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Case Series

Radiology Section

Imaging of Postpartum Reversible Cerebral Vasoconstriction Syndrome- A Case Series

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ABSTRACT

Postpartum Reversible Cerebral Vasoconstriction Syndrome (RCVS), previously known as postpartum angiopathy, presents within four weeks of delivery. Recurrent episodes of severe headache, seizures, focal neurologic deficits and photophobia are the common presentations of RCVS. Hereby the authors report a series of patients (30-year-old, 26-year-old and 29-year-old females) who presented with neurologic symptoms ranging from severe headache to altered sensorium and seizures. Imaging revealed a multitude of findings, from normal-appearing parenchyma to lobar and subarachnoid haemorrhage. Computed Tomography (CT) and Magnetic Resonance Angiographic (MRA) studies showed multiple areas of vasoconstriction and beaded appearance of the cerebral arteries in all three patients, which completely resolved within 12 weeks of the onset of symptoms, suggesting RCVS. Imaging in the acute setting plays a pivotal role in identifying the vasoconstriction and ruling out other neurological emergencies in the postpartum period.

Keywords: Angiopathy, Arterial beading, Puerperium

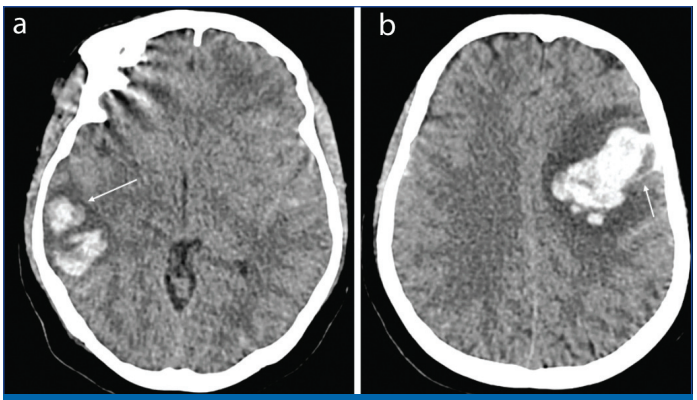
INTRODUCTION

Postpartum RCVS occurs in normotensive females within four weeks of delivery [1]. It was previously called by various names, including Call-Fleming syndrome, postpartum angiopathy and postpartum angiitis [2]. RCVS commonly presents with sudden onset of severe headache, focal neurologic deficits, seizures, or photophobia. Dysregulation of cerebral vascular tone from sympathetic overactivity, endothelial dysfunction and oxidative stress are attributed to the pathogenesis of RCVS [3]. Complications of RCVS include intraparenchymal or subarachnoid haemorrhage and infarction. Angiographic studies show multifocal short-segment narrowing in the cerebral arteries, which resolve spontaneously within 12 weeks [4]. Early recognition of this entity and appropriate treatment reduces maternal morbidity and mortality [2].

