

# Case report of a rare condition of thunderclap headache

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Submitted: 02 June 2020

Approved: 09 June 2020

Published: 12 June 2020

**How to cite this article:** Lwin Z. T., Yong Z., Case report of a rare condition of thunderclap headache. *G Med Sci.* 2020; 1(2): 001-004. <https://www.doi.org/10.46766/thegms.neuro.20060201>

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## ABSTRACT

This is a report to create awareness among clinicians on a very rare condition of thunderclap headache. A 51-year-old lady presented with recurrent thunderclap headaches over the preceding 3 days. This presentation sounds like Subarachnoid haemorrhage initially, but further investigation revealed reversible cerebral vasoconstriction syndrome. In this report, the clinical presentation, investigation and management for Reversible Cerebral Vasoconstriction Syndrome (**RCVS**) including its outcome are described.

**Keywords:** Thunderclap, Reversible Cerebral Vasoconstriction Syndrome, subarachnoid haemorrhage

## INTRODUCTION

Reversible Cerebral Vasoconstriction Syndrome is underdiagnosed and can be misdiagnosed. RCVS is characterized by reversible multifocal narrowing of the cerebral arteries heralded by sudden thunderclap, severe headaches with or without neurological deficits. RCVS are clinically important because they affect young persons and can be complicated by ischemic or hemorrhagic strokes. RCVS is usually self-limited, and resolution of headaches and vasoconstriction occurs over a period of days to weeks.

## CASE REPORT

A 51-year-old lady presenting with severe headache was admitted to Acute medical unit, Ninewells hospital, after suffering from 3 days at home. On further questioning, she described the pain as thunderclap in nature lasting for a few hours. It was accompanied by nausea and vomiting but no fever, no neck rigidity nor blurred vision. Between episodes, she was totally free of pain. On examination, there was no neurological deficit and fundus scope examination revealed normal. Her blood tests were also normal.

In view of thunderclap headache, she underwent an urgent CT brain within 10 hours of admission and followed by LP and both of which reported as normal findings. However, as her symptoms were not better and ongoing, hence, history and physical examination were reviewed again and proceeded MRI head and MRA [figures 1 and 2] on second day of admission which reported some apparent focal narrowing of the branches of PCA's on both sides and left MCA suggesting possibility of RCVS, particularly in the given clinical context.

After the proper diagnosis a couple of days after admission, she had been commenced on Nimodipine 30 mg QDS with adequate analgesia on the same day of diagnosis and consequently, the pain was significantly improved in 24 hours. As a treatment, she continued nimodipine for 3 weeks with tapering doses. Surprisingly, her symptoms resolved completely following 3 weeks of the treatment and repeated MRI head scan which was carried out after 3 weeks of completing nimodipine showed no abnormality.

## DISCUSSION

Reversible cerebral vasoconstriction syndrome (RCVS) is a group of disorders characterized by severe headaches and a narrowing of the blood vessels in the brain. RCVS is reversible and patients often recover within three months; the condition is frequently missed and is more common than most physicians realize. Serious complications, such as a stroke, can be associated with RCVS if not promptly diagnosed and treated.

RCVS happens when persistent contraction of the blood vessels (vasoconstriction) causes arteries to narrow. This reduces blood flow and oxygen delivery to the affected area of the body. When vasoconstriction affects the blood vessels of the brain, it is called cerebral vasoconstriction. symptoms may include seizures, changes in vision, difficulty in speaking and weakness on one side of the body. Most patients with RCVS recover completely. In terms of gender, females aged 20 to 50 are most common.

There are some possible external factors such as over-the-counter or illegal drugs that are related to RCVS. Some examples of prescription medications are SSRIs, Triptans, Bromocriptin etc. Illegal drugs in association with RCVS are Marijuana, cocaine, Ecstasy, AMphetamine and Lysergic acid Diethylamide (LSD).

Since the most common clinical manifestation of RCVS is recurrent thunderclap headache, the diagnosis should always be considered in this setting. The immediate differential diagnosis of RCVS is subarachnoid haemorrhage which is more common and that can be diagnosed by CT head, followed by CSF examination. Remember that CT head will be normal in majority of patients with RCVS. MRI and magnetic resonance angiography brain and vessels of the head and neck can be helpful to narrow the differential diagnosis.

