

# Call-Fleming syndrome

Avni Kalangott Padmanabhan Skandhan, Kollengode Gopalakrishnan Ramakrishnan<sup>1</sup>, Rajeev Anand<sup>1</sup>

Department of Radiology, MIMS Kottakkal, Malappuram, <sup>1</sup>Radiologist and HOD, Malabar Institute of Medical Science, Govindapuram, PO Calicut, Kerala, India

**Correspondence:** Dr. Avni K. P. Skandhan, Department of Radiology, MIMS Kottakkal, Malappuram - 676 503, Kerala, India.  
E-mail: avniskandhan@gmail.com

## Abstract

Call–Fleming syndrome is a part of reversible cerebral vasoconstriction syndrome (RCVS) group and is thought to be of idiopathic origin. It is classically described to be having multisegmental, focal vasospasms in the cerebral arteries. It is characterized clinically by the sudden onset of severe headache, classically described as thunderclap headache, with or without associated neurological deficits. The importance of it lies in that it is a potentially reversible cause of this clinical presentation, unlike its other counterparts, aneurysmal subarachnoid hemorrhage (SAH) or vasculitis.

**Key words:** Call–Fleming syndrome; reversible cerebral vasoconstriction syndrome RCVS; vasospasm

## Introduction

Reversible cerebral vasoconstriction syndrome (RCVS) is a recently described term. It is a large group of various clinical entities that includes multiple different entities with a common clinical presentation and radiological appearance. Clinically these patients present with thunderclap headache and may not have neurological deficits. There has to be a strong clinical suspicion of this entity to evaluate it further, as many conditions present with similar kind of clinical findings. The evaluation of RCVS is done by cerebrospinal fluid (CSF) analysis and imaging. Of these, the CSF analysis is most often normal or near normal and the classical radiological appearance is of reversible multifocal and segmental arterial spasms. There may or may not be associated hemorrhage. On follow-up and treatment, these spasms are reversed, thus explaining the name.

## Case Report

A 30-year-old Indian male presented with acute onset of orbito-frontal throbbing headache, which the

patient described as the worst pain in his life. There was no photophobia or nausea associated. The pain was not relieved with positional change or on taking medications (acetaminophen). He had no similar history prior to this. On examination, his vitals were found to be stable. He did not have any neurological deficits.

Magnetic resonance imaging (MRI) brain was done which demonstrated hyperintensity involving right precentral and superior frontal sulci on the fluid-attenuated inversion recovery (FLAIR) axial sequence [Figure 1]. Rest of the cisterns and sylvian fissures were normal. There were no altered parenchymal intensities, areas of restricted diffusion, or parenchymal hemorrhage.

Further, a magnetic resonance angiogram (MRA) was done which showed multiple areas of vasospasm involving the supraclinoid segment of internal carotid artery (ICA), M1 segment of middle cerebral artery, A1 segment of anterior cerebral artery on the right side [Figure 2], and the A1 segment of anterior cerebral artery on the left side. These findings were reconfirmed with cerebral angiogram under digital subtraction angiography (DSA) [Figures 3 and 4].

The laboratory investigations including CSF analysis were found to be inconclusive. Based on the imaging findings, a provisional diagnosis of RCVS was put forth and the patient was put on Nimodipine 30 mg, 3 times daily. The follow-up MRI which was taken 8 weeks later showed complete resolution of the above-mentioned findings [Figure 5]. This confirmed the diagnosis of RCVS.

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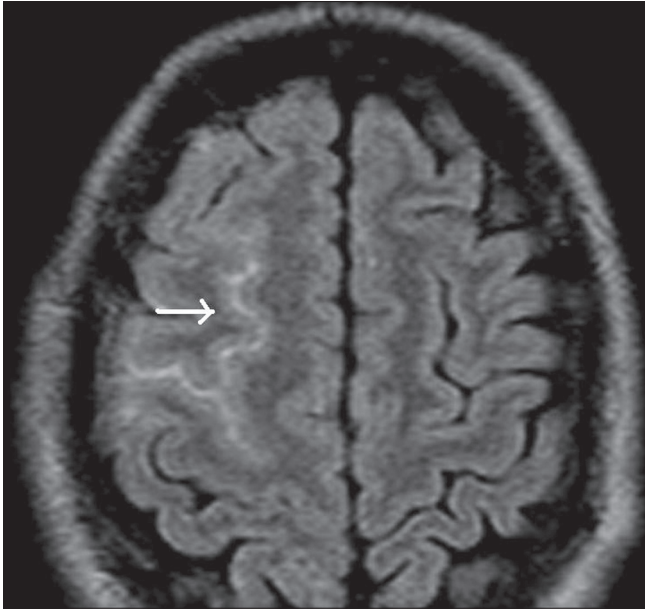
On a detailed history taking and evaluation, the patient was found to have no other co-morbidities and was not on any medications. Hence, the final diagnosis of Call–Fleming syndrome was made.

