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Reversible Cerebral Vasoconstriction Syndrome Triggered by Contact With Water in a Swimmer

Andrea Ndoka[1], Deepali Gupta, MD[2], Vivek Mehta, MD[2], and Amr Hosny, MD[2]

Background: Reversible cerebral vasoconstriction syndrome (RCVS) is a disorder characterized by segmental vasoconstriction of cerebral arteries, which leads to thunderclap-type headaches. These headaches peak in intensity within one minute and are often brought on by common triggers, such as contact with water, Valsalva maneuver, physical exertion, sexual activity, and use of vasoactive drugs, among others.

Case Report: A 40-year-old woman presented with a history of several episodes of severe headaches precipitated by swimming at work. After an inconclusive computed tomography, magnetic resonance imaging, and magnetic resonance angiography of the brain, transcranial Doppler revealed constriction of the left middle cerebral artery. Treatment with verapamil led to resolution of symptoms after 2 months.

Conclusions: RCVS is likely underdiagnosed and carries with it a small risk for potentially fatal complications. Differentiating RCVS headaches from migraines, as well as other headache disorders, is essential, and the present case provides an opportunity to further clarify the delineation between them.

Key words: Reversible cerebral vasoconstriction syndrome, RCVS, thunderclap headache, blood pressure surge, migraine

BACKGROUND

Reversible cerebral vasoconstriction syndrome (RCVS) is a cerebrovascular disorder that typically presents with severe acute headache with or without other neurological symptoms. Patients most commonly present with thunderclap headaches, which are characterized as acute severe headaches that peak in intensity in less than one minute (1). In RCVS, these headaches are usually recurrent and resolve spontaneously within 3 months (2). Around 80% of patients also describe specific triggers for their headaches, such as sexual activity, physical exertion, intense emotions, urination, bowel movements, sudden standing or sitting, and contact with water (3).

Though the prognosis is typically excellent for 78% to 90% of patients, according to a recent review of case reports (4,5), complications, such as subarachnoid hemorrhage, infarction, and cerebral edema do occur and a mortality rate between 1% and 5% exists. RCVS is thought to be underdiagnosed, underscoring the need for a complete workup in potential RCVS cases to prevent complications. Diagnosing this condition promptly can be a challenge though, due to how it can be easily mistaken for a migraine (3). In the present case, the patient's triggers of swimming, bowel movements, and abrupt standing are in line with commonly reported triggers; she was diagnosed with RCVS by transcranial Doppler (TCD) after several inconclusive studies.

CASE

The patient provided the Health Insurance Portability and Accountability Act of 1996 compliant informed con-

From: [1]Touro University Nevada, Henderson, NV; [2]The Advanced Headache Center, New York, NY

Corresponding Author: Deepali Gupta, MD, E-mail: deepali927@gmail.com
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sent. A 40-year-old woman with a past medical history significant for smoking 4 packs per week presented to our headache center with a new onset 2-week severe headache that started while doing a breath hold for her dive test at work. She is a zoologist and performs regular swim tests. The week prior to the onset of her headache she states she was swimming without any issue. On March 20, 2021, she dove into the pool and began experiencing a sharp severe pain in the center and side of her head, described as the worst headache of her life. She states that this happened again while swimming, as well as several times over the next 2 weeks while having a bowel movement or sitting up abruptly. She went to the emergency room where her blood pressure was 177/120. Her blood pressure is usually around 110/70. Computed tomography (CT) of the head was done and per the patient, was normal. She was discharged and advised to follow-up with a headache specialist.

In our office, she presented without a headache. Her blood pressure was taken: 179/120 on the left wrist; 149/110 on the right wrist. History and physical exam did not demonstrate any other significant findings. She denied exposure to water where parasites may be found. A full workup, including labs and magnetic resonance imaging (MRI) of the brain, was ordered. She had another episode the following day and went to the emergency room where her workup was done, including an MRI and magnetic resonance angiography (MRA) of the brain. This came back inconclusive. A TCD was then ordered at which point severe constriction of her left middle cerebral artery (MCA) was noted. She was diagnosed with RCVS and started on verapamil 40 mg 3 times a day, increased to 180 mg daily after 2 weeks. Her symptoms resolved after 2 months.

DISCUSSION

Considering RCVS as a differential diagnosis in a patient presenting with severe headache is important given the potential for serious or fatal complications. Reported complications include subarachnoid hemorrhage, intracerebral hemorrhage, cerebral aneurysm , brain herniation secondary to cerebral edema, and cerebral ischemia, and hemorrhagic stroke (5-7). Though only few fatal cases have been reported, 12 cases as of 2019, it is of note that 92% of those cases were women at an average of 42 years old (5). It is well reported that RCVS is more common in women than in men and is most common in women during the fourth decade of life (2,8); our patient is a 40-year-old woman. Postpartum and pregnant women, in particular, seem to be at a higher risk for RCVS and complications that can accompany it, so much so that it has been called "postpartum angiopathy" in this demographic (9,10). Of the 12 fatal cases mentioned, 6 of them were in pregnant or postpartum women (5).

