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Case Report
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Idiopathic Reversible Cerebral Vasoconstriction Syndrome Complicated by Sub-Arachnoid Bleed

Soorya Hegde[1], Janak Nayak[2], Khadija Rehman[3] Prabhat Sood[1] and Michela Simoni[4]*

[1]Department of General Internal Medicine, County Hospital, United Kingdom

[2]Department of Acute Internal Medicine, County Hospital, United Kingdom

[3]Department of Respiratory Medicine, County Hospital, United Kingdom

[4]Department of Neurology, County Hospital, United Kingdom

Abstract

This report describes a case of Idiopathic reversible cerebral vasoconstriction syndrome with a biphasic clinical course, complicated by / presenting as sub-arachnoid haemorrhage (RCVS-SAH). In this report, we aim to describe the case of a previously healthy 52-year-old woman who presented with severe occipital headache of sudden-onset, where neuroimaging initially revealed a sub-arachnoid bleed and multi-focal, segmental vasoconstriction of cerebral arteries, raising the concern for reversible cerebral vasoconstriction syndrome. Over the course of the next 4 weeks of inpatient stay, the patient developed focal neurological signs and subsequent imagine revealed worsening of the infarct and vasoconstriction. RCVS was diagnosed after ruling out other causes of cerebral vasoconstriction like infections, vasculitis and aneurysmal sub-arachnoid haemorrhage (aSAH). Follow-up with subsequent imaging demonstrated the resolution of changes which were previously demonstrated along with sequelae of RCVS. The patient also reported significant clinical improvement, with only mild persisting weakness of her legs.

Abbreviations

RCVS: Reversible Cerebral Vasoconstriction Syndrome; SAH: Sub-Arachnoid Haemorrhage; CT: Computed Tomography; MRI: Magnetic Resonance Imaging; DSA: Digital Subtraction Angiography

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Correspondence:
Soorya Hegde, Department of General Internal Medicine, County Hospital, Weston Road Stafford, Staffordshire, England, United Kingdom, ST163SA,

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Introduction

Reversible Cerebral Vasoconstriction Syndrome (RCVS) is an entity that was first described by Gregory Call and Marie Fleming as a unique clinical and radiographic syndrome in a small case series of 4 patients presenting with acute headache and reversible cerebral artery vasoconstriction on serial imaging [1-3]. It mostly affects individuals 25–50 years of age, but cases have been reported in other age groups, including children [4,5]. This syndrome is characterised by severe thunderclap headache [6] with occasional neurological signs associated with widespread transient vasoconstriction of the cerebral arteries lasting a few weeks to months. It can occasionally present with various types of intracranial bleed [7]. While there are few known precipitating factors like post-partum period [8], use of alcohol / illicit drugs [9,10] and vasoactive medication [11], some cases are spontaneous [12]. Some of the underlying mechanism believed to play a role in the pathogenesis include sympathetic over activity, inflammation, oxidative stress and disruption of cerebral vascular tone [13]. The diagnosis is confirmed by resolution of changes on repeat imaging after 4-8 weeks.

Case Report

Presentation

A 52-year-old woman with an unremarkable past medical history initially presented to our hospital with a severe, unremitting frontal headache of sudden-onset (thunderclap), associated with neck stiffness, photophobia, and pins/needles sensation over the left arm. She reported no factors changing the intensity of the pain and denied other symptoms like fever, vomiting, visual disturbance, or weakness. She denied the use of medications/illicit drugs apart from Paracetamol for pain relief. General physical examination showed a Blood pressure of 187/57 mm Hg, Pulse rate of 79 bpm and a temperature of 36.7 °C. Neurological examination revealed a GCS of 15/15 with no focal neurological deficit.

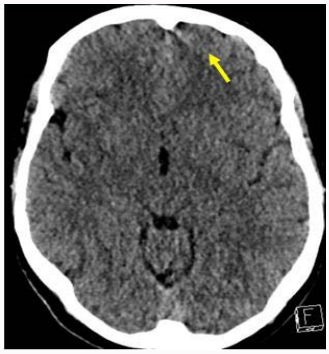

Figure 1: Axial view.

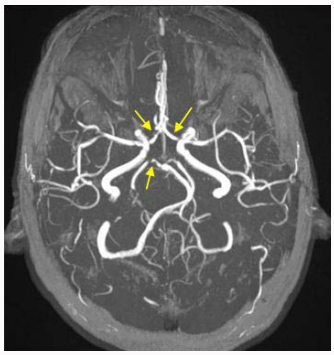

Figure 2: Transverse view.

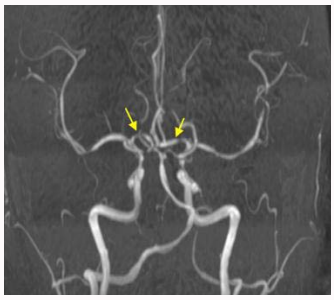

Figure 3: Coronal view.

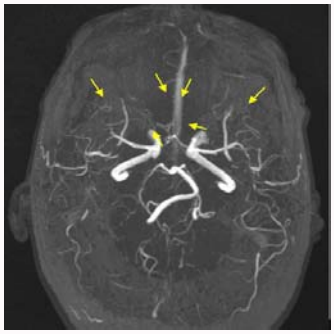

Figure 4: Transverse view.

