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Case Report

A Case of Reversible Cerebral Vasoconstriction Syndrome Requiring Differential Diagnosis with Migraine

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Abstract

A 32-year-old postpartum woman with a history of migraine-like headaches consulted us for throbbing headache which had started two days prior during defecation after alcohol drinking. Although computed tomography (CT) of her head revealed no abnormal findings, fluid-attenuated inversion recovery magnetic resonance imaging (FLAIR-MRI) showed sulcal hyperintensity. Magnetic resonance angiography (MRA) showed multiple cerebral vasoconstrictions, and cerebral angiography showed multifocal caliber changes of cerebral arteries, including vasodilatation and vasoconstriction. It took 13 days for her headache to resolve; cerebral vasoconstriction had disappeared on MRA performed 3 months later. The diagnosis was reversible cerebral vasoconstriction syndrome (RCVS), which has been reported to tend to be misdiagnosed as migraine. Although triptan drugs are widely used to treat the common disease of migraine, RCVS should be taken into consideration in the treatment of migraine-suspected patients because triptan drugs can induce or worsen RCVS, potentially resulting in ischemic stroke. In our case, a thunderclap-like onset pattern of headache and triggers such as being postpartum, alcohol drinking and Valsalva maneuver of defecation were useful information for the diagnosis of RCVS. Even in an era of advanced brain imaging, careful inquiry is still essential in treating severe headaches.

Keywords: Reversible cerebral vasoconstriction syndrome; Headache; Migraine

Introduction

Reversible Cerebral Vasoconstriction Syndrome (RCVS), for which the diagnostic criteria were not yet published in The International Classification of Headache Disorders, 2nd Edition (ICHD-2), but were published in The International Classification of Headache Disorders, 3rd edition, beta version (ICHD-3 beta) in 2013, is characterized by thunderclap headache and multifocal segmental cerebral vasoconstriction [1-3]. Moreover, it has been reported that RCVS can be misdiagnosed as migraine [3-5]. In this paper, we present a case of RCVS requiring differential diagnosis with migraine.

Case Presentation

In 2012, a 32-year-old woman with a history of migraine-like headache was transferred to our hospital due to a severe throbbing headache with nausea which started during defecation after drinking alcohol. She was three months postpartum and a habitual drinker. Loxoprofen sodium hydrate at 180 mg/day had been prescribed by the former doctor because she had no abnormal finding on physical examination, neurological examination or brain computed tomography (CT). However, she consulted us two days later without remission. Her blood pressure, heart rate and body temperature was 150/70 mmHg, 64/min and 36.8^{O}C, respectively. She was normal in physical and neurological exam without abnormal findings on repeat brain CT. However, fluid-attenuated inversion recovery magnetic resonance imaging (FLAIR-MRI) showed sulcal hyperintensity on the bilateral parietal lobe (Figure 1A). Magnetic resonance angiography (MRA) showed multiple vasoconstrictions of the cerebral arteries (Figure 1B). Cerebral spinal fluid (CSF) was watery clear with a pressure of 20 cm H_2O, 8 neutrophils/μl, 20 lymphocytes/μl, 64 mg/dl proteins and 58 mg/dl glucose. No abnormal value was recognized in her blood cell count, blood coagulation ability, blood chemistry or serum immunological tests including antinuclear antibody, perinuclear anti-neutrophil cytoplasmic antibody (P-ANCA) and cytoplasmic anti-neutrophil cytoplasmic antibody (C-ANCA).

After admission, she underwent intravenous drip infusion

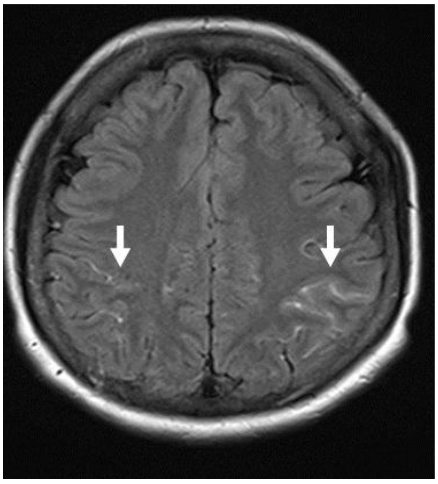

Figure 1A: The FLAIR-MRI on admission showed sulcul hyperintensity on the bilateral parietal lobes (arrow).

