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Case Report

# Reversible cerebral vasoconstriction syndrome manifesting as focal seizures without a thunderclap headache: A pediatric case report

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**Abstract**

We report a pediatric case of reversible cerebral vasoconstriction syndrome with focal seizures without a thunderclap headache. A 7-year-old girl had a mild acute headache with nausea after swimming. She subsequently developed hemi-convulsions followed by right hemiplegia. Brain magnetic resonance angiography revealed generalized vasoconstriction of the main cerebral peripheral arteries. Her hemiplegia was spontaneously resolved within 6 h. Over the next 24 h she suffered from recurrent and transient headaches, which recurred on days 3 and 5. Follow-up magnetic resonance angiography on day 3 documented the multifocal narrowing of the main cerebral arteries, which was observed to have diminished at 12 weeks after her initial presentation. She did not have any headaches or neurological deficits after day 5. This case indicates that reversible cerebral vasoconstriction syndrome should be considered in children with focal seizures even when they do not present with thunderclap headaches. The timely and appropriate evaluation by magnetic resonance angiography and imaging is essential for diagnosing reversible cerebral vasoconstriction syndrome.


*Keywords:* Reversible cerebral vasoconstriction syndrome; Focal seizure; Hemiplegia

## 1. Introduction

Reversible cerebral vasoconstriction syndrome (RCVS) is a clinical and radiographic syndrome that is characterized by thunderclap headache due to reversible cerebral vasoconstriction. In adulthood, the prevalence of RCVS increases with aging; the mean age of the RCVS patients in the literature is 42–43 years (range: 4 months–65 years) [1–4]. The syndrome rarely affects infants or children [5–11] (Table 1). RCVS is diagnosed based on the observation of multifocal narrowing of the cerebral arteries on cerebrovascular imaging; this usually diminishes within 12 weeks after the onset of symptoms. We herein report a pediatric case of RCVS in a patient with focal seizures without thunderclap headache, which was probably triggered by the physical stress of swimming.

## 2. Case report

A 7-year-old girl suffered an acute mild headache and nausea after undergoing extensive physical stress from 2-h of swimming. She subsequently experienced a hemi-convulsion which spontaneously resolved within 5 min. She was transferred to our hospital after experiencing another hemi-convulsion followed by right-sided hemiplegia, which spontaneously resolved within

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Table 1
Clinical and radiological features of children with reversible cerebral vasoconstriction syndrome in the published literature.

| Age sex | Triggers | TCH | Clinical symptom | BP | Underlying disease | Migraine patient/family | Brain bleeding |
|---|---|---|---|---|---|---|---|
| 13 M [5] | Deep diving | YES | Vomiting Photophobia | N.D. | None | NO/YES | NO |
| 10 M [6] | Nasal spray | YES | Vomiting | 130/92 | None | NO/YES | NO |
| 16 M [6] | Defecation | YES | None | 152/87 | None | NO/N.D. | NO |
| 10 M [6] | Head banging | YES | Phonophobia | Normal | None | NO/N.D. | NO |
| 15 M [7] | Hypertension | YES | Photophobia Phonophobia | 210/140 | None | NO/N.D. | NO |
| 12 M [8] | Eletriptan | YES | Paralysis of lower extremities | N.D. | None | NO/YES | YES SAH |
| 7 F [9] | Cyclosporine | YES | Blurred lower right vision | 150/100 | Aplastic anemia | NO/N.D. | YES IPH |
| 13 M [10] | Swimming Sumatriptan | YES | None | N.D. | None | YES/NO | NO |
| 9 M [11] | Subclavian artery dissection | YES | Vomiting/seizure visual disturbance | 154/101 | LDS | N.D./N.D. | YES SAH |
| 7 F (this case) | Swimming | NO | Vomiting | Normal | None | NO/YES | NO |

M, male; F, female; TCH, thunderclap headache; BP, blood pressure; N.D., not described; SAH, subarachnoid hemorrhage; IPH, intraparenchymal hemorrhage; LDS, Loeys–Dietz Syndrome.

6 h. On admission, a blood analysis (complete-blood-count, electrolytes, liver and renal function tests) and cerebrospinal fluid examination revealed no abnormalities, and immunological analysis in serum were all negative such as rheumatoid factor, or anti-nuclear, anti-cardiolipin and anti-neutrophil-cytoplasmic antibodies. Electroencephalography demonstrated low amplitude of background activity in the left hemisphere without apparent paroxysmal discharges. Brain magnetic resonance imaging (MRI) revealed several trivial ischemic lesions, none of those seemed to be responsible for the patient's right-sided hemiplegia. The abnormal findings by DWI were almost correlated to FLAIR signals and showed a relationship with the clinical symptoms. The constant modality of MRI applied was 1.5T every time in the serial studies. Brain magnetic resonance angiography (MRA) revealed generalized vasoconstriction of the left cerebral arteries (Fig. 1). She had recurrent headaches that spontaneously resolved three times within the 5 days after the first attack. An MRA study on day 3 revealed multifocal and tapered narrowing of the main cerebral arteries and abnormal dilated segments of the peripheral arteries. The generalized vasoconstriction of the left cerebral arteries was not apparent at this time (Fig. 2A). On day 6, she was neurologically intact and was discharged from our hospital. An MRA study at 12 weeks after this episode showed no cerebral vasoconstriction or ischemic changes in the brain (Fig. 2B), and the abnormal signals in FLAIR almost disappeared at 12 weeks after the onset. She was doing well without any neurologic deficits or seizures.

## 3. Discussion

The present case highlights two important clinical issues. First, children with RCVS might present with focal seizures without a thunderclap headache. Second, an MRA study is especially important for making a proper diagnosis of RCVS.

According to the recent review articles [1–4], thunderclap headache occurs in almost all patients with RCVS. For this reason, it is considered to be a key symptom for the diagnosis of RCVS. RCVS is usually triggered by specific conditions or events, including pregnancy, orgasm, acute stressful or emotional situations, bathing, physical exertion, or the intake of vasoactive drugs. Generalized convulsions are less common at the time of presentation [1–4]; it is estimated that convulsion is caused by ischemia in the motor cortex due to decreased blood flow, which is caused by a spasm of the cerebral arteries. Although the present case did not present with a thunderclap headache, serial MRI and MRA studies demonstrated findings characteristic to RCVS. About 30–70% of patients with RCVS initially showed no abnormal findings by brain imaging soon after the