Symptomatic Migraine with Prolonged Visual Aura and Unruptured Occipital Arteriovenous Malformation

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Abstract

A 35 year-old woman had a two-year history of recurrent headache with clinical presentations of visual aura in her left visual field followed by right-sided throbbing headache. The patient suffered from a similar attack but her visual aura-like symptoms persisted for over 48 hours. The concurrent electroencephalogram demonstrated focal non-epileptiform rhythmic slow waves in the right occipital region. The magnetic resonance images showed prominent parenchymal edema in the right occipital area. The cerebral angiographic study proved a small cerebral arteriovenous malformation. This illustrated case showed that cerebral arteriovenous malformation produces headaches mimicking migraine with visual aura. The acute vascular flow change and the parenchymal edema trigger a prolonged visual aura with coinstantaneous evidence of cortical depression shown on the electroencephalogram.

Keywords: **Symptomatic migraine; Prolonged visual aura; Unruptured arteriovenous malformation**

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INTRODUCTION

It is known that intracerebral hemorrhage and epileptic seizures are the most common complications of cerebral arteriovenous malformation (cAVM). Chronic non-hemorrhagic headache as an initial presentation of cerebral AVM is reported in 14.9-47% of patients with cAVMs, [1-3] and this warning symptom is often neglected. For decades, many clinicians reported that patients with cAVM experienced recurrent migrainous headaches, especially cAVM located in the occipital lobe[3,4]. The pathophysiology of the concurrence between the cerebral AVM and the migrainous headache with aura is not clear, and there is speculation that the complex vascular structure and vascular flow change of cAVM can trigger cortical spreading depression (CSD), activate the trigeminovascular nerve afferents, and generate the pain. Here, we illustrated an unusual case experiencing recurrent migraine with aura and suffering from a prolonged visual aura when a small occipital AVM without obvious hemorrhage developing severe regional cortical edema. While the patient suffered from prolonged visual aura, the electroencephalogram showed focal slow waves in the corresponding area, which indicated regional cortical

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depression. To our knowledge, cAVM causing migrainous headache and prolonged visual aura has not been reported before.

CASE ILLUSTRATION

A 35 year-old healthy woman started to have headaches about two years before this admission. Her attacks were always proclaimed by visual symptoms consisting of phosphenes in her left visual field for 20 minutes, while right-sided throbbing headache developed shortly after the scintillating visual bright spots subsided. The throbbing headache usually subdues about 30-60 minutes later. No pain-relieving medicines for the headache attacks were taken. The headache attacks were not frequent, but mostly provoked by stress, poor sleep, menstruation, or not drying her wet hair immediately. She asked for medical consultation during her first and early pregnancy, and the neurological examinations and electroencephalography (EEG) were normal. She remained headache-free though her pregnancy and on her maternity leave. When she was ready for work about three months after her child-birth, she had a sudden and severe headache along with persistent blurred vision. This time she had to visit one local clinic for pain-relieving medicine. However, her visual symptoms did not vanish. She described a persistent visual snow-like blur in the entire visual field, which was mingled with paroxysmal scintillating white spots emerging over the left visual field lasting for 10 to 15 minutes every one hour. She also noted intermittent throbbing but tolerable headaches over right posterior head every one to two hours. Two days later, she revisited our clinic for the prolonged and annoying visual symptoms. Neurological examinations revealed visual blockage over her left lower visual field. She received a magnetic resonance image (MRI) study of the brain soon after her transferring to the emergent unit. Prominent parenchymal edema over right occipital region was revealed on MRI T2 sequence images without obvious hemorrhage noted on T2*-weighted gradient-echo (GRE) sequences. (Figure 1A, 1B) Also, a few small high-flow vessels were seen on Time Of Flight (TOF) magnetic resonance angiography

