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Moyamoya Disease in a 28 Year Old Pregnant Women

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1. Abstract

MoyaMoya Disease (MMD) is a cerebrovascular condition that can cause bilateral occlusion of distal internal carotid arteries, leading to ischemic or hemorrhagic stroke. The incidence of MoyaMoya Disease in Japan is reported to be 0.35 per 1 lac compared to the US where incidence is 0.086 per 1 lac [1]. There are two peaks of incidence with different clinical presentations. In children, ischemic symptoms are more common, while adults tend to present with intracranial hemorrhage [7]. Pregnant women with MMD are at increased risk due to pregnancy-associated changes such as increased cerebral perfusion pressure and thrombotic risk [3]. However, no guidelines exist for managing MMD in pregnancy, despite several case studies [2].

2. Case Presentation

We report a case of a 28-year-old pregnant female, gravida 4 para 2 live births 2 and 1 abortion, at 6 months of gestation who presented with sudden onset headache and vomiting. She had a history of seizure with right hemiparesis on the 11th day post-partum of previous pregnancy 7 years ago for which she was started on Tablet Levetiracetam 500 mg twice daily and was compliant. The patient was conscious, cooperative, and oriented to time, place, and person. On neurological examination, her higher mental function, cranial nerve examination, and sensory system were within normal limits. However, she had brisk knee reflexes bilaterally, and plantar reflex showed extensor response bilaterally. MRI revealed features of MoyaMoya Disease with occlusion of the left distal internal carotid artery and Moyamoya vessels in the left middle cerebral artery territory.

Pregnant women with MoyaMoya Disease are at an increased risk of cerebrovascular events, and management of MMD during pregnancy is challenging. This case highlights the need for vigilance in pregnant women with a history of MMD and emphasizes the importance of appropriate diagnostic imaging, such as MRI, to aid in diagnosis and management.

3. Key Messages

- MoyaMoya Disease can be regarded as a rare cause of stroke in pregnancy. However, the incidence of cerebrovascular events in a patient diagnosed with MMD is high at 50–75% [1].
- Several case studies describe MoyaMoya Disease in pregnancy but no guidelines exist to date for management of MoyaMoya Disease in pregnancy [2].
- Although there is no evidence to support the theory that MoyaMoya Disease would increase the maternal risk of morbidity and/or mortality, it is assumed that the normal physiologic changes that occur during pregnancy would make women with MoyaMoya Disease more susceptible to cerebrovascular events.
- The pathophysiology of MoyaMoya Disease and pregnancy poses specific concerns including a pregnancy-associated increased risk of thrombosis as well as increased cerebral perfusion pressure, stresses of labor and hypertensive diseases [3].
- The development of severe preeclampsia could cause a rapid deterioration of symptoms due to a cerebral vasospasm, hypertensive encephalopathy, and a heightened sympathetic drive. High

cerebral perfusion pressures overcome the limits of intrinsic autoregulation leading to endothelial damage, vasogenic edema, and barotrauma, which increase the risk of hemorrhagic stroke [5].

● Successful pregnancy outcomes may be achieved in women treated with Aspirin/Dipyridamole, Nifedipine and magnesium sulfate throughout the duration of pregnancy and in the postpartum period.

● The role of a surgical intervention is palliative rather than definitive and is used to treat the effects of cerebral ischemia.

4. Background

MoyaMoya Disease (MMD) is a cerebrovascular condition that can cause bilateral occlusion of distal internal carotid arteries, leading to ischemic or hemorrhagic stroke. The incidence of MoyaMoya Disease in Japan is reported to be 0.35 per 1 lac compared to the US where incidence is 0.086 per 1 lac [1]. There are two peaks of incidence with different clinical presentations.

In children, ischemic symptoms are more common, while adults tend to present with intracranial hemorrhage [7].

The incidence of cerebrovascular events in a patient diagnosed with MMD is high at 50–75% [1].

Pregnant women with MMD are at increased risk as there are concerns regarding pregnancy-associated changes such as increased cerebral perfusion pressure and thrombotic risk [3]. However, no guidelines exist for managing MMD in pregnancy, despite several case studies [2].

The pathophysiology of MMD involves the development of fragile collateral blood vessels due to changes in circulatory patterns and shear stresses at bifurcations at the base of the brain. The development of collateral vessels may be due to the upregulation of proangiogenic enzymes in response to increased blood flow [1]. Pregnancy is associated with an increased risk of thrombosis due to elevated levels of estrogen, decreased fibrinolytic activity, and increased plasma volume [4]. High cerebral perfusion pressures overcome the limits of intrinsic autoregulation leading to endothelial damage, vasogenic edema, and barotrauma, which increase the risk of hemorrhagic stroke [5].

Intracranial hemorrhage is prone to occur during the antepartum period, especially at ‡24 weeks, and cerebral infarction tends to occur postpartum.

The gold standard for diagnosing and grading Moyamoya disease is catheter-based DCA, with Suzuki's staging system used to classify its development into six stages.

As the angiographic stages progress, clinical deterioration often follows, with children more likely to progress than adults.

