Odd Neurology – An Atypical Thunderclap Headache
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Introduction
As all acute medical physicians are aware, sudden and/or thunderclap headaches are a common presentation to the emergency department (ED). Reversible cerebral vasoconstriction syndrome (RCVS) is an under-diagnosed but emerging cause of such life-threatening presentations. Diagnosis is time-critical as treatment implementation in the acute setting may help optimise patient outcome and prognosis. The following case is a unique presentation encompassing the clinical clues and time-sensitive presentation of RCVS.

Case Study
A 57-year-old Caucasian lady presented with 4 episodes of severe, sudden-onset, generalised headache over a 5-day period, lasting up to an hour each episode. She denied associated symptoms or suffering from headaches in the past. Past medical history included anxiety disorder, vertigo, hypertension and hypothyroidism. Systems and neurological examination were normal. Given the nature of the headache, she was admitted for a CT brain and subsequent lumbar puncture (LP) performed to assess for SAH. Findings were normal, albeit mildly elevated protein was noted on her CSF studies. A few hours post-LP, she complained of non-specific, intermittent right-sided drift with associated weakness, dizziness, and feeling unsteady with poor coordination. She was kept in overnight for observation. Neurology was consulted and a cranial MRI/MRA was performed on Day 2 of admission to exclude an infarct. The imaging showed no acute pathology, flow obstruction, stenosis or aneurysm. She was reassured that there was no organic evidence for cause of her symptoms and queried if it was functional in nature. She was discharged and advised to return to the emergency department if any worsening of symptoms.

Three days later, she represented with complaints of ataxia with secondary falls, persistent headache and feeling ‘muddled’. On clinical examination, she had poor coordination and an anomia aphasia. Cranial MRI/MRA was repeated for evaluation of stroke, which showed multiple foci of cortical infarction in the vascular territories of both right and left parietal and frontal lobes. Other changes noted were multifocal beading of the intracranial vasculature, affecting the anterior and posterior circulation, in keeping with large vessel vasculopathy. These findings were new since previous imaging 4 days prior. An urgent neurological opinion identified the diagnosis as RCVS. She was subsequently transferred to the care of Neurology, admitted to SCU for specialist management.

Neuro-Imaging

![FIG 1. DWI of patient's brain at time of initial presentation (A) showing no evidence of restricted diffusion, in comparison to scan performed 4 days later (B), showing multifocal infarcts involving the right posterior and left medial parietal lobes. On coronal plane MRI (C), shows multifocal ‘beading’ appearance (yellow arrows) of intracranial vasculature. Patient was diagnosed with RCVS and commenced on Nimodipine. There was subtle improvement noted on repeat MRA (D) 8 days later.](image)

Discussion
RCVS is an under-recognised cause of sudden and severe ‘thunderclap’ headaches. Clues to the diagnosis are recurrent thunderclap headaches, often over the course of 1-2 weeks, with or without focal deficits or seizures. Alternative presentations to the ED may include non-aneurysmal SAHs or cryptogenic strokes – especially when the patient also has a headache, and/or is post-partum or after use of vasoactive drugs. These are termed ‘catastrophic RCVS’ as they often carry a high mortality rate at time of presentation. Catastrophic RCVS will have evident findings on neuroimaging given the aetiology of their presentation.

However, as in this case study, complaints maybe non-specific, sub-acute in its presentation with all investigations being normal or negative. Key to diagnosis is in obtaining a full history, as most patients present with complaints of several thunderclap headaches over the course of a few days, and neuroimaging series and neuro-specialist input should be sought when there’s a high clinical suspicion. Multiple angiographies maybe necessary as changes can present two to three weeks after clinical onset of symptoms.

Studies concerning RCVS cases in the setting of emergency departments or headache clinics are being conducted to further understand the epidemiology, pathophysiology and precipitants of the condition.

Treatment and Prognosis
Nimodipine, a calcium-channel blocker, seems to reduce the number and intensity of thunderclap headaches within 48 hours of administration. It doesn’t, however, affect the complications of RCVS, including haemorrhage or ischaemia, or the time course of vasoconstriction.

Additional medications such as anti-convulsants for seizures may need to be used. Patients may need admission to ICU in severe cases.

In some cases, observation and symptomatic management may be all that is required as RCVS is usually a self-limiting process with spontaneous resolution within 3 months.

Long-term prognosis is determinable by occurrence of stroke. Less than 5% develop life-threatening forms with several strokes and uncontrolled massive brain edema.

Outcome
Patient in case study underwent extensive MDT input and demonstrated ongoing impairment in visuospatial and higher level executive functioning domains. She is currently continuing with offsite rehabilitation.

References