Post-Partum Angiopathy presenting as Ischemic Stroke

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Abstract
Post-partum angiopathy is grouped within the category of reversible cerebral vasoconstriction syndromes. It is considered to be a rare but under-recognized cause of stroke especially in pregnancy. We present the case of a 24 year old female who presented with hemiparesis and seizure, and turned out to be a case of post partum angiopathy.

Introduction
A acute stroke is a rare but potentially devastating complication in pregnancy or puerperium. Unlike an incidence of 10.7 strokes per 100,000 woman-years among non-pregnant women of comparable age, one study showed a threefold increased risk of stroke during pregnancy. 1 Post-partum angiopathy (PPA), a cerebral vasculopathy is considered as a rare but under-recognized cause of stroke in pregnancy. 2 PPA is often grouped with other conditions under the category of reversible cerebral vasoconstriction syndromes (RCVS) even though these conditions may have a different pathophysiology.

Case
A 24 year female presented with an episode of generalized seizures at 36 weeks of gestation. The seizure was preceded by severe headache for 2 days. At a local hospital, she was found to have hypertension (BP 180/90 mmHg; no past history of hypertension) and immediately, a caesarean section was done. She remained unconscious after that and her attendants noticed decreased movement of right side of body. She was then transferred to our center in ICU after 5 days of LSCS. There was history of a spontaneous abortion one year back. At presentation, she was responding to painful stimuli with movement of left side of body only. There was gaze preference towards left side and bilateral plantars were extensor. We considered the possibility of cerebral venous thrombosis and an Magnetic Resonance Venogram (MRV) was done, which was normal. But the MRI Brain showed multiple acute infarcts in left Anterior cerebral artery (ACA), bilateral Middle cerebral (MCA) and left Posterior cerebral artery (PCA) territories and a large acute infarct between Left ACA and MCA territory (Figure 1). Magnetic Resonance Angiography revealed multiple segmental areas of stenosis involving ACA, MCA and PCA, suggestive of Post-partum angiopathy (Figure 2). In addition the patient had elevated liver enzymes (Total Bilirubin 5.07 mg/dl, Direct 3.35 mg/dl, AST: 151 IU/L, ALT: 264 IU/L, Alk. Phosphatase: 350 IU/L), deranged renal function tests (serum creatinine 1.8 mg/dl), low platelets (60000 per cu mm) and altered coagulation parameters (INR 2.6). Urine was negative for protein. There was no pedal edema. She also had a large hematoma anterior to uterus which was managed conservatively in view of coagulopathy. We did not give her heparin or antiplatelet for CVA and managed her with antiepileptic, calcium channel blockers, plasma and blood transfusion. Patient’s sensorium improved gradually and subsequently, the hemiparesis also improved partially. Her metabolic parameters normalized after two weeks of admission and abdominal hematoma also resolved. Work up for systemic vasculitis and genetic markers of thrombophilia which was negative. This is a rare case of Reversible vasoconstriction syndrome which presented as post-partum angiopathy with complication of Ischemic stroke. In addition this patient some had features overlapping with Pre-eclampsia/ Eclampsia with HELLP syndrome (like elevated liver enzymes, low platelets, seizure). She had a favorable outcome without use of heparin or steroids.

Discussion
Traditional cerebrovascular risk factors (e.g., hypertension, smoking, hyperlipidemia) contribute to the etiology of stroke in pregnancy, but there are additional factors that pertain to the unique physiology of pregnancy.