Magnetic resonance angiography displays diffuse segmental arterial constriction in up to 90% of cases, and a second study may be required 1-2 weeks later to demonstrate the abnormalities. Large and medium-sized arteries are more commonly affected. Catheter angiography remains the gold standard test to demonstrate the characteristic 'string of beads' pattern of alternating areas of arterial stenosis and dilation in cases where noninvasive vascular imaging is inconclusive. By definition, the cerebrovascular abnormalities are transient, and should demonstrate complete resolution on repeat imaging 1-3 months later. Treatment of RCVS is currently based on expert opinion and reported case series. Provided that any illicit or over-the-counter medications cause this problem, discontinuation of those offending drugs is prudent. The commonly used drugs are Calcium channel blockers, such as nimodipine, nifedipine or verapamil. A commonly reported regimen consists of an initial intravenous administration of nimodipine at a rate of 1-2 mg/h, followed by an oral regimen (30 - 60 mg every 4 h) tapered over the course of several weeks. Treatment is typically given for 4-8 weeks, although the optimal duration is unclear. The efficacy of treatment varies considerably between reports, ranging from 40% to 80%. The variability in reported treatment success may result from small sample sizes and/or variable definitions of treatment success, which have included cessation of headaches, resolution of vasospasm, or lack of further neurological symptoms and/or stroke. In refractory cases, intra-arterial nimodipine, papaverine or milrinone have been used with good results reported, although experience is limited to case reports.

## Images for reversible cerebrovascular vasoconstriction

### Figures 1 and 2 displaying Reversible Cerebrovascular Vasoconstriction

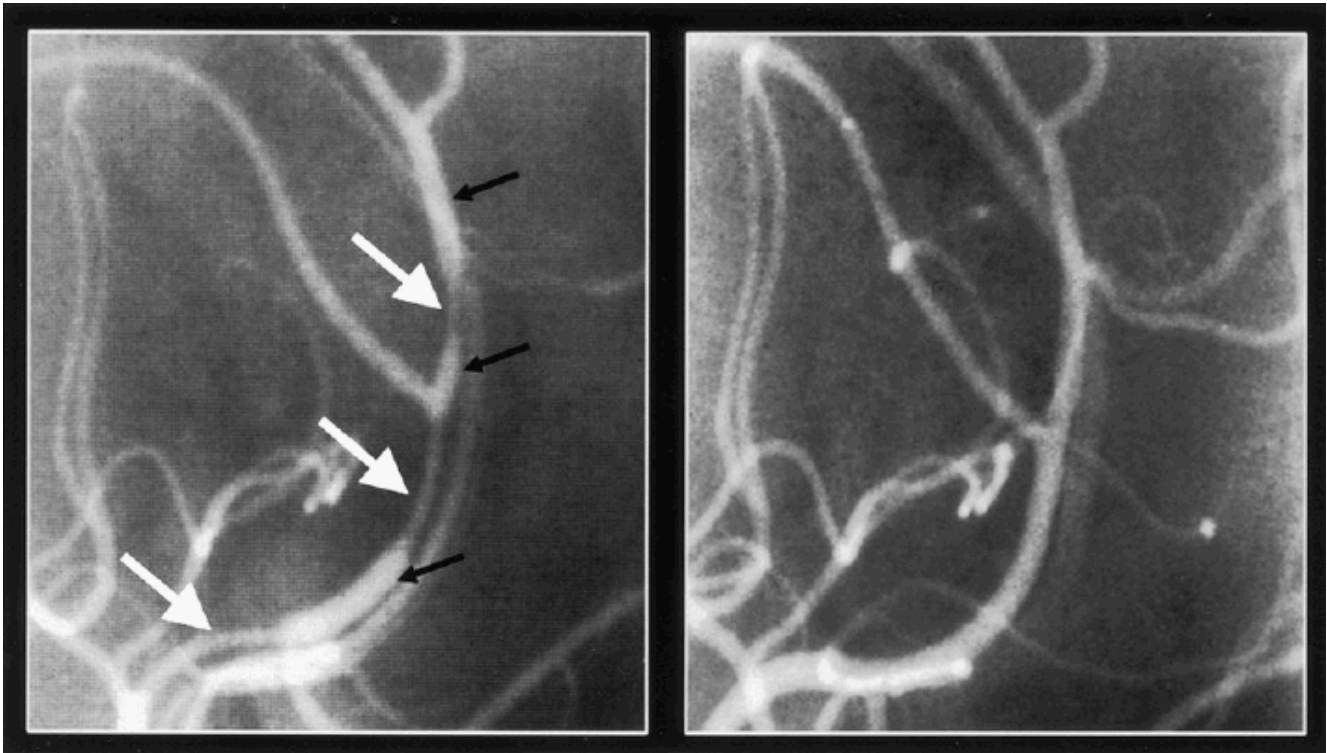


Fig 1

Fig 2

## Conclusion

Thunderclap headache is a very common presentation for acute medical admission. We are aware of excluding subarachnoid haemorrhage first for patients with thunderclap headache. However, clinicians should bear in mind that reversible cerebral vasoconstriction may present with thunderclap headache and this is one of the differential diagnosis despite being rare because its management is different from subarachnoid haemorrhage (SAH).

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