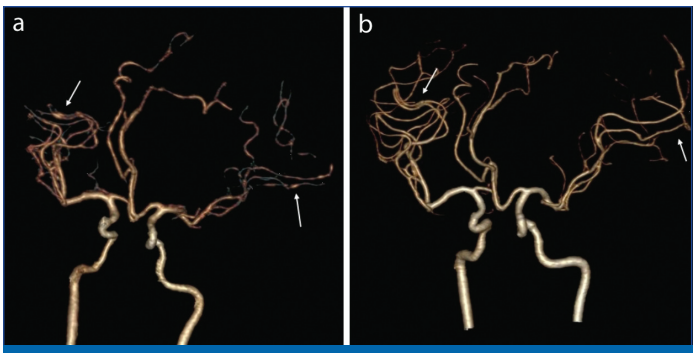
CASE SERIES

Case 1

A 30-year-old female, gravida 2, para 2, presented with altered sensorium, one episode of generalised tonic-clonic seizures lasting for two minutes, and right-sided weakness within 24 hours after normal vaginal delivery. Her antenatal check-ups were unremarkable, and there was no history of seizure disorder, preeclampsia, or gestational diabetes mellitus in her previous and present pregnancy. She was not a smoker or an alcoholic. On examination, her vitals were stable, and Glasgow Coma Scale (GCS) score was 15/15. Patient was aphasic, with right upper and lower limb strength rated at 2/5 and 3/5, respectively. A plain CT scan showed left frontal and right temporal intraparenchymal haemorrhages and thin subarachnoid haemorrhage in the adjacent sulcal spaces [Table/Fig-1a,b]. A Computed Tomography Angiogram (CTA) of the intracranial vessels showed multifocal narrowing of the cortical branches of the bilateral Anterior Cerebral Arteries (ACA) and Middle Cerebral Arteries (MCA) [Table/Fig-2a]. The patient was kept under strict neurological observation and started on 60 mg nimodipine every four hours through nasogastric tube, along with intravenous magnesium sulphate (2 g every 6 hours) for four days, followed by oral nimodipine for four weeks. A repeat CTA performed at 12 weeks showed complete resolution of the arterial narrowing [Table/Fig-2b]. Her right upper and lower limb muscle power gradually improved with physiotherapy over a period of six months.



[Table/Fig-1]: Plain CT image (a) Showing an irregular parenchymal haemorrhage in the right temporal lobe (white arrow); (b) Showing parenchymal and thin subarachnoid haemorrhages in the left frontal region.

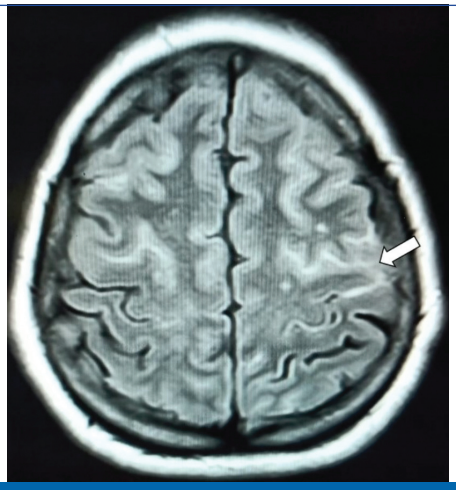


[Table/Fig-2]: Volume rendered CTA images (a) Showing multiple areas of luminal narrowing with beaded appearance in the distal cortical branches of bilateral MCA and ACA (white arrows). Repeat CTA volume rendered image; (b) Showing complete resolution of the beaded appearance of the arteries (white arrows).

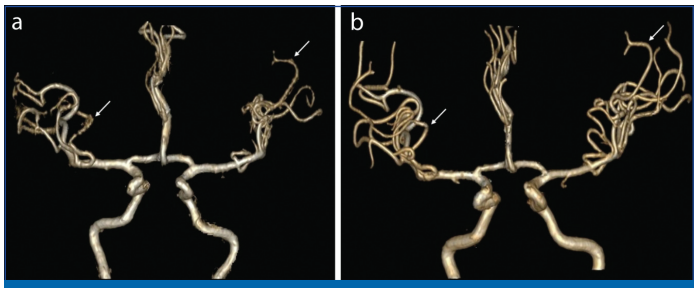
Case 2

A 26-year-old primiparous woman presented with severe thunderclap headache, vomiting, and altered behaviour three days after an emergency Lower Segment Caesarean Section (LSCS) for pregnancy-induced hypertension. She had no history of migraine or diabetes mellitus. On examination, her vital signs were normal, with GCS of 15/15. Complete haemogram, C-reactive protein, liver function tests and urine examination were normal. Magnetic Resonance Imaging (MRI) showed Fluid-attenuated Inversion Recovery (FLAIR) hyperintensities with blooming on susceptibility-weighted images in

the bilateral frontal and parietal sulci, features consistent with acute subarachnoid haemorrhage [Table/Fig-3]. MRA showed multifocal areas of short segment mild luminal beading in the M3 and M4 segments of bilateral Middle Cerebral Artery (MCA) [Table/Fig-4a]. The patient was given levetiracetam 500 mg and nimodipine 60 mg every four hourly for six weeks. A repeat MRA performed after eight weeks showed complete resolution of the bilateral arterial narrowing [Table/Fig-4b].