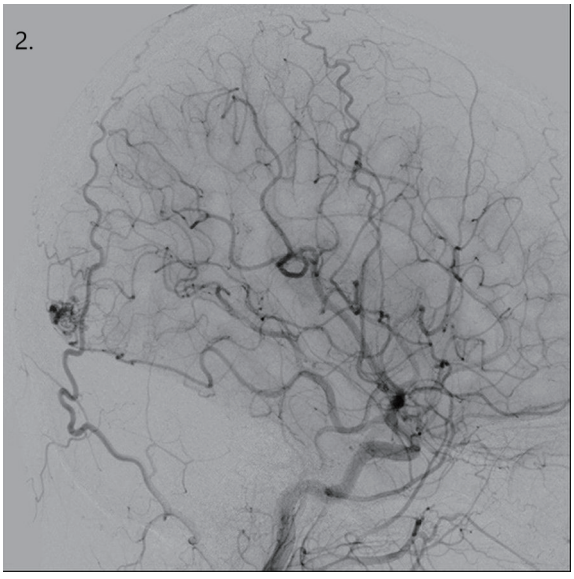

Figure 2. Lateral right external carotid angiogram demonstrated arterial supply to the right occipital AVM nidus from the right posterior cerebral artery branch.

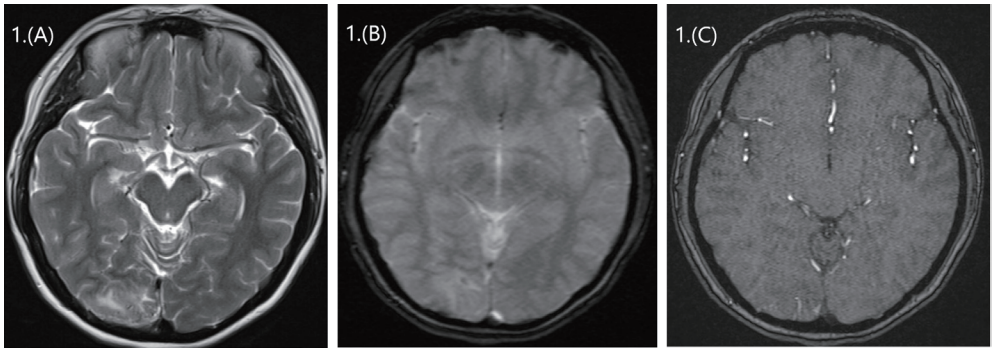

Figure 1. (A) Axial T2-weighted and (B) Axial T2* GRE MR image revealed edematous change at the right occipital lobe without evidence of parenchymal hemorrhage. (C) TOF MRA axial source image showed some small high-flow vessels.

(MRA) sequences (Figure 1C), and a vascular structure lesion located near the right occipital area was suspected. Following cerebral angiography, a small nidus in the right occipital region with a feeding artery and drainage veins from right posterior cerebral artery was disclosed, which confirmed the diagnosis of an occipital arteriovenous malformation with Sptzler-Martin grade 2. (Figure 2) In the meantime, the emergent EEG revealed focal non-epileptic rhythmic theta waves with focus on the right occipital area. (Figure 3A, 3B) Her visual symptoms did not respond to either steroid or Sumatriptan, but began to subside after getting Lamotrigine treatment. The prolonged visual symptoms completely disappeared for about 10 days after she took 50 mgs of oral lamotrigine twice a day. After the discussion of the strategies of evacuating the AVM, she preferred radiosurgery. A few months after receiving gamma-knife radiosurgery, although she still had few brief headaches over the right occipital region, there were barely any visual aura symptoms or throbbing characters. All the follow-up EEGs were normal without any focal slow waves.