It is not uncommon for those who are diagnosed with RCVS to have a history of migraines (2,3,11,12), making it sometimes difficult to differentiate a migraine headache from a thunderclap headache seen in RCVS. In addition, headaches associated with RCVS can have characteristics that overlap with features commonly associated with migraines, such as photophobia, phonophobia, nausea, and vomiting (2). Blurry vision and visual field defects have also been reported in RCVS patients (13,14), similar to symptoms classically seen in those who have migraines with auras (1).

Our patient did not have any ophthalmological symptoms, however, she did have multiple recurrent headaches over a 2-week period. While there is no agreed upon strict diagnostic criteria, recurrent thunderclap headaches over the span of several days is proposed to have near 100% sensitivity and specificity in diagnosing RCVS (12). With regards to differentiating RCVS from migraine headaches, high frequency of attacks, particularly 2 or more attacks occurring over several days, should raise suspicion for RCVS and prompt diagnostic testing.

Despite the mentioned similarities, certain qualities of thunderclap headaches can also be used as differentiating factors. While migraines can have acute onsets, thunderclap headaches have very acute onsets and are characteristically described as severe headaches that peak in intensity within one minute (15). In terms of triggers, RCVS headaches are associated with several common precipitants. These frequently described triggers include chronic cannabis use, vasoconstrictive drug use, selective serotonin reuptake inhibitor use, contact with water, Valsalva maneuver, defecation, emotional stress, physical exertion, and sexual activity (2,4,16). Of these common triggers, only emotional stress, physical exertion, and sexual activity are described as being common precipitants of migraines as well (17).

Diagnosing RCVS requires first recognizing the thunderclap headache and ruling out other headache types, followed by comprehensive imaging of the brain. The importance of ruling out migraine cannot be overstated since treatment with triptans, as is routine in patients

with migraines, can enhance vasoconstriction and worsen symptoms in RCVS patients (3). Thunderclap headaches are always considered to be secondary to other disorders, commonly subarachnoid hemorrhage, unruptured cerebral aneurysms, pituitary apoplexy, ischemic stroke, intracerebral hemorrhage, and RCVS; thus, necessitating a complete workup that includes vascular imaging studies (6,15).

MRA and CT angiography are commonly used to reveal the segmental vasoconstriction present in RCVS patients and have about 80% sensitivity in detecting RCVS, although they have decreased sensitivity in detecting vasoconstriction in distal vessels (18). In our patient, CT and MRI of the brain, as well as MRA of the brain, returned inconclusive results. TCD revealed constriction of our patient's left MCA and confirmed the diagnosis of RCVS. In a prospective study (19) of 67 patients who were diagnosed with RCVS, only 9% had normal initial TCD and MRA studies. In such cases where noninvasive imaging yields no results, cerebral digital subtraction angiography (DSA) can be considered; DSA is believed to have 100% sensitivity in detecting vasoconstriction, especially in distal vasculature that is otherwise difficult to visualize (18).

Though the precise pathophysiology remains unclear, RCVS is characterized by reversible segmental vasoconstriction of cerebral vasculature (20). Around one-third of patients experience blood pressure surges during acute attacks, just as this patient did when she presented to the emergency room (2,3). This patient presented to our office with a marked surge in blood pressure, but, interestingly, had an episode the following day. Such a surge in blood pressure the day before an attack has not been reported. This raises the possibility that triggers and/or surges in blood pressure may not always immediately lead to episodes, but may sometimes precede an episode by at least one day. One of the prevailing hypotheses of the pathogenesis of RCVS suggests that sympathetic overactivation, caused by a trigger of some sort, leads to transient deregulation of cerebral vascular tone and a subsequent thunderclap headache (3,20). This hypothesis implicates very little temporal delay between sympathetic activation and cerebral arterial tone dysregulation. The delay seen in this case further confirms what many investigators have noted; the pathogenesis of RCVS is unclear and proposed mechanisms are speculative.

CONCLUSIONS

This case presents an opportunity to highlight the need for a prompt and comprehensive workup in patients, particularly women in their 40s and 50s, presenting with severe headache. Though fatality is uncommon, a complete workup with cerebral DSA is necessary in differentiating RCVS from a severe migraine attack. It has been reported that RCVS-related headaches have been misdiagnosed as migraine attacks and treated with triptans, which are vasoconstrictors and may worsen RCVS symptoms. Especially considering the female preponderance for both migraines and RCVS, women presenting with severe recurrent headaches must receive a full diagnostic workup, which includes neuroimaging. Failure to properly workup and diagnose RCVS could lead to inappropriate use of vasoconstrictive drugs, such as triptans, subsequent exacerbation of RCVS symptoms, and complications that could have otherwise been avoided.

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