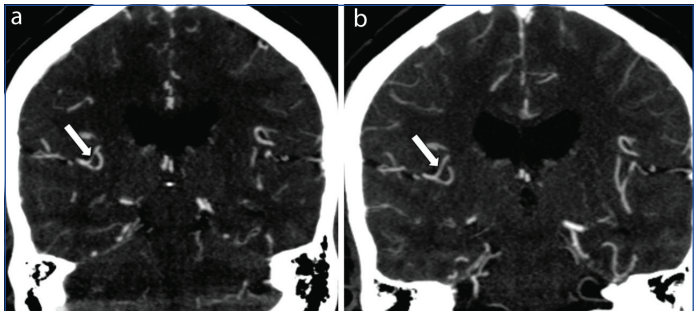
[Table/Fig-3]: Axial FLAIR MR image showing sulcal hyperintensity in the bilateral frontal sulci (white arrows).



[Table/Fig-4]: Volume rendered MRA images (a) Showing multiple areas of beading in the distal cortical branches of bilateral MCA and ACA (white arrows). Repeat MRA volume rendered image; (b) Showing complete resolution of the beaded appearance of the arteries (white arrows).

Case 3

A 29-year-old female, gravida 2, para 2, presented with recurrent episodes of what she described as the severe headache of her life one week after normal vaginal delivery, each episode lasting 40-50 minutes. Her antenatal period was unremarkable. On examination, her blood pressure was 116/76 mmHg, pulse rate 76 beats per minute, oxygen saturation 99% and GCS 15/15. No history of migraine, preeclampsia, or diabetes mellitus. The plain CT of the brain was unremarkable. CTA showed multifocal short-segment luminal narrowing involving the bilateral MCA branches [Table/Fig-5a]. The patient was conservatively treated with nimodipine 60 mg every four hours for six weeks. A repeat CTA after ten weeks showed complete resolution of the findings [Table/Fig-5b].



[Table/Fig-5]: Coronal CTA images (a) Showing luminal narrowing with beaded appearance at two sites in the cortical branches of right MCA (white arrows). Repeat CTA coronal image; (b) Showing complete resolution of the beaded appearance of the arteries (white arrows).

DISCUSSION

The RCVS represents a group of disorders characterised by severe headache with or without other acute neurological symptoms, and the presence of reversible luminal narrowing of the cerebral arteries [4]. This unifying term was proposed to include various syndromes with similar clinical presentation and radiological findings. Transient dysfunction of the cerebral autoregulation, along with sympathetic hyperactivity in response to endothelial injury, has been implicated in the pathogenesis of this condition [5]. An increase in anti-angiogenic factors during puerperium, in an already altered cerebral arterial tone secondary to pregnancy-associated hormones, results in postpartum RCVS [6].

Numerous vasoactive agents, like serotonergic and adrenergic drugs, cannabis and cocaine abuse, and catecholamine-producing tumours, were identified as putative precipitants for this condition [7,8]. Postpartum RCVS classically presents in normotensive women within four weeks after an uneventful pregnancy. A rapid fall in the hormones, particularly progesterone, is believed to precipitate this syndrome [9]. About one-third of the patients with postpartum RCVS also had exposure to bromocriptine to inhibit lactation or vasoconstrictors for epidural anaesthesia and postpartum haemorrhage [8]. Acute onset of thunderclap headache is the most common and often the only manifestation of postpartum RCVS. Patients usually experience multiple episodes of severe headache, with each episode lasting one to three hours [7]. Two of our cases had severe headache as the presenting symptom, former presented with persistent thunderclap headache, while the latter with multiple episodes of headache lasting 40-50 minutes. Other common neurological manifestations include seizures, vomiting and photophobia. RCVS is usually self-limiting, and the symptoms resolve within two to three months in most patients. Transient or persistent focal neurological deficits are seen in 8-43% of the patients [3]. Occasionally, RCVS is complicated by the development of ischaemic stroke and intraparenchymal or subarachnoid haemorrhage [10]. In the appropriate clinical context of a postpartum female with severe headache, RCVS may be a straightforward diagnosis; however, imaging plays a vital role in the documentation of reversibility of vasospasm and to exclude the mimics of RCVS.