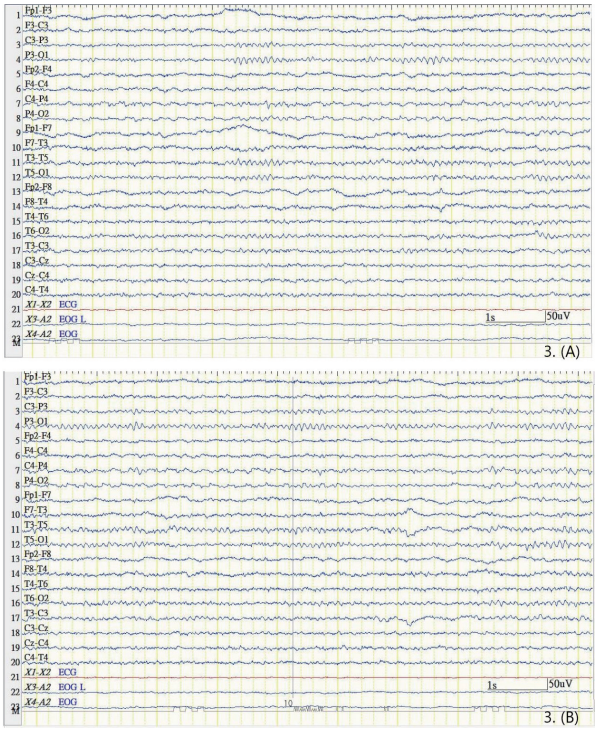

Figure 3. (A and B) The electroencephalograms reveal focal non-epileptiform rhythmic theta waves at right occipital area.

DISCUSSION

Our presenting patient was first diagnosed of migraine with aura, because her clinical characteristics were typical migraine-like headache with visual aura which fulfilled the official criteria by the International Classification of Headache Disorders 3nd Edition (ICHD-III). Until the development of an unusual prolonged visual aura, thorough studies were arranged and finally confirmed a small cerebral AVM located in the corresponding occipital area. The association between the migrainous headache and the cerebral vascular lesion is worth discussing.

Much research has been done on the concurrence of migraine with aura and unruptured cerebral AVM since 1970. A few retrospective studies or registered data from the neurovascular centers also disclosed a high incidence of migrainous headache in the patients with cAVM. One study even revealed a high incidence of patients with migraine-like headaches and non-ruptured cAVM if the lesions were located in the occipital lobe (56.1%, 23/41) [4]. Furthermore, one study revealed a strong positive correlation (88.8%) between the site of AVM and the side of the migraine type headache[1]. Although the effects of surgical removal or radiosurgery of the cAVM on migrainous attacks were not consistent[2,4], many patients resulted in dramatic or total resolution of all symptoms. The mechanism of the concurrence of cerebral AVM and migrainous headache with aura is still unclear although several hypotheses for the pathophysiology were proposed.

Besides, CSD is known as the electrophysiological basis of migraine aura. The CSD, first described by Aristides Leão in 1944, is a slow propagating wave of strong neuronal and glial depolarization accompanied by the depression of EEG activities[6]. Very few clinical and EEG feature correlations were reported. One study described that the concomitant EEG abnormalities of the five patients suffering from prolonged aura all showed continuous focal slow waves over the corresponding regions, and the EEG depression activities resolved when the aura symptoms subsided.[7] The CSD activates trigeminal nociception and thus triggers a headache[6,8,9]. More interestingly, evidence from animal studies proved that CSD is highly associated with changes of the regional cerebral blood flow[6,10]. Also, our patient's clinical symptoms and EEG features correlated well with the

evolution of the focal parenchymal edema related to the unruptured cAVM.

As for why lamotrigine was chosen to control the prolonged aura in our case, there were studies with accumulating evidence suggesting that lamotrigine was highly effective in reducing the frequency and intensity of migraine aura[11,12]. It was speculated that lamotrigine interfered with presynaptic voltage-gated ionic channels, which were the keys for spreading depolarization and glutamate release during CSD[13].

If the regional blood flow change of cAVM could initiate CSD and activate the trigeminal-vascular nerve afferents, then it was possible that the eradication of cAVM should greatly relieve headache symptoms. Two studies reported that the effectiveness of headache alleviation after the evacuation of cAVM in over 85% of the patients with recurrent migrainous attacks[2,4]. There were clinical improvement in our presenting case, too.

Our illustrating case showed an unpredictable vascular flow and hemodynamic change of a small unruptured cAVM evolved in severe focal brain swelling, triggered CSD, and produced the prolonged visual symptoms clinically. The patient's unusual clinical manifestations and coinstantaneous EEG findings revealed that a pathological structure like cAVM could produce symptoms mimicking migrainous headache and visual aura.

CONCLUSION

Vascular structural lesions as occipital unruptured cAVMs can be presented as migraine-like headaches with visual aura. Acute vascular changes of the unruptured cAVM with focal parenchymal edema can produce prolonged aura-like symptoms.

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