### Discussion

Our patient presented with bi-fronto-occipital throbbing headache, which was very severe and of sudden onset, and as described by the patient was one of the worst pains the patient ever suffered. This is classically described

as thunderclap headache. The most common cause of thunderclap headache is subarachnoid hemorrhage (SAH), the other causes being cerebral vasculitis, carotid artery dissection, cerebral dural venous thrombosis, and pituitary apoplexy. RCVS is a relatively rare and less-known entity that also presents in a similar manner.

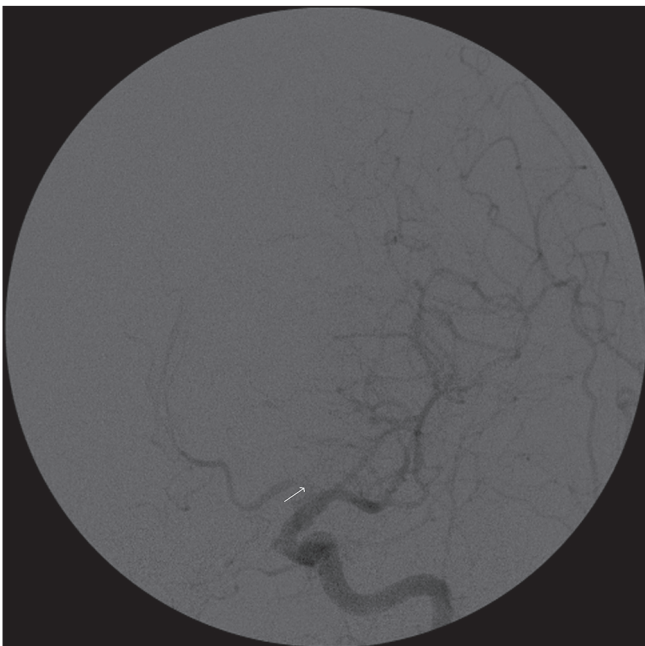
RCVS is a cerebrovascular disorder and it encompasses a large group of conditions which are characterized by, as the name suggests, intracerebral vascular spasms that are reversible. It encompasses the various syndromes like Call–Fleming syndrome, migrainous vasospasm, benign angiopathy of the central nervous system (CNS) and drug-induced arteritis.<sup>[1]</sup> It can also be seen in association with postpartum status, exposure to vasoactive substances, catecholamine secreting tumours, exposure to immunosuppressants, blood products or



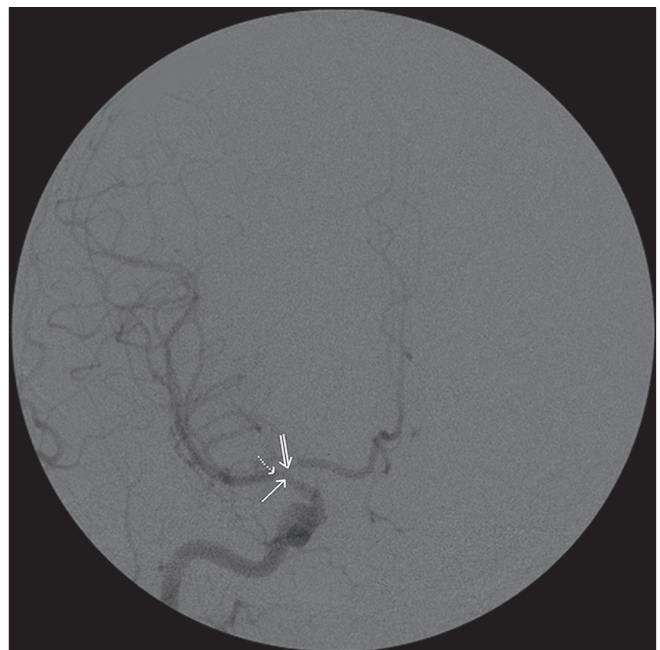
**Figure 1:** FLAIR axial images show focal SAH - blood in the precentral sulci and superior frontal sulci (arrow). The cisterns and sylvian fissures were free