In uncomplicated RCVS, plain CT and MRI are usually unremarkable. Thin convexity Subarachnoid Haemorrhage (SAH) is seen in 10-20% of cases [11]. Intraparenchymal haemorrhage can occur, which may be single or multiple. Infarctions typically occur in the arterial watershed areas and can rarely have territorial distribution [12]. Digital Subtraction Angiography (DSA), though not routinely indicated, remains the gold standard for identifying vasoconstriction and has a very high sensitivity in detecting the changes in the distal cortical branches. Owing to its invasive nature and procedure-related complications, DSA is reserved for the patients when non invasive angiographic study (CT/MRA) do not reveal any abnormalities [12]. CT and MR angiogram have been demonstrated to be 80% sensitive in detecting the vasoconstriction of RCVS, compared to DSA [8]. The pathognomonic imaging feature of RCVS is segmental narrowing and dilatation (resembling a string of beads) in one or more cerebral arteries. Both the anterior and posterior circulation arteries can be affected [5]. Complete or atleast substantial resolution of these findings is observed on repeat angiogram performed 12 weeks after the onset of symptoms. The first case presented with intraparenchymal haemorrhage, the second case with SAH and the third case with no finding on plain CT. In all three cases, CT/MRA typically showed multifocal involvement of the medium-sized arteries (branches of ACA and MCA). On follow-up imaging at 8-12 weeks, all three cases showed complete resolution of the segmental narrowing.

Aneurysmal SAH is the most common cause of severe thunderclap headache, and its imaging features overlap with that of RCVS. Recurrent episodes of severe headache, classically seen in RCVS,

are unusual in aneurysmal SAH. In aneurysmal SAH, imaging often identifies the aneurysm in the circle of Willis, and the associated vasospasm involves a long segment of proximal arteries. RCVS shows diffuse beaded appearance, preferentially affecting the second and third-order cerebral branches [12]. Posterior Reversible Encephalopathy Syndrome (PRES) is a closely related condition that can clinically mimic RCVS. Both these syndromes are believed to be secondary to cerebral autoregulation dysfunction. On imaging, PRES shows vasogenic oedema in the cortical and subcortical regions, predominantly in the parieto-occipital lobes [7]. Other conditions, like cerebral venous thrombosis and pituitary apoplexy, can present with severe headache in postpartum women, but both have distinctive imaging features [2,9]. RCVS needs to be differentiated from Primary Angiitis of the Central Nervous System (PACNS), as both share similar clinical and imaging features. PACNS presents with insidious onset and slowly progressive headache, in contrast to the thunderclap headache of RCVS. CSF analysis shows elevated protein levels and white blood cell count in PACNS [13]. Both PACNS and RCVS show multifocal beading of the arteries. The arterial narrowing in RCVS shows improvement with intra-arterial vasodilators and complete resolution within 12 weeks. In contrast, the arterial narrowing of PACNS shows no improvement with vasodilators and no significant change on follow-up imaging [12].

The management of RCVS is primarily symptomatic, with analgesics and rest; antiepileptics are added in patients with seizures [9]. Precipitants, like vasoactive drugs or bromocriptine, should be discontinued. Calcium channel blockers, like nimodipine, have been used to relieve vasoconstriction. Nimodipine is administered orally or by nasogastric tube at a dosage of 30-60 mg every 4 hours, and it should be tapered over 4-8 weeks [14,15]. Intra-arterial nimodipine and balloon angioplasty have been tried in severe and refractory cases [7,10].

CONCLUSION(S)

Postpartum RCVS presents with a multitude of clinical and imaging manifestations which overlap with various other conditions, particularly PRES and PACNS. Early recognition of this entity by CT or MRA is the linchpin for initiating the appropriate treatment. Documentation of reversibility of the cerebral arterial narrowing by 12 weeks confirms the diagnosis of RCVS.

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