MRI is useful for visualizing areas of acute ischemia, and the «ivy sign» on the FLAIR sequence is diagnostic for MMD.

Perfusion imaging has been helpful in both pre- and postoperative evaluations, while TCD and intraoperative ultrasound can be used to assess the success of revascularization surgery [8].

There are no prospective trials that address the safest mode of delivery for women with MoyaMoya Disease. During labor, Valsalva and intracranial pressure can cause hemodynamic stresses on the fragile cerebral blood vessels, predisposing the patient to hemorrhagic stroke. Hyperventilation, alkalosis, and hypocapnia can cause a resultant decrease in the cerebral perfusion pressure, predisposing the patient to ischemic stroke.

Successful pregnancy outcomes may be achieved in women treated with Aspirin/Dipyridamole, Nifedipine and magnesium sulfate throughout the duration of pregnancy and in the postpartum period.

MoyaMoya Disease has no cure and surgical intervention is only palliative. Surgical treatment involves extracranial-intracranial revascularization, either through direct or indirect methods.

● The direct revascularization is between the superficial temporal artery and the middle cerebral artery (STA-MCA) combined with placement of the temporal muscle over the brain surface, a procedure called encephalomyosynangiosis (EMS)

● The indirect method is technically easier and hence is preferred. It includes EDAS, encephaloduroarteriosynangiosis and pial synangiosis. [4] Symptomatic progression occurs in 2.6% of patients undergoing surgery compared to 66% of patients without treatment.[1] Success of EDAS is dependent on the development of neovascularization, taking 3 to 6 months postoperatively for any reperfusion benefit to take place.[6]

Surgical revascularization is recommended at least 6 months prior to conception to maximize the benefit of surgery and reduce the risk of intracranial hemorrhage during pregnancy.

5. Case History

Our case was of a 28 year old female, gravida 4 para 2 live births 2 and 1 abortion, at 6 months of gestation who came to EMS at 8 pm complaining of sudden onset Headache associated with 2 episodes of vomiting since 8 hrs.

Headache was acute in onset, continuous, severe in intensity, throbbing type, occipital and frontal in location not associated with blurring of vision or diplopia.

Vomiting was projectile containing food particles, non-bilious, non-blood stained.

No other neurological history suggestive of seizure, LOC, Altered sensorium, limb weakness was obtained. She did not complain of any fever, chest pain, palpitation, syncope or presyncope.

Past medical history included a history of seizure with right hemiparesis on the 11th day post-partum of previous pregnancy 7 years ago for which she was started on Tablet Levetiracetam 500 mg twice daily and was compliant. Details regarding the cause were not available. Patient reported increased blood pressure reading in her previous pregnancy. The right limb power was regained over

time.

The patient was G4P2L2A1. Her first child was a 7 year old healthy male born out of a full term normal vaginal delivery. After the delivery of the 1st child, she experienced an abortion at 6 weeks for which Dilation and curettage was also done. The second child was a 2.5 year old male child born out of a full term normal vaginal delivery.

Her current pregnancy was not registered with her last menstrual period or Estimated Delivery date unavailable. No significant medical history related to pregnancy.

The patient was conscious, co-operative and oriented to time, place and person. Afebrile

P: 84 bpm, regular rhythm, BP 94/60 MM Hg in right upper limb with no postural drop.

RR 15 per minute with thoracoabdominal pattern. JVP was not raised. Neck stiffness was present.

On neurological examination, her higher mental function and cranial nerve examination was within normal limits. On assessing the motor system, the patient looked well nourished and well built, with a normal tone in all 4 limbs and a power of 5/5 in all 4 limbs. All superficial and deep tendon reflexes were 2+ EXCEPT B/L Knee reflexes 3+. Plantar reflex showed extensor response bilaterally. Cerebellar signs were absent. Sensory system examination was also within normal limits. On cardiovascular examination S1, S2 heart sounds were heard normally. No murmurs were heard.

Respiratory examination showed equal air entry bilaterally and no other adventitious sounds were heard. Abdominal examination revealed a uterus of around 20 weeks. Relaxed tone. Fetal movements were perceived.

From laboratory parameters her complete blood count showed a hemoglobin of 10.2 g/dl, a total leukocyte count of 8,300 cells/mm3 and 160,000 platelets/mm3. Her blood urea nitrogen and serum creatinine were 5 and 0.8 respectively. Her serum sodium, potassium and chloride counts were 138 mEq/L, 3.5 mEq/L and 110 mEq/L respectively. Her serum calcium and magnesium were 8.6 mg/dl and 1.50 mg/dl. Her lipid panel also showed normal levels. Normal TSH and free T3/T4 levels.

Serum homocysteine levels had shown a mild rise above normal limits. A set of rheumatological tests for anti-beta2 glycoprotein-1 IgM and IgG antibody, antiphospholipid IgM and IgG antibody and anticardiolipin IgG and IgM antibody had been found to be negative. Weakly positive antinuclear antibody levels of >1:80 with speckled cytoplasmic pattern had been found. Anti-double stranded DNA antibodies had been negative.

Ultrasonography for fetal viability had shown a single live intrauterine gestation with an estimated gestational age of 23 weeks. Malformation scan had shown no abnormalities.

