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# MOYA MOYA DISEASE PRESENTING IN POSTPARTUM PERIOD

KEY WORDS:

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**ABSTRACT**

Moya moya disease (MMD) is a rare progressive vaso-occlusive disorder that involves the internal carotid arteries and their branches. MMD can present as both ischemic stroke and intracranial hemorrhage, and pregnancy may increase the risk of these adverse events. We present the case of a 30 year old woman with Moyamoya disease who presented as post partum cerebral venous thrombosis. It is suggested that clinical suspicion for Moyamoya disease should be high for any stroke in pregnancy.

## INTRODUCTION

Moya moya disease (MMD) is a poorly understood occlusive disease of the cerebral blood vessels involving large intracranial arteries, especially the terminal portion of the internal carotid arteries and the stem of the anterior and middle cerebral arteries.[1] A dense network of collateral circulation form around the steno-occlusive lesion, giving rise to the characteristic angiographic appearance. MMD can present as both ischemic stroke and intracranial hemorrhage. Pregnancy may increase the risk of these adverse events, particularly if it is complicated by hypertension.[2] There are limited case reports of Moya moya vasculopathy in pregnancy due to rarity of this condition.[3-5] We present the case of a 30 year old woman who developed post partum cerebral venous thrombosis and was subsequently diagnosed to have Moyamoya disease.

**Case presentation.**

The above-mentioned patient presented with sudden onset weakness of the right side of the body and loss of speech 3rd day postpartum in November 2016. There was no history of fever, convulsion, vomiting, visual disturbance or any symptom suggestive of raised ICP. She was known diabetic and hypertensive, and her blood glucose and BP was well controlled with medications during pregnancy. On examination, the patient was conscious, pupils bilaterally reacting, plantar nonresponsive on the right. There was complete hemiplegia on the right side, without sensory involvement, and she had sensory aphasia.

A noncontrast CT scan of brain revealed acute infarct in the left parietal lobe, in the left ACA-MCA watershed area. Contrast MRI of brain with MR venogram revealed ill-defined hyperintensity in the left superior parietal cortex and subcortical region with diffusion restriction, and asymmetric filling of superior left parafalcine cortical veins, suggestive of isolated cortical venous thrombosis with acute venous infarct.

The patient was managed conservatively, with systemic anticoagulation, initially LMWH followed by oral warfarin for 12weeks, empiric antibiotics, prophylactic anticonvulsants, and physiotherapy. The patient improved clinically with improvement in both muscle power as well as in speech. The patient was discharged in a hemodynamically stable condition.

Subsequently the patient presented in the outpatient department with weakness and tingling sensation in her left upper limb, which resolved spontaneously within few hours. There was no documented hypoglycemia or electrolyte disorder. A follow-up CT angiogram of brain revealed short steno-occlusive segments involving proximal portions of P2 segment of left PCA, M1 segment of left MCA, and poor run-off with thinning of M2 and M3 segments of left MCA; long segment stenosis involving A1 and A2 segments of left ACA, bilateral PCom, and the right MCA. The right MCA was transformed into multiple small collateral twigs immediately after its origin with no distal reformation. The cavernous and supraclinoid parts of the internal carotid arteries, the basilar artery, and the intracranial part of the vertebral arteries were normal. Great neck vessels on both sides were free of steno-occlusive disease.

With the above imaging findings, the possibilities in this age group for young stroke were: (1) vasculitis; (2) connective tissue disease; and (3) Moya moya disease.

The tests for ANA (Hep-2 method), APLA, ANCA, Factor V (Leiden) mutation, Prothrombin gene mutation, Antithrombin III, and Protein C & S deficiency were all negative.

Digital subtraction angiography of Brain shows stenosis of both supraclinoid internal carotid arteries. Collateral channels between PCA-MCA and PCA-ACA are seen. Pial collaterals from Moyamoya vessels faintly fill MCA with slow forward flow.

The patient was finally diagnosed as a case of Moyamoya disease and was treated conservatively with a combination of oral antiplatelet and warfarin. Patient was referred to neurosurgeon for further opinion and management.

## DISCUSSION.

Our patient was unique in the sense that cerebral venous thrombosis is a very rare presentation of MMD. An association between the two conditions has been suggested by few isolated case reports and small case series, [6-8] although a direct causal relationship is difficult to establish. According to Boundel et al, prothrombotic abnormalities are found in 4 out of 10 non-Japanese children with Moyamoya syndrome.[9] The presence of prothrombotic abnormalities in MMD can increase the tendency of both arterial and venous thrombosis, including cerebral venous thrombosis. Therefore we have screened our patient for all common inherited and acquired thrombophilic conditions, but all her reports were negative. It is possible that cerebral venous thrombosis might have occurred in the setting of pregnancy as pregnancy itself is a known prothrombotic state. But the role of co-existing MMD, as an additional risk factor contributing to the thrombotic event, cannot be downplayed.

The relationship between pregnancy and MMD is poorly understood. Though pregnancy has no direct casual relation with MMD, but a consistent pattern is reported in two major reviews of MMD in pregnancy [10]: the first in 1998 by Komiyama et al, who found 23 cases where MMD was diagnosed during pregnancy. Three of these 23 patients died and eight had ongoing neurologic impairment. By contrast, those who were diagnosed with MMD before pregnancy (31 case reports) had good outcomes with only one adverse event.

A nationwide survey of 280 centers in Japan, reporting MMD outcomes over five years, found 64 cases, of whom 59 women had MMD diagnosed before pregnancy, and five newly diagnosed. [11] In this report too, patients with previously diagnosed MMD had clearly better pregnancy outcomes compared to the newly diagnosed group.

In our case too, the patient presented with stroke in pregnancy and had permanent neurologic impairment.

It is not clear whether pregnancy itself alters the risk of intracerebral events in MMD, but there are suggestions that preeclampsia may. [12] Cerebral autoregulation is impaired in MMD. While the cerebral blood flow improves post bypass surgery, the autoregulatory mechanisms will not be restored by a direct extracranial-intracranial bypass. Severe hypertension (as with preeclampsia), hypotension, hyper- and hypocapnia may impair cerebral perfusion and autoregulation, predisposing to stroke. [13-14]

## CONCLUSION.

In conclusion it is suggested that Moyamoya disease should be considered as a differential diagnosis of any stroke in pregnancy, and wherever possible an MR angiography should be ordered as a part of diagnostic work up to identify this condition.

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