Prolonged Symptoms in Sporadic Hemiplegic Migraine: Aura or Migrainous Infarction?

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Abstract-

Purpose: Sporadic hemiplegic migraine is a rare form of migraine associated with motor weakness during the aura phase. While the aura usually lasts <1 hour, patients with sporadic hemiplegic migraine frequently have prolonged weakness.

Case Report: A 60-year-old male had sporadic hemiplegic migraine after a head injury at the age of 14. He presented to our emergency department with a typical migraine attack except prolonged right limbs weakness and numbness (>1 day). Brain magnetic resonance imaging showed an acute infarction in the left posterior medial pons. He recovered completely from motor weakness but still complained of residual numbness in his right limbs three months later.

Conclusion: We report the first adult case of sporadic hemiplegic migraine with migrainous infarction located in the pons. Since patients with hemiplegic migraine often have prolonged aura, it is easy to be confused with a migrainous infarction. The case report highlights that migrainous infarction is a complication difficult to diagnose and treat early, especially in patients with hemiplegic migraine.

Key Words: brainstem, cortical spreading depression, migrainous infarction, migraine with aura, sporadic hemiplegic migraine

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INTRODUCTION

Hemiplegic migraine is defined as a subtype of migraine with aura (MwA) associated with motor weakness during the aura period[(1)]. Based on the presence of similar symptoms in a first or second degree relative, the diagnosis of hemiplegic migraine can be classified as familial or sporadic hemiplegic migraine (SHM). SHM is an uncommon form of migraine with an estimated prevalence of 0.002%[(2)]. SHM differs from other types of MwA in the duration of the aura. While most auras last <1 hour, SHM auras can last >1 day. Thus, its diagnosis can be confused with that of migrainous infarction (MI)[(2)]. SHM is associated with various paroxysmal and permanent neurological symptoms such as epilepsy and cerebellar dysfunction[(3)], and occurs extremely rarely in conjunction with MI.4 We report the first case of SHM with MI in an adult and discuss the

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possible mechanism underlying MI development.

CASE REPORT

A 60-year-old male had a history of hypertension and smoking. He suffered from MwA SHM after receiving a head injury at the age of 14, without family history of similar symptoms. The presentation of his aura was typical and included blurred vision, right limbs numbness and weakness, dysarthria and diplopia. The aura lasted nearly one hour, followed by a severe throbbing headache with nausea, vomiting, photophobia, and phonophobia which lasted 4-24 hours. These symptoms occurred two or three times a year, but he did not seek medical help. In January 2011, he had another similar aura attack followed by a headache. However, the symptoms of weakness and numbness persisted and he visited our emergency department the next day. Neurological examination revealed right hemiparesis MRC (Medical Research Council) grade 4/5 and hypesthesia. Brain magnetic resonance imaging (MRI) showed an acute infarction in the left posterior medial pons (Fig. 1). Concomitant magnetic resonance angiography (MRA) revealed no significant vascular lesion in the vertebrobasilar system but a focal stenosis in the left distal middle cerebral artery (Fig. 2). He recovered completely from motor weakness four days after the episode. A MRI one month later confirmed the diagnosis of cerebral infarction (Fig. 3). Three months later, he still complained of residual numbness in his right limbs.

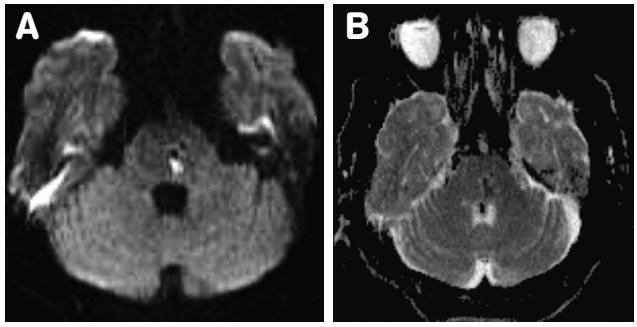

Figure 1. (A) Diffusion-weighted imaging and (B) apparent diffusion coefficient scans revealed an acute infarction in the left posterior medial pons.