Fundus examination had been significant with no papilloedema.

6. MRI Brain Had Shown

● acute intraparenchymal bleed in right thalamus, posterior limb of internal capsule, lentiform nucleus, peritrigonal region extending into both lateral ventricles, 3rd and 4th ventricle without midline shift.

● Encephalomalacia with perifocal gliosis had been seen in the left thalamus and peritrigonal region secondary to prior hemorrhage.

MR angiography of brain had revealed:

● Mild luminal narrowing of left CCA with high-grade stenosis of left ICA from its origin in its cervical course (90%)

● Severe luminal narrowing of intracranial left ICA in petrous, lateral, and Cavernous segments with non-visualization of supraclinoid ICA. Severe narrowing of the right supraclinoid ICA with multiple collaterals bilaterally.

● Non-visualization of bilateral ACA and right MCA and left PCA (Figures).

The patient had been admitted to the Medicine floor. Daily regular vital signs and neuromonitoring had been done. The patient had been given the following treatment:

1. Injection mannitol 100 ml intravenously three times a day

2. Tablet Levetiracetam 500 twice daily mg had been continued

3. Tablet ondansetron 4 mg had been given as required for vomiting

4. Tablet paracetamol 500 mg as required/ injection tramadol 500mg as required

5. Syrup lactulose 30 gm twice daily

6. Syrup glycerol 30 ml three times daily

7. Tablet Clarithromycin 500 mg twice daily

8. Multivitamin B complex tablets

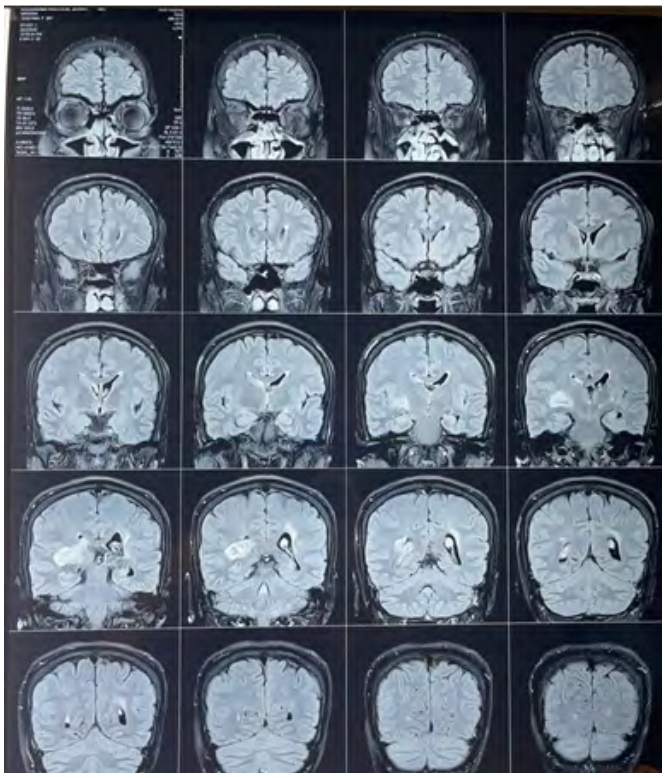
9. Bolus insulin regimen

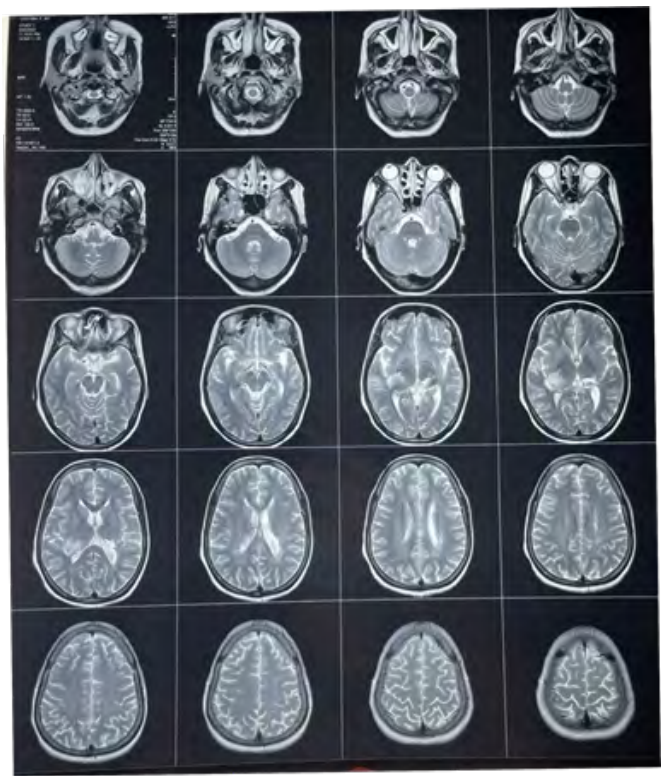
Nausea and vomiting had improved by the second day of admission, and headache had gradually improved by the 7th day. Occupational therapist referral had been done for lower extremity weakness, and recommendations had been taken. