Reversible cerebral vasoconstriction syndrome in the backdrop of cerebral venous thrombosis: A post-partum catastrophe

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ABSTRACT

Reversible cerebral vasoconstriction syndrome (RCVS) is an under-recognized condition characterized by the association of acute severe headaches, often of the thunderclap type, with or without additional neurological symptoms, and multifocal constriction of cerebral arteries, that resolve spontaneously within 1–3 months. Here, we report a rare case of post-partum stroke, where cerebral venous thrombosis was followed in quick succession by RCVS. A 24-year-old lady in her immediate post-partum period developed headache, seizure, and papilledema. After a subsequent episode of a severe headache, she had right hemiparesis and dysarthria. Computed tomography and magnetic resonance venography confirmed the presence of thrombosis. Afterward, she had another episode of a severe headache with altered sensorium. MR imaging of the brain revealed widespread infarct in bilateral medial frontal areas, and MR angiography was suggestive of segmental constriction of cerebral arteries bilaterally. The subsequent clinical course was uneventful with slow recovery. Repeat brain imaging, after 4 weeks, revealed near-total disappearance of diffusion restriction and subsidence of the beading pattern in the cerebral vasculature. This rare case depicts the co-occurrence of venous and arterial angiopathies in the setting of a post-partum headache with the stroke that led to a challenging clinical scenario.

Keywords: *Cerebral venous thrombosis, Headache, Reversible cerebral vasoconstriction syndrome*

Reversible cerebral vasoconstriction syndrome (RCVS) is an under-recognized condition characterized by the association of acute severe headaches, often of the thunderclap type, with or without additional neurological symptoms, and multifocal constriction of cerebral arteries, that resolve spontaneously within 1–3 months [1-3]. More than half of the cases of RCVS occur in special circumstances, such as exposure to post-partum or vasoactive substances. The pathology of this peculiar cerebral angiopathy is still obscure [4].

Cerebral venous thrombosis (CVT) is an infrequent disorder characterized by a wide clinical spectrum, which includes a headache in the vast majority of cases [5]. No obvious physiological link has so far been established between these two forms of angiopathy. We report a case of post-partum stroke where CVT was followed in quick succession by RCVS, representing a rare clinical situation.

CASE REPORT

A 24-year-old female in her immediate post-partum period developed holocranial headache associated with vomiting that started 3 days after the delivery. She was admitted in a local health-care hospital following 1 attack of generalized tonic-clonic convulsion which occurred on the day 8 of postpartum. Neurological examination revealed bilateral papilledema and no focal neurologic deficit. On the next day, she had a second bout of a severe headache followed by the right-sided complete hemiparesis along with dysarthria. She was subsequently referred to the higher center and was admitted under our care.

When the patient reported to our department, her clinical examination was done and the vitals were within the normal ranges.

Computed tomography scan showed hemorrhage in the left caudate nucleus along with an intraventricular extension of bleed (Fig. 1). Magnetic resonance venography (MRV) confirmed the presence of thrombosis involving superior sagittal sinus and the left transverse sinus (Fig. 2). Routine blood parameters revealed anemia (hemoglobin 9.8 g/dl) without any other abnormality. All blood tests for thrombophilic conditions were negative or normal, including antinuclear antibodies, anti-DNA antibodies, antiphospholipid antibodies, C protein, S protein, antithrombin-3 and genetic screening for mutation G1691A of factor V, mutation G20210A of factor II, mutation V617F of JAK2, and mutation of MTHFR. Anticoagulation was initiated, thereafter, with low molecular weight heparin (enoxaparin).

On the 4th day after admission, she developed an exacerbation of a headache with altered sensorium. After regaining consciousness, she was found to have developed the akinetic mute state. Spasticity was present in both the lower limbs along with brisk deep tendon reflexes in all four limbs and bilateral extensor

plantar response. MR imaging of the brain, at this point, revealed widespread diffusion restriction and infarct, mainly in the bilateral medial frontal (parasagittal) region (Fig. 3). MR angiography was suggestive of segmental constriction and dilatation of cerebral arteries on both the sides (Fig. 4). A final diagnosis of post-partum stroke was made, where CVT was followed in quick succession by RCVS.