Fig. 1: Diffusion weighted Imaging (DWI) and apparent diffusion coefficient (ADC) images of brain MRI showing left ACA-MCA watershed territory acute infarct

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Received: 23.03.2015; Revised: 25.08.2015; Accepted: 28.10.2015
Neurologic symptoms that occur respectively. Seizures can be inaugural, reported in 8–43% and 1–17%, persistent, and seizures have been phonophobia frequently occur. Focal nausea, vomiting, photophobia, and location followed by diffuse pain. Typical headache is bilateral (although it can be unilateral), with occipital status and/or focal neurologic deficits.1 Typical headache is bilateral (although it can be unilateral), with occipital location followed by diffuse pain. Nausea, vomiting, photophobia, and phonophobia frequently occur. Focal deficits, which can be transient or persistent, and seizures have been reported in 8–43% and 1–17%, respectively.1 Seizures can be inaugural, and recurrence is rare. Infarctions occur mainly in arterial watershed regions of the cerebral hemispheres, often between the posterior circulation and the carotid territories. Cerebellar infarcts can also occur. In two-thirds of cases, this starts during the first week after delivery, after an uneventful pregnancy or one complicated by proteinuria or HELLP (hemolysis, increased concentrations of liver enzymes, low platelet count) syndrome. Hormonal fluctuations (sudden fall in concentrations of estrogens and progesterone) might trigger the syndrome.2 Hypertension is commonly but not invariably present at initial clinical presentation.2 Neurologic symptoms that occur late in pregnancy and in puerperium often warrant a clinical diagnosis of eclampsia (if concomitant hypertension and proteinuria are present) or even “atypical eclampsia” in the absence of these features. Brain imaging is not always performed in these circumstances, and many patients are treated with intravenous magnesium sulfate with resolution of symptoms.2 If done for deficits lasting more than 24 hours, three types of stroke can be seen—convexity subarachnoid hemorrhage, intracerebral hemorrhage, and cerebral infarction with or without reversible brain edema. A combination of lesions can be present, and different types of lesion can develop successively. In one study, 55% of patients had a normal initial CT or MRI scan, but 81% had visible lesions when imaging was repeated.3 Multifocal areas of vessel narrowing involving multiple large and medium-sized arteries are characteristic and should raise suspicion for the diagnosis of PPA. Vessel caliber may be dilated or normal between areas of constriction. When dilated, the appearance may be that of “beads on a string.” The anterior cerebral, middle cerebral, distal basilar, and superior cerebellar arteries are most commonly affected.2 Large- and medium-sized arteries are more commonly affected, and the degree of affection to smaller arteries remain unclear as the angiographic techniques are not sensitive to small arteries. Cerebrospinal fluid (CSF) examination, extensive serological tests, and biopsies of the brain and temporal arteries have shown no abnormalities in patients with RCVS.4 Differential diagnosis include Pre-eclampsia/eclampsia, CVT, Pituitary apoplexy, Aneurysmal SAH, carotid artery dissection, CNS vasculitis and CSF Leak hence all these conditions should be ruled out. The pathophysiology underlying PPA is unknown. Because most patients recover rapidly, definitive pathologic evaluation is usually not possible. Postmortem examination has been performed in three fatal cases and these have revealed no inflammatory changes or evidence of vasculitis.2 Similarities seen in PPA and eclampsia have led to speculation that they share a common pathophysiology and thus might be part of the same disease process.2 Prognosis of RCVS and PPA is good but it has been reported that the presence of stroke is the major determinant of persistent morbidity. Fulminant vasoconstriction resulting in progressive symptoms or death has been reported in exceptional cases. Recurrence of this syndrome has been reported only rarely.3 Treatment should include analgesics for headache, antiepileptic drugs for seizures, monitoring of blood pressure, and admission to intensive-care units in severe cases. Drugs nimodipine, verapamil and magnesium sulphate targeted at vasospasm can be considered when cerebral vasoconstriction has been detected. Duration of treatment ranges from 4 to 12 weeks.3

Conclusion
RCVS affects patients of all ages and has a female preponderance. The syndrome should be suspected in any patient who presents with recurrent thunderclap headaches or cryptogenic stroke, especially in post-partum period or after the use of vasoactive drugs. Further research form standard guidelines for treatment. Our case presented as post-partum angiopathy associated with some features of HELLP syndrome with complication (ischemic stroke) and recovered well.

References