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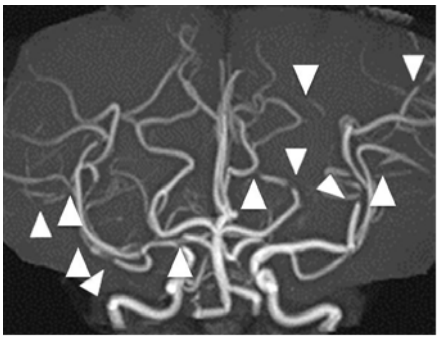

Figure 1B: The MRA on admission showed no aneurysms, but multifocal vasoconstrictions of the cerebral arteries (arrowhead).

of lactate Ringer solution for inappetence and continued taking loxoprofen sodium hydrate. On the 4th day of illness, cerebral angiography showed multifocal arterial caliber changes including both vasodilatation and vasoconstriction (Figure 2). Pregabalin at 150 mg/day was additionally administered but was ineffective against her headache. On the 8th day of illness, 20mg of sumatriptan nasal spray following lomerizine hydrochloride administration at 10 mg/day starting on the 7th day of illness remarkably improved her headache. She was discharged from our hospital without neurological deficit on the 9th day of illness. Her headache had completely disappeared by the 11th day of illness except transient tolerable headache on the 13th day of illness. She consulted us 3 months later for follow-up FLAIR-MRI and MRA, which showed disappearance of the sulcal hyperintensity and the multiple vasoconstrictions (Figure 3). Her symptoms were diagnosed as headache attributed to RCVS.

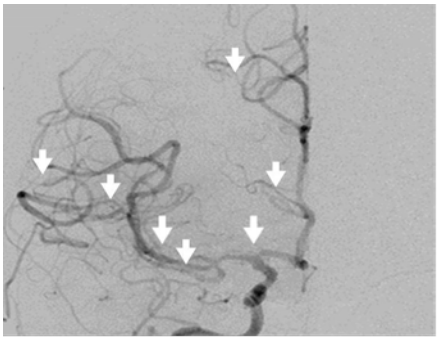

Figure 2A: The anteroposterior view of right carotid angiography showed multiple caliber changes of the cerebral arteries (arrow).

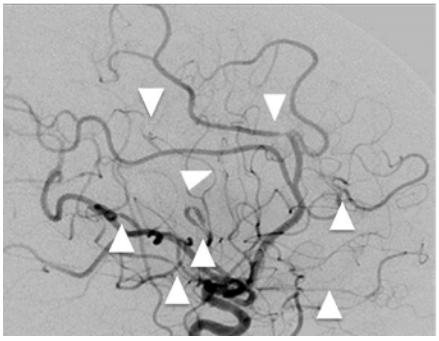

Figure 2B: The lateral view of right carotid angiography showed multiple caliber changes of the cerebral arteries (arrowhead).

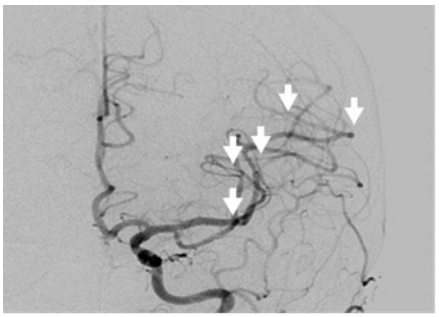

Figure 2C: The anteroposterior view of left internal angiography showed multiple caliber changes of the cerebral arteries (arrow).

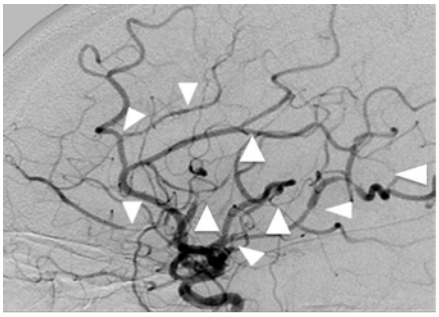

Figure 2D: The lateral view of left carotid angiography showed multiple caliber changes of the cerebral arteries (arrowhead).