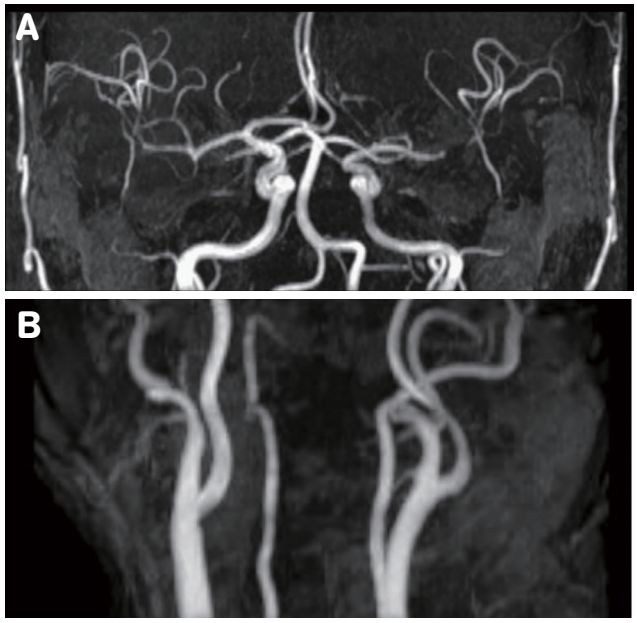

Figure 2. (A) Intracranial magnetic resonance angiography (MRA) showed focal stenosis of left distal middle cerebral artery. No significant atherosclerosis was noted in the vertebral and basilar arteries. (B) Extracranial MRA disclosed no obvious atherosclerosis.

DISCUSSION

This report describes the case of an elderly male patient with a history of SHM who developed MI in the pons. Patients with SHM could have prolonged neurological deficits, sometimes associated with hyperintensity changes in diffusion weighted imaging and reduction in apparent diffusion coefficient values (Fig. 1)[4]. We repeated brain MRI one month later and confirmed the diagnosis of established MI (Fig. 3). His residual right limbs numbness three months later supported the diagnosis. To our knowledge, there is only one similar case in a 10-year-old child[5]. A study of 105 patients with SHM found that all patients had motor aura lasting 7 hours as an average and 8% of them had weakness lasting longer than one day[2]. Clinicians may lose the opportunity to diagnose and treat MI early because the initial presentation of MI can easily be mistaken for prolonged aura.

Migraine with aura is a well-established risk factor

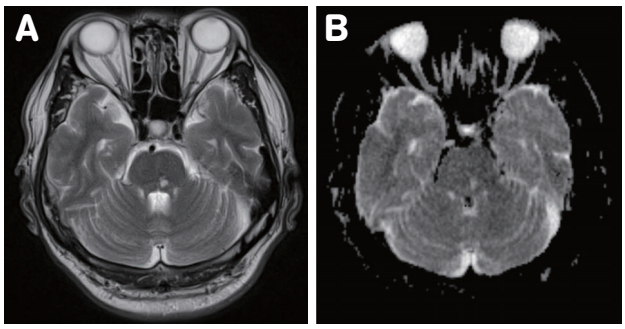

Figure 3. Magnetic resonance imaging one month later confirmed the established cerebral infarction in (A) T2-weighted and (B) apparent diffusion coefficient images.

for cerebral infarction but its mechanism is controversial[6]. One of the hypothesis is that cortical spreading depression (CSD), the probable cause of migraine aura[7], is the mechanism underlying the development of MI. CSD is associated with transient hyperemia followed by prolonged oligemia, tissue damage, and hypoxia in animal studies[8,9]. The location of MI in the posterior circulation also suggests that CSD is the mechanism leading to MI, since the occipital cortex is the likely source of visual aura where CSD usually occurs[1,7,10]. Other possible factors include patent foramen ovale, vasospasm, artery dissection, and use of triptans or ergots. Considering the location of MI in this patient, cardioembolism was not likely. We did not find evidence for vertebral or basilar artery dissection, though transient vasospasm could not be completely excluded. The patient denied using triptans or ergots to treat his headache.

In our patient, MI occurred at an uncommon location (the pons). In the largest recent Nordic study, two out of 33 patients (6%) had MI in the brainstem[10]. Based on the following reasons, we propose that CSD propagated to the brainstem is the cause of MI in our patient. First, both of the Nordic patients had basilar-type aura, as did our patient (diplopia). The link between basilar-type aura and MI in the brainstem seems to support our hypothesis, though the number of cases is too small to warrant a definite conclusion. Second, patients with SHM may have higher chances for their CSD propagating to the brainstem. It has been reported that patients with SHM have multiple types of aura, occurring mainly in the sequence-visual, sensory, motor, aphasic, and basilar-type[2]. This phenomenon suggests that CSD originates in the occipital cortex and propagates forward to other brain areas such as the sensory cortex, motor cortex, and brainstem. Although basilar-type aura is the least frequent type of aura in patients with MwA (10%)[3], it occurs in 72% of patients with SHM[2]. By definition, patients with SHM must have motor aura, indicating propagation of CSD through the central sulcus to the primary motor cortex. It suggests that CSD in patients with SHM is more capable to propagate, because the central sulcus has been proved to be a barrier to CSD propagation[11]. In our patient, the other possibility was that CSD occurred in the brainstem[12]. A previous study found the absence of visual aura in 9% of patients with SHM, which means CSD does not always involve the occipital cortex[2]. Though our patient experienced right limbs numbness, the location of MI seemed to be distant from the medial lemniscus and spinothalamic tract. The reason could be individual variation in anatomy.