The subsequent clinical course was uneventful with slow recovery. She was started on nimodipine, and anticoagulation was withdrawn. Repeat brain imaging, after 4 weeks, revealed near-total disappearance of diffusion restriction and subsidence of the beading pattern in the cerebral vasculature (Fig. 5).

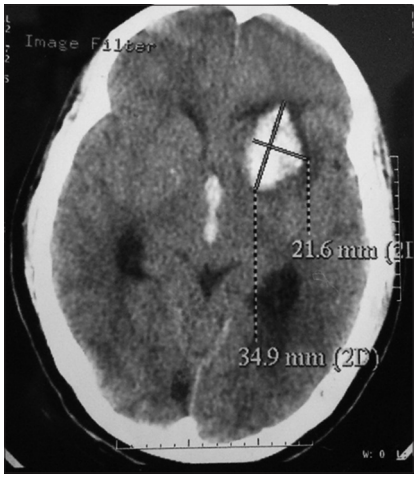

Figure 1: Computed tomography brain shows hemorrhage in the left caudate nucleus

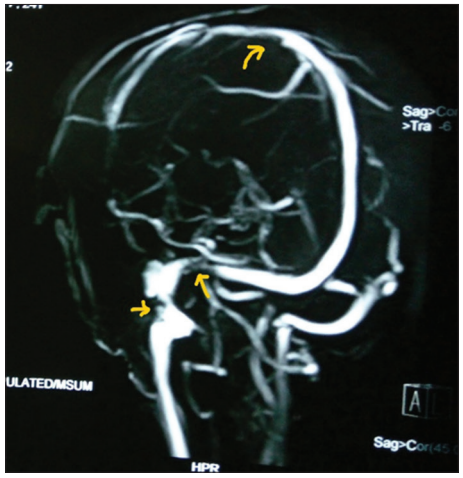

Figure 2: Magnetic resonance venography revealed thrombosis (yellow arrows) involving superior sagittal sinus along with left transverse sinus and left sigmoid sinus

DISCUSSION

A cerebral venous thrombosis is an uncommon form of stroke with a wide range of clinical manifestations. The risk factors are pregnancy, intravenous drug abuse, infection, and dehydration. 90% of cases have a headache as the main symptom. Seizures are fairly common in CVT. Papilledema is found in around 30% of cases and is attributed to raised intracranial tension. Focal neurologic deficits do occur in CVT as consequence of infarcts and less commonly, due to hemorrhages. Deep gray matter hemorrhages mostly occur in thrombosis of straight sinus and vein of Galen [6].

RCVS is a rare form of angiopathy and has been mostly reported in the background of childbirth. Segmental arterial spasm is the basic mechanism which gives rise to an excruciating headache. Infarcts and hemorrhages both have been reported in the context of RCVS. However, hemorrhage in RCVS precedes infarct. Basal ganglia hemorrhage can frequently occur in RCVS, resulting from a spasm in the proximal part of arterial circulation [7].

Our patient developed diffuse holocranial headache of increasing intensity in the immediate post-partum period, which was followed by an appearance of generalized seizure. The subsequent detection of bilateral papilledema was suggestive of a rise in intracranial pressure in this case. The constellation, the above-mentioned clinical features, fits well with CVT which was later confirmed by MRV. However, the second bout of a

