also occurred in the patient's father and cousins. After detailed anamnesis, the patient was diagnosed with familial hemiplegic migraine and treatment was started with acetazolamide, topiramate, sodium valproate, lamotrigine and non-steroidal anti-inflammatory drugs. On the second day of hospitalization, although the patient's consciousness was confused, she was extubated when it was observed that her spontaneous respiratory effort was sufficient. On the 3$^{rd}$ day of the patient's hospitalization, it was observed that the speech disorder resolved and muscle strength returned to normal.

The patient was informed about the case presentation and the consent form was signed by her on 15/01/2023.

## Discussion

FHM is a rare condition. A systematic study conducted for the epidemiology of FHM stated that the prevalence of FHM was around 0.003% [3]. In a study on FHM conducted in Denmark with 147 patients from 44 families, it was reported that women were twice as likely to experience FHM as men, and the age of onset of FHM ranged between 3 and 55 years [4]. In our case, patient was a 26-year-old woman.

The International Headache Society developed specific diagnostic criteria for SHM/FHM in 2004 in order to fully characterize and differentiate FHM disease from other subtypes of migraine and updated these diagnostic criteria in 2013 (Table 1) [5].

Diagnosis of FHM requires at least 2 attacks presenting with reversible motor weakness and at least 1 other transient neurological symptom. Motor symptoms usually last less than 72 hours but may last for days [5]. The patient in our case complies with these criteria; she had speech impairment, seizures, and muscle weakness on one side of her body, and at the end of the 3rd day, her speech impairment and muscle weakness returned to normal. The attacks in the patient's anamnesis comply with the diagnostic criteria in terms of both symptoms and duration.

For the diagnosis of FHM, in addition to the attacks, at least 1 first-degree or second-degree relative must have similar attacks. It was determined that the father and cousins of the patient in our case had similar attacks.

In a study investigating the clinical findings and first attack patterns of HM; It has been stated that HM may affect any age group, and its first symptoms often occur in pediatric age. It has been determined that early transient neurological symptoms can occur between the ages of 1 and 4 [6]. In the anamnesis of the patient in our case, it was determined that the first attack was nausea, visual impairment and weakness in movement at the age of 2-3, and the aura symptoms lasted 2-3 hours, and the muscle weakness lasted at least half an hour and at most a week. It was reported that she had a seizure at the age of 7.

In many studies conducted for HM; various factors may play a triggering role in an HM attack, and it has been reported that there is a triggering factor in up to 46% of patients. Overall, a history of minor head trauma has been identified in 45% of HM patients, and emotional stress ranks second among possible triggers [6]. Other possible triggering factors include: intense physical activity, pregnancy and fever [6,7]. In our case; since pregnancy and emotional stress trigger FHM attacks, we think that the patient had attacks every 2 months during the pregnancy.

A study by Thomsen Let al reported that coma can occur in about a third of patients with FHM, which may be accompanied by respiratory insufficiency requiring intubation, in addition to unilateral muscle weakness [4]. The patient in our case had frequent attacks during pregnancy, and in her last attack, she was admitted to intensive care due to coma and respiratory distress in addition to seizures. The most common differential diagnoses of HM are cerebrovascular diseases and epilepsy [1]. In patients with headache and neurological symptoms, brain imaging can help in the differential diagnosis of intracranial hemorrhage, ischemic stroke, tumors, and abscesses [8]. It has been reported that brain imaging results are normal in most of the attacks of FHM patients [9]. In our case; the magnetic resonance imaging performed on the day of the patient's attacks was normal, and there were no findings in favor of hemorrhage, ischemia, tumor, abscess in the postcesarean CT report. All other reported conditions that mimic HM are focal neurological deficits associated with

hypercapnia, hyponatremia, liver-renal failure, meningitis [1]. In our case, the patient's liver function tests, kidney function tests, electrolyte levels and examination results for meningitis-encephalitis were normal.

HM patients often have EEG abnormalities; these abnormalities consist mainly of asymmetric, occipital slow wave activity. In some cases, the EEG trace is normal [7]. In our case, it was learned that the EEG performed at the age of 7 years during an episode accompanied by seizure was found to be normal. Although the prognosis of FHM is thought to be benign because the symptoms are reversible, Russell MB et al. reported that in severe hemiplegic migraine attacks, impaired consciousness and prolonged aura symptoms may last from a few days to months until complete recovery is achieved [10]. In our case, the patient's treatment continued in the intensive care unit due to a severe attack accompanied by confusion and seizures, and the patient's speech impairment and muscle strength returned to normal on day 3.

## Conclusion

The diagnosis of hemiplegic migraine is a diagnosis of exclusion and is made on the basis of anamnesis with routine biochemical, hematologic, lumbar puncture and imaging methods. It is observed that the FHM diagnostic criteria are based on the physical examination findings of the patient and the history of the disease. Therefore, we believe that obtaining detailed anamnesis from relatives of coma patients in intensive care unit is very important in the diagnosis of familial hemiplegic migraine.

## References

1. International Headache Society (IHS). The International Classification of Headache Disorders, 3rd ed.; International Headache Society: London, UK, 2018; Volume 38, pp. 1–211.
2. de Boer I, Hansen JM, Terwindt GM. Hemiplegic migraine. Handb Clin Neurol. 2024;199:353-365.
3. Eriksen M.K., Thomsen L.L., Olesen J. Implications of Clinical Subtypes of Migraine with Aura. Headache 2006, 46, 286–297.
4. Thomsen LL, Eriksen MK, Roemer SF, Anderson I, Olesen J, Russell MB. A population-based study of familial hemiplegic migraine suggests revised diagnostic criteria. Brain. 2002;125:1379-1391.
5. Headache Classification Subcommittee of the Inter- national Headache Society. The International Clas- sification of Headache Disorders, 3rd edition (beta version). Cephalalgia. 2013;33:629-808.
6. Toldo I, Brunello F, Morao V, Perissinotto E, Valeriani M, Pruna D, et al. First Attack and Clinical Presentation of Hemiplegic Migraine in Pediatric Age: A Multicenter Retrospective Study and Literature Review. Front. Neurol. 2019, 10, 1079.
7. Barros J, Ferreira A, Brandão A.F, Lemos C, Correia F, Damásio J. et al. Familial Hemiplegic Migraine Due to L263V SCN1A Mutation: Discordance for Epilepsy between Two Kindreds from Douro Valley. Cephalalgia 2014, 34, 1015–1020.
8. Evans R.W. Diagnostic Testing for Migraine and Other Primary Headaches. Neurol. Clin. 2019, 37, 707–725.
9. Bonemazzi I, Brunello F, Pin JN, Pecoraro M, Sartori S, Nosadini M, Toldo I. Hemiplegic Migraine in Children and Adolescents. J Clin Med. 2023 May 31;12(11):3783.
10. Russell MB, Ducros A. Sporadic and familial hemiplegic migraine: Pathophysiological mecha- nisms, clinical characteristics, diagnosis, and management. Lancet Neurol. 2011;10:457-470.