Is cervicocephalic dissection a part "postpartum vasculopathy" spectrum? A case of postpartum posterior reversible encephalopathy syndrome, bilateral petrous carotid artery dissections, and stroke

Sir,

The postpartum dissection of large and medium sized arteries is a rare entity, being increasingly recognized due to the widespread availability of non-invasive imaging. Coronary arteries and cervicocephalic arteries (carotid and vertebral arteries) are commonly involved. Although single vessel dissection is reported, involvement of two or more arteries is not uncommon. Involvement of both the coronary and cervicocephalic arteries in the same patient at the same time possibly represents a unique predisposition shared between these two arteries. The association of postpartum posterior reversible encephalopathy syndrome (PRES) with cervicocephalic dissection is rare. We report the fourth case of cervicocephalic artery dissection associated with PRES.

A 29-year-old female patient had severe pregnancy-induced hypertension with albuminuria underwent preterm lower segment caesarean section (LSCS) at 31 weeks of pregnancy. She developed multiple seizures during the immediate postpartum period. She underwent magnetic resonance imaging (MRI) at a local hospital, which showed multiple ill-defined T2 hyperintensities in bilateral gangliocapsular regions (L>R) and in bilateral cerebral hemisphere white matter [Figure 1]. The MRI pattern was suggestive of PRES, for which she was treated conservatively in the local hospital. During the 14th postpartum day, she developed sudden onset left hemiplegia with reduced consciousness, suggestive of right middle cerebral artery (MCA) territory stroke. She was referred to our hospital, and was evaluated with a repeat MRI. MRI revealed a right MCA territory large, acute infarct [Figures 2 a-e]. There was minimal residual fluid-attenuated inversion recovery hyperintensity in the left caudate nucleus [Figure 2a arrow], corresponding to the previous findings of PRES, and complete resolution of rest of the gangliocapsular region and white matter hyperintensities. Time-of-flight (TOF) MR angiogram (MRA) showed occlusion of the right internal carotid artery (ICA) and MCA[Figure 2f]. Mild short segment stenosis (<50%) was seen at the origin of the left anterosuperior division of MCA. Double lumen was noted in the left distal petrous ICA [Figure 2f, arrow] with a dissection flap in-between. Axial T1-weighted image showed a hyperintensity within the false lumen of the dissected segment of left petrous ICA [Figure 2g, arrow] with reduced calibre of normal flow void. Time of flight (TOF) MRA source images [Figure 2h] showed a T1 hyperintense signal in the right petrous ICA wall (long arrow).

Her vasculitis workup (antinuclear antibody, Anti-double stranded DNA antibody, phospholipid antibody-IgG and IgM) was negative. Serum electrolytes, liver function tests, renal function tests, random glucose test, bleeding parameters, and the platelet count were normal. Her total count was elevated, with predominant neutrophilia. Her erythrocyte sedimentation rate (ESR) was 77 mm/h. Human immunodeficiency virus, hepatitis B surface antigen, and venereal disease research laboratory tests were negative.

The postpartum vasculopathy represents a group of disorders consisting of pre-eclampsia/eclampsia, PRES, and reversible cerebral vasoconstriction syndrome (RCVS). Postpartum coronary and cervicocephalic dissection also appear to be a part of the spectrum of postpartum angiopathy/vasculopathy.[1] The coexistence of PRES/RCVS with cervicocephalic dissections favors this hypothesis. While multiple gene mutations are known to predispose to large artery (aorta) dissections,[2] apart from collagen vascular disorders such as Ehlers–Danlos and Marfan syndrome, no gene mutations predisposing to coronary cervicocephalic artery dissections are identified. As

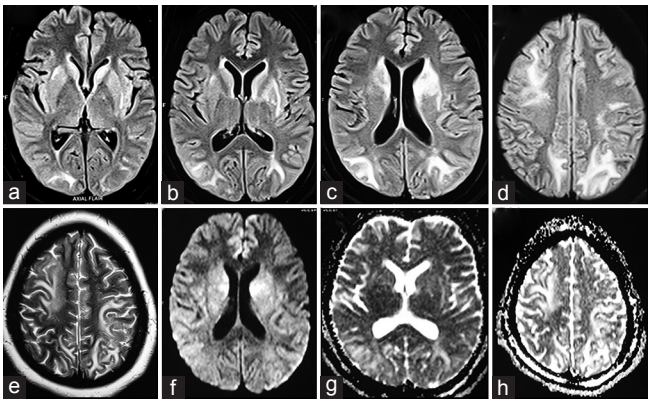

Figure 1: Immediate postpartum MRI. (a-d) Axial FLAIR MRI of the brain showing multiple hyperintensities in bilateral gangliocapsular regions (L>R) and white matter. (e) T2-weighted image showing white matter hyperintensities. (f) DWI and (g,h) apparent diffusion coefficient (ADC) images showing diffusion facilitation