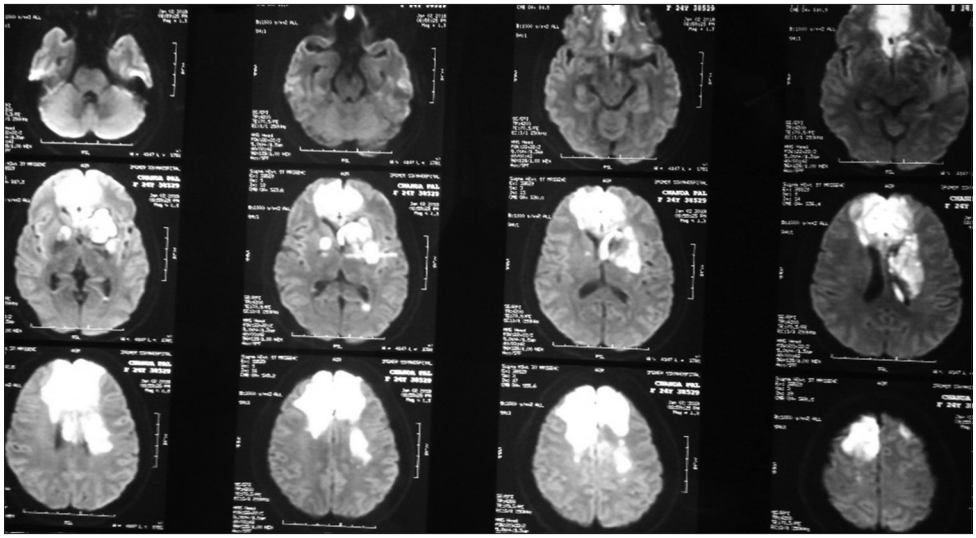

Figure 3: Magnetic resonance imaging of the brain, after the third episode of headache, revealed widespread diffusion restriction, mainly in bilateral medial frontal (parasagittal) region

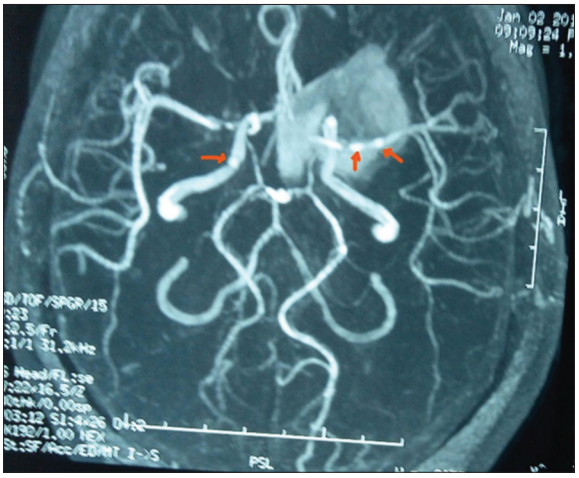

Figure 4: Magnetic resonance angiography was suggestive of segmental constriction and dilatation of cerebral arteries on both the sides (red arrows)

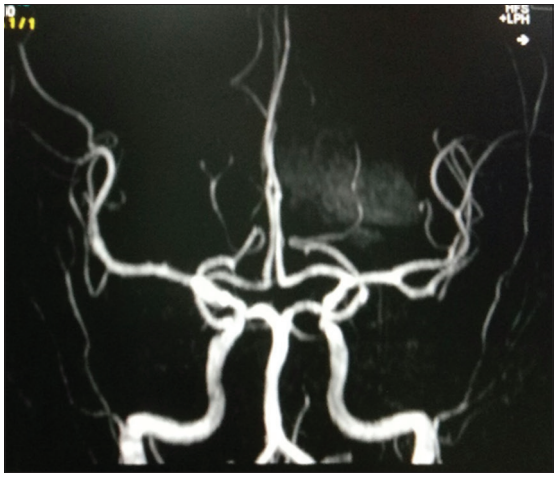

Figure 5: Repeat brain imaging, after 4 weeks, revealed near-total disappearance of the beading pattern in cerebral vasculature

severe headache with the left basal ganglia bleed and the right hemiparesis cannot be explained by MRV finding of superior sagittal sinus thrombosis alone. Rather, taking into account the subsequent development of widespread infarct in both frontal lobes along with obvious beading pattern in multiple cerebral arteries on MRA, one has to consider the possibility of RCVS. The resolution of diffusion restriction and vascular segmental constriction in follow-up MRI brain after 2 months further supported the idea of RCVS. We speculate that our patient developed RCVS at the point of the second bout of a headache. The left caudate bleed was an early hemorrhagic manifestation of RCVS in this case.

There are two likely possibilities in the present case: RCVS and CVT were co-existent or CVT gave way to RCVS. The association between CVT and RCVS has been previously reported in two women during immediate postpartum [8,9]. In both the cases, the authors concluded that CVT and RCVS were probably distinct pathophysiologically, although Katzin *et al*. hypothesized that rapid changes in CSF pressure due to CVT may be mechanistically involved in triggering RCVS [9]. Recently, another such case showing an association between these two forms of angiopathy has been described in a young lady who had a stent in her lateral venous sinus for the management of idiopathic intracranial hypertension [10].

CONCLUSION

This case depicts the co-occurrence of venous and arterial angiopathies in the setting of post-partum headache with a stroke that led to a challenging clinical scenario.

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