



## A NEAR MISS CASE REPORT OF MOYAMOYA DISEASE IN PREGNANCY : A MULTIDISCIPLINARY APPROACH.

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**ABSTRACT** **Summary:** Moyamoya disease in pregnancy is a rare, progressive, chronic, vaso-occlusive disorder that affects the internal carotid arteries and their branches. As known, pregnancy, being an hypercoagulable state, may increase the risk of these occurrence. **Case description:** We present a case report of Primigravidae, Asian, 30 year old, 34 weeks b/d unregistered, unimmunised presented with pain in abdomen since 5-6 hours, headache, epigastric pain since 3 hours in Obstetric emergency Unit of our hospital. General condition of the patient on arrival was pulse 98 beats per minute, blood pressure 170/110 mm Hg; Urine albumin 3+ on bedside urine dipstick, Deep Tendon reflexes brisk. Per abdominal examination was Uterus 30 weeks size, cephalic presentation, Fetal heart Rate 100 beats per minute, Non-Reactive CTG, per vaginal examination revealed Poor Bishop score in labour. Like most common cause of such presentation in obstetric emergencies unit, patient was diagnosed Severe preeclampsia with Nonreactive CTG with IUGR and Decision for Emergency LSCS was taken. On postoperative Day 2, patient developed sudden episode of right upper monoparesis with facial deviation with focal seizures. With multidisciplinary approach and detailed investigations, diagnosis of Moyamoya disease was made. Patient was started on medications accordingly and discharged with baby on day 14 after suture removal, She followed up till 6 weeks and was doing well. **Clinical Significance:** Although clinical findings can mislead us, a rare condition like Moyamoya disease should be kept in mind especially in high risk cases presenting as HTN disorder in pregnancy.

**KEYWORDS :** Moyamoya, Postpartum, CVA, Maternal Morbidity, Multidisciplinary

### INTRODUCTION:

Moyamoya means “puff of smoke” in Japanese literature and it means the tangled appearance of tiny vessels compensating for the blockage, is a chronic progressive, non-inflammatory vasculopathy. It can present with various clinical symptoms like ischemic changes, seizures or cerebral haemorrhage<sup>1</sup>. During pregnancy, increase in the hormonal levels of estrogen and progesterone promote vasodilation which leads to activation of the renin-angiotensin-aldosterone system (RAAS) which increases cerebral blood flow<sup>2</sup>. Studies have stated that risk of developing cerebral hemorrhage in pregnant women increases by 5.6 times more as compared to that in non-pregnant women<sup>3</sup>. In patients with Moyamoya disease, fragile cerebral vessels are more sensitive to normal physiological changes of pregnancy, and thus the risk of CVD is higher<sup>4</sup>.

### CASE REPORT:

We present a case report of 30 year old Asian by origin, Primigravidae with 34 weeks b/d unregistered, unimmunised presented with pain in abdomen since 5-6 hours, headache, epigastric pain since 3 hours in Obstetric emergency Unit of our hospital. Patient was seen by Senior doctor. General condition of the patient on arrival was pulse 98 beats per minute, blood pressure 170/110 mm Hg; Urine albumin 3+ on bedside urine dipstick, Deep Tendon reflexes brisk. Per abdominal examination was Uterus 30 weeks size, cephalic presentation, Fetal heart Rate 100 beats per minute, Non-Reactive CTG, per vaginal examination revealed Poor Bishop score. Like most common cause of such presentation in obstetric emergencies unit, patient was diagnosed Severe preeclampsia with Nonreactive CTG with IUGR and Decision for Emergency LSCS was taken. Investigations were traced with Haemogram showed Hb 9.2 g, Rest Investigations like liver function test, Renal function test, coagulation function test all were within normal limit. The surgery was uneventful except persistently raised Blood pressure post operatively. In spite of dual antihypertensive drugs used, it was difficult to maintain blood pressure below 150/110 mmHg. Postoperative Patient was shifted in Intensive care unit for monitoring. However, postoperative 12 hours in Intensive Care Unit, patient developed sudden focal seizures with right upper monoparesis with slight facial deviation of the angle of the mouth. Emergency Multidisciplinary help was called, and after Medicine consultation, CT brain with MR diffusion was done. It suggested acute infarct of left frontal lobe, old lacunar infarct in anterior aspect of right lentiform nucleus. MR angiography shows stenosis of bilateral ICA including proximal, mainstem, and middle supratentorial branches. (Figure 1). With multidisciplinary approach and detailed investigations, diagnosis of Moyamoya disease was made. Patient was started on Medication

accordingly and discharged with baby on day 14 after suture removal, She followed up till 6 weeks and was doing well.

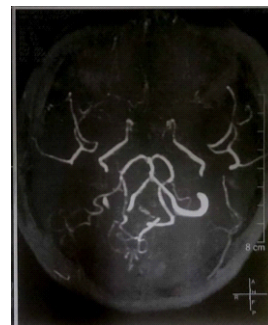
### DISCUSSION:

Differential diagnosis especially in high risk cases like severe preeclampsia, previous history of CVD, along with pregnancy, although rare Moyamoya disease should be kept in mind. Also, it has been found that in a patient diagnosed with Moyamoya disease, the risk of developing CVD after delivery is more<sup>5</sup>. Therefore, vigilant and long-term postpartum surveillance is necessary for women with Moyamoya disease.

**Consent:** Ethical principles were maintained throughout the case and consent of the patient was taken in local language.

**Source of support:** Nil.

**Conflict of interest:** None.



**Figure 1 :** MR angiography image showing stenosis of the C6-C7 segment of ICA in case of Moyamoya Disease patient.

### REFERENCES:

- Zhang H, Zheng L, Feng L. Epidemiology, diagnosis and treatment of moyamoya disease (Review). *Exp Ther Med*. 2019;17:1977–84.
- Takahashi JC, Ikeda T, Iihara K, Miyamoto S. Pregnancy and delivery in Moyamoya disease: results of a nationwide survey in Japan. *Neurol Med Chir (Tokyo)*. 2012;52(5):304–10.
- Miyakoshi K, Matsuoka M, Yasutomi D, Tanaka M, Yakubo K, Fukuiya T, Yoshimura Y. Moyamoya-disease-related ischemic stroke in the postpartum period. *J Obstet Gynaecol Res*. 2009;35(5):974–7.
- Galambosi PJ, Gissler M, Kaaja RJ, Ulander V-M. Incidence and risk factors of venous thromboembolism during postpartum period: a population-based cohort-study. *Acta Obstet Gynecol Scand*. 2017;96(7):852–61.
- Jeong Y, Jung YJ, Noh E, et al. The risk for future cerebrovascular disease in pregnant women with Moyamoya disease: a nationwide population-based study in South Korea. *BMC Pregnancy Childbirth* 22, 433 (2022)