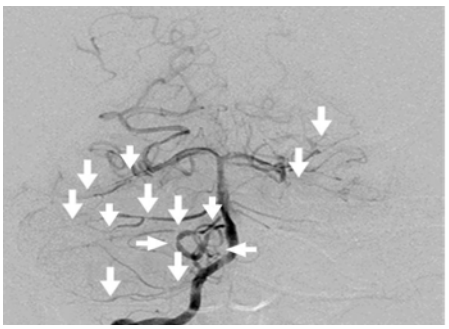

Figure 2E: The anteroposterior view of right vertebral angiography showed multiple caliber changes of the cerebral arteries (arrow).

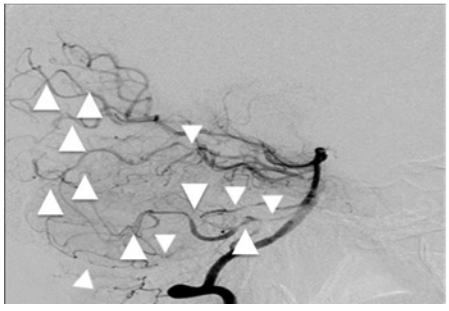

Figure 2F: The lateral view of right vertebral angiography showed multiple caliber changes of the cerebral arteries (arrowhead).

Discussion

Our patient had a history of migraine-like headaches, and her severe throbbing headache had lasted 2 days before presentation.

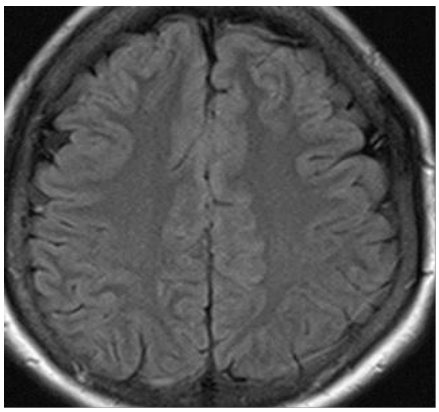

Figure 3A: The FLAIR-MRI performed 3 months later showed the disappearance of sulcul hyperintensity from bilateral parietal lobes.

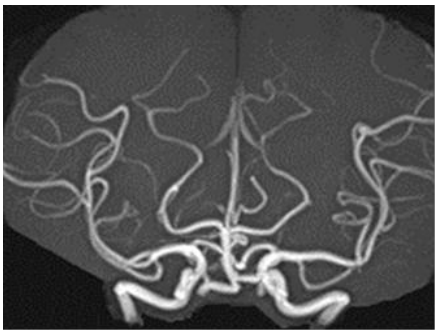

Figure 3B: The MRA performed 3 months later showed the disappearance of multifocal vasoconstrictions of the cerebral arteries.

Without MRI, we might have diagnosed her headache as migraine. Although the exact time to the peak of her headache was unknown, her headache reached a peak during the short time of her defecation. Therefore, her headache seemed to meet the definition of a thunderclap headache, which reaches a peak within one minute and lasts more than 24 hours [2]. Thunderclap headaches occur secondary to underlying diseases such as cerebral aneurysm, dissection of cerebral artery, subarachnoid hemorrhage (SAH), pituitary apoplexy, cerebral venous thrombosis, central nervous system angiitis and benign angiopathy of the central nervous system [2]. In this case, sulcal hyperintensity on FLAIR-MRI proved not to be SAH by CSF examination. Dissections were considered unlikely because the caliber changes of cerebral arteries revealed by MRA and cerebral angiography were reversible and atypically located for dissection [6]. MRI did not reveal pituitary apoplexy. Cerebral angiography did not reveal cerebral aneurysm or cerebral venous thrombosis. Central nervous system angiitis was considered unlikely because her headache improved without steroids or immunosuppressant drugs [2]. The tests of antinuclear antibody, P-ANCA and C-ANCA did not indicate systemic vasculitis [2]. The criteria of benign angiopathy of the central nervous system (BACNS) are diffuse, severe headache of abrupt or progressive onset with or without focal neurological deficits and/or seizures; a string-and-beads appearance on angiography and subarachnoid hemorrhage ruled out by appropriate investigations; and a headache (and neurological deficits, if present) that resolves spontaneously within 2 months [2]. Although her headache met these criteria, the concept of RCVS, not yet listed in ICHD-2, was also considered. RCVS is characterized by thunderclap headache, multifocal segmented vasoconstriction of cerebral arteries, resolution of vasoconstriction within 12 weeks, non-appearance of SAH by ruptured cerebral aneurysm, and normal CSF test [7,8]. RCVS occurs more often in women from 20s to 60s and in postpartum period [9], and hardly ever occurs in persons 15 years or younger [3]. Although approximately 30% of RCVS occurs without triggers, it has been reported that alcohol drinking, bathing, sexual activity, Valsalva maneuver, triptan preparation and stimulant drugs induce RCVS [1,3,9-11].