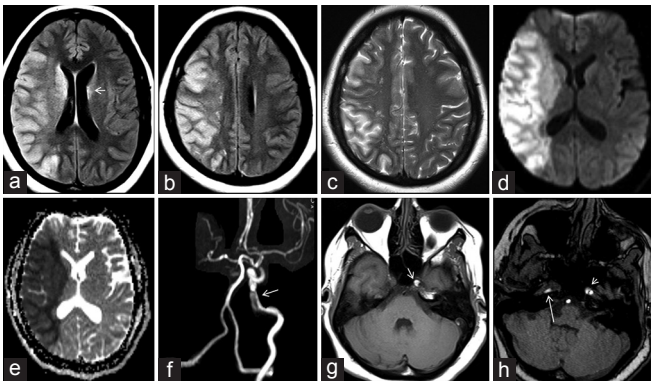

Figure 2: MRI after stroke onset. (a, b) Axial FLAIR and (c) Axial T2-weighted MRI showing residual left caudate nucleus hyperintensity (arrow) and resolution of rest of the previously noted abnormalities. (d) DWI and (e) ADC images showing a large right MCA territory acute infarct. (f) TOF MR angiogram showing occlusion of the right ICA and MCA, and the dissection flap in the left petrous ICA (arrow). (g) Axial T1-weighted image showing a hyperintense blood signal in the left petrous ICA wall (arrow). (h) Source images of TOF MRA showing the hyperintense intramural blood in the right petrous ICA wall (long arrow), and a double lumen in the left petrous ICA (short arrow)

possible genetic predisposition is more likely in postpartum vasculopathy, genetic studies in such cases are required for better understanding of the pathophysiology and treatment. A single case report of a four vessel cervicovertebral artery dissection in association with haemolysis with elevated liver enzymes and low platelet count (HELLP; hemolysis, elevated liver enzymes, low platelet count) syndrome has been described,[3] where a possible inadequate immune response was hypothesised as the possible predisposing cause. However, this theory was not supported by other case reports or case series. The head and neck manipulation as an etiology for the occurrence of arterial dissection is unlikely in our case because the dissections were in the petrous ICA segment, which is the most immovable portion of ICA.

There is no predisposed segment of vertebrobasilar and carotid arteries that undergoes frequent dissections. In our case, bilateral petrous ICA segment was involved, whereas in the literature, involvement of different segments of the intra- and extracranial portions have been described. A few cases associated with reversible cerebral vasoconstriction syndrome have also been reported.[4,5] An association between PRES and cervicocephalic artery dissection is extremely rare, with only three cases being reported in the literature till date.[4,6] We report the fourth case of cervicocephalic artery dissection following PRES.

In our case, the patient had eclampsia prior to delivery, developed PRES during the immediate postpartum period, as well as bilateral carotid artery dissections. The carotid wall intramural haemorrhage (dissection) was showing T1/T2 hyperintensity, suggestive of late subacute stage of the haemorrhage. It is likely that both dissection and PRES would have started at the same time, even though stroke manifested clinically 2 weeks later. In most of the previously reported cases as well as in our case, the onset of dissection and PRES appears to be simultaneous; however, Burrus *et al*.,[7] attributed dissection as a predisposing factor for PRES due to failure of the baroreceptor reflex. Elevated ESR may be due to the postpartum status or secondary to unknown, subtle, vascular inflammatory processes not detectable during the routine vasculitis workup. Although MRI was done during the immediate postpartum period, angiogram was not done at that time. Avoiding a large stroke could have been possible if the dissection had been identified at an earlier stage. Blood pressure management in such cases, where the two entities (PRES and cervicocephalic dissection) coexist, is tricky because aggressive blood pressure (BP) reduction has been suggested for PRES, whereas BP reduction aggravates brain ischemia predisposing to infarct. Minimal short segment stenosis of the left anterosuperior division of MCA was suspicious of a coexisting RCVS as well. It is likely that PRES, RCVS, and cervicocephalic/coronary artery dissection are the different components of the postpartum vasculopathy spectrum; at times, two or more components coexist, the frequency of which is unknown at present. Routine performance of an angiogram for all patients with PRES/RCVS, preferably a computed tomography/catheter based angiography, particularly including the extracranial cervicocarotid arteries can guide in further management. The presence of a coexisting dissection warrants the administration of anticoagulants/antiplatelet agents; however, this aspect should be explored in further prospective studies. Uniquely, no reports of PRES associated with isolated coronary artery dissections have been published until now. The usefulness of the routine use of coronary angiogram in helping to identify the coexisting silent/life-threatening coronary artery dissections remains to be explored.

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Conflicts of interest
There are no conflicts of interest.

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