Our patient reported suffering from headaches after receiving a head injury when he was 14 years old. Head injury has been noted as a trigger in patients with SHM but not in patients with other types of MwA[3]. It has been shown that CSD may occur after head injury and the relationship between SHM and head injury deserves further investigation[13].

Considering the rare occurrence of MI in patients with SHM, it is prudent to distinguish traditional atherosclerotic cerebral infarction from MI. Our patient had several cardiovascular risk factors: male, relatively old age, hypertension, and smoking. His MRA revealed focal stenosis of left distal middle cerebral artery; however, no obvious atherosclerotic changes were noted on his MRA in the vertebrobasilar system and extracranial carotid arteries (Fig. 2). We could not exclude the possibility that these risk factors and atherosclerosis interact with CSD to cause MI. The two diseases, SHM and cerebral infarction, might coincidently occur in the patient, but the temporal relationship argued against the possibility.

In brief, this is the first case of MI in an adult patient

with SHM that highlights the difficulty in differentiating MI from prolonged aura. Patients and clinicians should be aware of the potential risk of MI to treat early.

Conflict of interest: The authors declare that there are no conflicts of interest.

REFERENCES

1. Headache Classification Committee of the International Headache Society. The International Classification of Headache Disorders, 2nd ed. Cephalalgia 2004;24:1-160.
2. Thomsen LL, Ostergaard E, Olesen J, Russell MB. Evidence for a separate type of migraine with aura: sporadic hemiplegic migraine. Neurology 2003;60:595-601.
3. Russel MB, Ducros A. Sporadic and familial hemiplegic migraine: pathophysiological mechanisms, clinical characteristics, diagnosis, and management. Lancet Neruol 2011; 10:457-470.
4. Toldo I, Cecchin D, Sartori S, Calderone M, Mardari R, Cattelan F, Laverda AM, Drigo P, Battistella PA. Multimodal neuroimaging in a child with sporadic hemiplegic migraine: A contribution to understanding pathogenesis. Cephalalgia 2011;31:751-756.
5. Ribeiro RT, Pinto MM, Villa TR, Gamba LT, Tengan CH, de Souza-Carvalho D. Migrainous infarction as a complication of sporadic hemiplegic migraine in childhood. Arq Neuropsiquiatr 2009;67:906-908.
6. Schurks M, Rist PM, Bigal ME, Buring JE, Lipton RB, Kurth T. Migraine and cardiovascular disease: Systematic review and meta-analysis. BMJ 2009;339:b3914.
7. Hadjikhani N, Sanchez Del Rio M, Wu O, Schwartz D, Bakker D, Fischl B, Kwong KK, Cutrer FM, Rosen BR, Tootell RB, Sorensen AG, Moskowitz MA. Mechanisms of migraine aura revealed by functional MRI in human visual cortex. Proc Natl Acad Sci USA 2001;98:4687-4692.
8. Gursoy-Ozdemir Y, Qiu J, Matsuoka N, Bolay H, Bermpohl D, Jin H, Wang X, Rosenberg GA, Lo EH, Moskowitz MA. Cortical spreading depression activates and upregulates MMP-9. J Clin Invest 2004;113:1447-1455.
9. Takano T, Tian GF, Peng W, Lou N, Lovatt D, Hansen AJ, Kasischke KA, Nedergaard M. Cortical spreading depression causes and coincides with tissue hypoxia. Nat Neurosci 2007;10:754-762.
10. Laurell K, Artto V, Bendtsen L, Hagen K, Kallela M, Meyer EL, Putaala J, Tronvik E, Zwart JA, Linde M. Migrainous infarction: a Nordic multicenter study. Eur J Neurol 2011;18:1220-1226.
11. Grenier E, Dronne MA, Descombes S, Gilquin H, Jaillard A, Hommel M, Boissel JP. A numerical study of the blocking of migraine by Rolando sulcus. Prog Biophys Mol Biol 2008;97:54-59.
12. Richter F, Bauer R, Lehmenkuhler A, Schaible HG. Spreading depression in the brainstem of the adult rat: Electrophysiological parameters and influences on regional brainstem blood flow. J Cereb Blood Flow Metab 2008; 28:984-994.
13. Lauritzen M, Dreier JP, Fabricius M, Hartings JA, Graf R, Strong AJ. Clinical relevance of cortical spreading depression in neurological disorders: migraine, malignant stroke, subarachnoid and intracranial hemorrhage, and traumatic brain injury. J Cereb Blood Flow Metab 2011;31:17-35.