Her CSF test indicated an elevation of proteins and white blood cells. It has been reported that proteins and white blood cells increase in 10 to 50% of RCVS patients [12,13]. She was the appropriate age for RCVS and was a postpartum habitual drinker. She had drunk 500 ml of beer just before the onset, and her headache started during a Valsalva maneuver of defecation. Therefore, RCVS was a more appropriate diagnosis for her headache than BACNS. Moreover, our case met the diagnostic criteria of RCVS listed in the ICHD-3 beta published in 2013 [1].

The dysfunction of cerebral vasoconstriction has been regarded as a cause of RCVS [3,13]. While approximately 30% of RCVS are accompanied by intracranial hemorrhage, vasoconstriction occurs even without hemorrhage [12].

RCVS tends to be misdiagnosed as postpartum angiopathy, BACNS and especially migraine [13]. It is difficult to differentiate migraine and RCVS only with brain images because sulcal hyperintensity on FLAIR-MRI and multifocal cerebral vasoconstrictions in migraine patients have been reported [8,14,15].

The average time to peak of the migraine has been considered to be 60 to 90 minutes [16]. Although the time to peak of her headache was considered to be too short to fit this profile, thunderclap headache in migraine patients has been reported [17]. Snyder first reported a case of young woman's cerebral vasoconstriction that was difficult to distinguish from central nervous system angiitis in 1978 [3,18]. Similar diseases have been reported as Call-Fleming syndrome, postpartum angiopathy and drug-induced vasoconstriction [3,10]. The concept of RCVS was proposed by Calabress et al. in 2007 and 16 to 52% of RCVS patients have histories of migraine, which is a trigger of RCVS. Therefore, migraines with sulcal hyperintensity on FLAIR-MRI, vasoconstriction or thunderclap headache might easily be confused with RCVS [8-10,14,19].

Importantly, the mechanism of headache due to RCVS has not yet been clarified; vasoconstriction seems not to be the cause of the headache because the headache does not necessarily coincide with the vasoconstriction. No specific medicine has been established, but steroid, calcium blocker and magnesium sulfate have been reported to be effective [3,20]. On the other hand, it has been reported that the throbbing headache is due to the dilated portions of the cerebral arteries [21]. Therefore, triptan, which is a 5-hydroxytryptamine agonist and a specific medicine for migraine, might be effective to relieve the throbbing headache in our case. However, the vasoconstrictor action of the triptan drugs might worsen the vasoconstrictions of cerebral arteries and induce ischemic stroke. We suspected that depolarization of the cerebral cortex might be involved in her headache and administered pregabalin because pregabalin

exerts analgesic action by inhibiting cerebral cortex depolarization and is not a vasoconstrictor [22]. Then, we prescribed sumatriptan in concert with a calcium channel blocker of lomerizine hydrochloride and intravenous drip infusion of lactate Ringer solution to prevent excessive vasoconstriction and ischemic stroke, after informed consent with the patient and her mother. However, while sumatriptan seemed very effective on her headache, it might have been a spontaneous remission because the headaches of 90% of RCVS patients resolve spontaneously within a week [3,8,10]. Although triptan drugs are widely used as the specific medicine for migraine, the possibility of RCVS should be taken into consideration because triptan drugs can not only trigger RCVS but also worsen the vasoconstriction and induce ischemic stroke [11,23]. The complete remission rate of RCVS has been reported as approximately 95% [24]. However, the incidence rates of cortical SAH, intracranial hemorrhage and cerebral infraction have been reported as approximately 30%, 20% and 5%, respectively, and approximately 40% of RCVS patients with hemorrhagic strokes suffer from disability [10,12]. In addition, it has been reported that the patients of RCVS with histories of migraine have a high risk of hemorrhagic stroke [12]. The preventive therapy for RCVS is removal of triggers; hence, we encouraged our patient to discontinue drinking and not to strain during defecation [20,25].

Our case had a typical onset pattern of headache and triggers which were useful information for the diagnosis of RCVS. Even in an era of advanced brain imaging technology, careful inquiry is still essential in treating severe headaches.

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