Recurrence Transient Hemiparesis in a Patient with a Giant Persisting Eustachian Valve and Patent Foramen Ovale: Atypical Hemiplegic Migraine or Paradoxical Cerebral Embolism?

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Abstract

We encountered a patient with the overlapping disorders of migraine with aura, migraine-triggered seizures and recurrent transient hemiparesis caused by atypical hemiplegic migraines with motor weakness during headache attacks, but not during the aura period, or paradoxical cerebral embolism. The patient displayed a giant Eustachian valve and patent foramen ovale, through which a spontaneous right-to-left shunt was revealed on transesophageal echocardiography. We considered that the overlapping disorders in the present case were closely related to the spontaneous right-to-left shunt caused by the giant Eustachian valve.

Key words: Eustachian valve, migraine with aura, migraine-related epilepsy, paradoxical cerebral embolism, patent foramen ovale, migrainous stroke

(Intern Med 52: 1523-1525, 2013)
(DOI: 10.2169/internalmedicine.52.0200)

Case Report

A 23-year-old woman presented with recurrent episodes of right hemiparesis associated with headaches. Bilateral pulsating headaches with photophobia, phonophobia and nausea had started seven years earlier. The headaches were initially accompanied by scintillating scotomas in the right visual hemifield that lasted 30 minutes. The patient’s symptoms were responsive to oral rizatriptan and aggravated by routine physical activity. The headaches occurred at a frequency of once per week and lasted 12 to 24 hours. The patient was diagnosed with MA. Recurrent episodes of right hemiparesis occurring since 20 years of age started in the right hand and traveled to the right side of the face and right leg over a few minutes followed by MA. The symptoms would reach maximum severity (3/5 strength) within 20 minutes, last from half an hour to a few hours and resolve spontaneously. The symptoms were not associated with any obvious provocation. Occasionally, the episodes of hemi-
Figure.  (A) Brain MR imaging with a FLAIR sequence showing a high-signal intensity lesion in the left basal ganglia (arrow). (B) \textsuperscript{99m}Tc-ECD SPECT performed three days after the development of transient right hemiparesis with a mild headache and impaired consciousness showed hypoperfusion in the left temporo-occipital lobe. (C) Transesophageal echocardiogram using contrast saline detected a giant persisting EV and PFO, through which a spontaneous right-to-left shunt was found without the Valsalva maneuver (below). No pulmonary hypertension was observed. RA: right atrium, LA: left atrium

paresis switched to the left side, developed suddenly and were not associated with migraine attacks. A close review of the patient’s medical history revealed that loss of consciousness with seizures had occurred occasionally within one hour from the resolution of migraine aura since 22 years of age. Episodes of both hemiparesis and loss of consciousness often coincided with the migraine attacks. The patient’s past medical history was negative for hypertension, diabetes mellitus, heart disease and smoking, and she did not use oral contraceptives or illicit drugs. She had no family history of either migraines or cerebrovascular disease.

On magnetic resonance (MR) imaging of the brain, fluid-attenuated inversion recovery (FLAIR) images showed a high-signal intensity lesion in the left basal ganglia (Figure A), although diffusion-weighted images and MR angiography were unremarkable. Electroencephalography revealed a few spiky waves in the bilateral central-parietal regions. \textsuperscript{99m}Tc-ethylcysteinate dimer single photon emission computed tomography (\textsuperscript{99m}Tc-ECD SPECT) performed three days after the development of transient right hemiparesis with a mild headache and impaired consciousness showed hypoperfusion in the left temporo-occipital lobe (Figure B). The remaining diagnostic workup, which included a complete blood count, coagulation studies, basic metabolic and thyroid function tests, an electrocardiogram, an X-ray of the chest and transthoracic echocardiography, was normal. However, transesophageal echocardiography detected a giant persisting EV and patent foramen ovale (PFO), through which a spontaneous right-to-left shunt was noted without the Valsalva maneuver (Figure C). No pulmonary hypertension was observed. Venous Doppler ultrasound of the lower extremities failed to reveal any venous thrombosis. The patient began taking clopidogrel to prevent cerebral embolism but declined percutaneous PFO closure. After several episodes, valproate therapy was started, which controlled both the epileptic episodes and migraine attacks. The headache attacks occurring once a month were attenuated with nonsteroidal anti-inflammatory drugs.

Discussion

HM is a rare form of MA associated with motor weakness during the aura period (2). The clinical manifestations of HM can occur as a sporadic or familial disorder and range from attacks with short-duration hemiparesis to severe forms with recurrent coma and prolonged hemiparesis, permanent cerebellar ataxia, epilepsy, transient blindness and mental retardation (2). The motor symptoms of HM often
begin insidiously in contrast to the vascular events and epileptic seizures that develop suddenly (2). The symptoms can be bilateral or unilateral, switching from attack to attack or always involving the same side (2). Although some of the clinical features in our case appear to be appropriate for a diagnosis of sporadic HM, the atypical presentation of motor weakness during the headache attacks, not the aura period, does not strictly fit the diagnostic criteria for ICHD-II (1). If an ischemic event cannot be excluded sufficiently, as in our case, the patient must be screened for thromboembolic disease.

The most intriguing aspect of our case was the presence of a giant persisting EV and PFO. Several data in the literature indicate that EV can initiate the development of a spontaneous right-to-left shunt and promote not only MA, but also paradoxical cerebral embolism in patients with PFO (3-5). Although we were unable to identify the origin of the thrombus, it may have originated in situ or resulted from adhesion of embolic material from a distant source. We considered both mechanisms of the expression of atypical HM and paradoxical cerebral embolism.

The relationships between MA, ischemic stroke and epilepsy are complex and bidirectional; therefore, no simple cause-effect links can be established (6). Migraines themselves may, in rare instances, trigger strokes or epilepsy or, exceptionally, both (7), as in the present case.

The pathophysiology of migrainous infarction is currently unknown. It has been hypothesized that cortical spreading depression (CSD), the neurophysiological phenomenon underlying migraine aura, may lead to migrainous infarction if coupling between neuronal metabolism and the cerebral blood flow is disturbed (8, 9). This hypothesis is consistent with the findings of previous reports showing that CSD in patients with HM occurs in the frontal lobe, including the motor cortex, of the affected hemisphere (10). Epilepsy is associated with cortical hyperexcitability. Human and animal in vitro epilepsy models suggest that epileptiform cortical hyperexcitability is often paralleled by the occurrence of spreading depression (10). This could explain the link between epilepsy and MA observed in the present patient. Regional oligemia consequent to CSD may further decrease the epileptic threshold of neurons, initiating epileptic discharge in already susceptible neurons (11). Once an ischemic lesion has developed, it may in turn cause cortical remodeling and become a source of focal epileptic activity.

The diagnostic workup revealed a FLAIR hyperintense lesion, which is often found in migraine patients (12), and PFO, whose prevalence is elevated in migraine patients (13). The decrease in the cerebral blood flow observed on SPECT must have decreased the epileptic threshold but remained above the ischemic threshold, since cerebral ischemia did not develop. Iizuka et al. reported that prolonged aura symptoms in patients with familial HM are frequently associated with hyperperfusion and middle cerebral artery vasodilatation (14), although the perfusion state can differ depending on the time course of the migraines or the timing of scanning in relation to CSD.

In brief, this is a rare case of MA with transient hemiparesis that highlights the difficulty in differentiating paradoxical cerebral embolism from atypical HM. If ischemic events cannot be excluded as the cause of hemiparesis in patients with HM, the patients must be screened for thromboembolic diseases, including EV and PFO.

The authors state that they have no Conflict of Interest (COI).

References