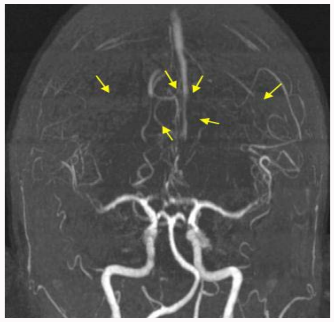

Figure 5: Coronal view.

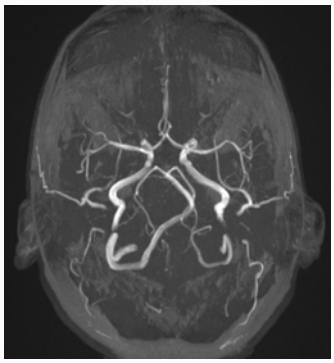

Figure 6: Transverse view.

Initial management

Initial testing including infection markers were within normal limits and a CT scan of the head revealed sub-arachnoid bleed over the left frontal cortex (Figure 1). CT Angiogram done subsequently did not show any evidence of aneurysm. Initial MR Angiogram however demonstrated the beginning of multifocal and segmental intracranial arterial stenosis raising the possibility of RCVS (Figure 2 and 3). The patient was initially managed with anti-hypertensives (Nimodipine) and pain relief.

Clinical course

Over the course of the next few days in the Hospital, the patient developed left-sided weakness, hypotonia and anosognosia. Imaging done in the form of MRI revealed new bilateral frontal and parietal infarcts along with worsening of arterial constrictions/beading as demonstrated before (Figure 4 and 5). This led to confirmation of changes associated with RCVS. Further investigations done to rule out DIFFERENTIALS including infections (in the form of CSF testing), Vasculitis (Blood tests and vessel wall imaging), aneurysmal SAH (imaging) were unremarkable. By exclusion, this was now diagnosed as a case of RCVS-SAH.

Outcome and Follow-up

The patient was discharged with Community physiotherapy and routine follow-up in the Stroke Clinic. Headache and weakness improved over the next 12 weeks and repeat imaging showed resolution of vasoconstriction (Figure 6 and 7). The patient reports being able to mostly manage her activities of daily living with persistence of mild leg weakness, and is still under regular Physiotherapy and Stroke clinic follow-up.

Discussion and Conclusion

RCVS is a rare clinical entity whose mechanisms remain poorly understood. The exact figures for incidence/prevalence are unknown due to lack of large epidemiological studies. It generally affects women more than men [12,14]. It can be Idiopathic or have a known precipitating factor [2,3,8-10]. The most characteristic clinical feature is thunderclap headache which can be exacerbated by change in

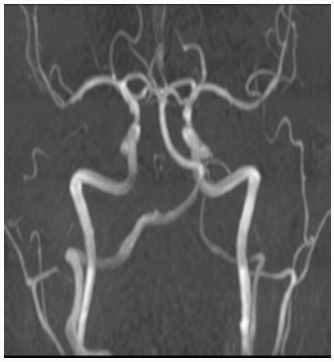

Figure 7: Coronal view.

posture, exercise etc. This can be accompanied by focal neurological signs. It can also present with seizures, intracranial haemorrhage [15,16], cerebral oedema and PRES (Posterior Reversible Encephalopathy Syndrome). Clinical worsening can be seen on rare occasions due to development of infarct/bleed or evolution of the same, as described in this case report. Seri al neuroimaging in the form of CT/MR Angiogram is preferred to visualise the findings characteristic to this syndrome, exclude differentials, and confirm diagnosis and to identify potential sequelae/complications. It is important to exclude potential differentials at the first instance as RCVS is mostly a diagnosis of exclusion, and is retrospective due to the requirement of resolution of changes on follow-up imaging for a confirmatory diagnosis. Digital Subtraction Angiography (DSA) although considered to be the gold standard [17], is not preferred due to the invasive nature of the same and the need for serial imaging for diagnosis as described. The treatment includes withdrawal of any causative agent(s), pain relief, and Blood pressure control. Calcium channel blockers like Nimodipine have been shown to help with symptomatic relief [18]. The long-term prognosis is good in 80-85% of cases with resolution of vasoconstriction expected by around 3 months, while a rarer, more debilitating course ending in death has been described in a minority of patients [19-21].

Important Learning Points

1. Although RCVS is rare and often under diagnosed, an increasing number of cases are being reported due to its associations / differentials and the similar set of investigations for the same.

2. Around 30-50% of all patients with RCVS have convexal SAH (referred to as RCVS-SAH) [22]. It is important to differentiate RCVS-SAH from aneurysmal SAH (aSAH), due to similar clinical (Thunderclap headache) and radiological presentation (arterial narrowing). In around 15% of the patients, no definite cause can be found for the SAH even after imaging. This is termed as Cryptogenic SAH (cSAH).

3. A previous study has shown that patients with RCVS-SAH are mostly women, around 10 years younger, have shorter hospital stays and better overall outcomes than their male counterparts with a SAH or cSAH. It is important to identify the same as they influence the management [22,23].

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