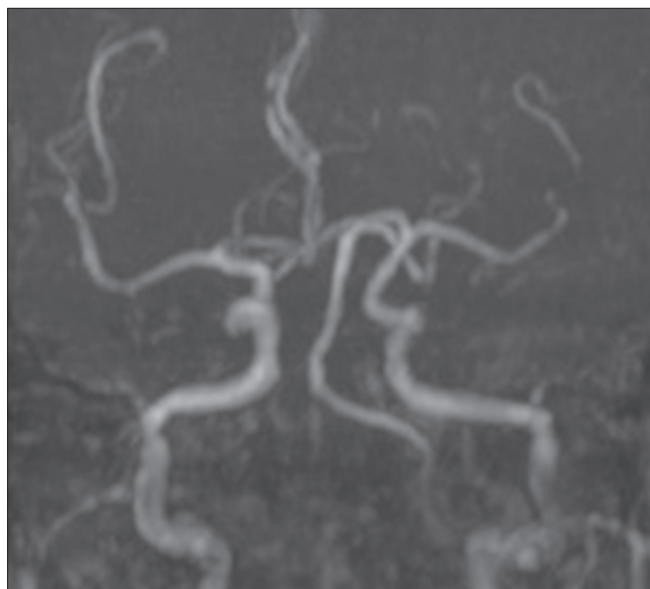
**Figure 2:** MRA TOF images shows vasospasms in right-sided A1 (black arrow) and left-sided A1 (white arrow)



**Figure 3:** Oblique view under DSA shows vasospasm in left A1 (arrow)



**Figure 4:** Frontal view cerebral angiogram under DSA shows vasospasms in right supraclinoid ICA (arrow), M1 (dotted arrow) and A1 (double arrow)



**Figure 5:** Eight weeks post treatment MR Angiogram TOF images appeared normal. Previously mentioned spasms are not visualized in the present scan

blood transfusion, extra- or intracranial large artery disorders, head trauma, neurosurgical procedures, strenuous physical activities, and deep diving in a swimming pool.<sup>[2]</sup> When this clinico-radiological entity is found to be idiopathic and occurs without any predisposing condition, then the condition is classified under Call–Fleming syndrome.

The radiographic appearance is of constriction of the arteries arising or forming the circle of Willis and their branches. The post-stenotic segments show dilatation. This may give an appearance of beaded or sausage-shaped arteries. This appearance with the resolution on the follow-up imaging is classical for this condition.<sup>[3]</sup> Recent articles have shown the association of SAH and intraparenchymal hemorrhage with RCVS,<sup>[4,5]</sup> more commonly subarachnoid haemorrhage which is seen in about 11-25% of the cases. Cortical SAH was the most common pattern seen.

Differentiation from various other similarly presenting conditions is important. The presence of SAH in our case necessitates differentiating vasospasms secondary to aneurysmal rupture from those secondary to RCVS. In RCVS, at the time of presentation, the patient has the multifocal vasospasms which are characteristic of the same. However, in cases of aneurysmal rupture with SAH, there was delayed vasospasm in a time setting of about 1-2 weeks after hemorrhage.<sup>[6]</sup> Ruptured aneurysmal SAH has various patterns depending on the region of involvement. An anterior communicating artery aneurysm is suggested by blood in the cisterna lamina terminalis, anterior pericallosal cistern, and interhemispheric fissure. In posterior communicating

artery aneurysms, blood is usually seen diffusely within the cisterns. Rupture of a middle cerebral aneurysm is characterized by blood in the sylvian fissure and a hematoma in the temporal lobe, which may rupture into the adjacent temporal horn. Posterior fossa aneurysms mostly have no characteristic localizing findings on computed tomography (CT) scan. In our case, the blood was found in the cortical sulci and there was no evidence of blood in the basal cisterns. Vasospasms can also occur in correlation to a ruptured aneurysm; these spasms are most commonly seen in the vessels closest to the site of leakage. Traumatic SAH also present similarly; however, our case had no history of trauma.

Primary angitis of the CNS has no characteristic radiological findings; it can have diffuse white matter lesions, multiple infarctions involving multiple vascular territories, and multiple intraparenchymal hemorrhages. The differentiation from RCVS is predominantly based on the clinical presentation. Primary angitis of the CNS tends to have a dull aching pain of insidious onset and is progressive in nature. Our case had a sudden and severe onset of headache, with MRI showing no parenchymal changes.

Therapeutic management is by calcium channel blockers;<sup>[7]</sup> however, they should be used with caution as there is a risk of infarction in the watershed regions.<sup>[1]</sup> Short course of high-dose glucocorticoid and magnesium sulfate as treatment are also advocated. Due to spontaneous resolution, the prognosis is good.<sup>[1]</sup> Identification and discontinuation of the trigger agents is advocated.<sup>[8]</sup>

As discussed earlier, the changes are reversible, with nil or minimal neurological defects. During the course, however, secondary to the vasospasms, stroke could occur and this is an important determinant of the morbidity of the patient.<sup>[1]</sup>

## Conclusion

Call–Fleming syndrome is an idiopathic cause for RCVS – a clinico-radiological entity. Thunderclap headache is one of its chief clinical presentations; however, it may have a spectrum of other findings too. The classical radiological presentation is that of multisegmental reversible vasoconstriction, followed by its resolution in a span of 3 months. However, a strong clinical suspicion is mandatory to evaluate the patient further and to reach the diagnosis.

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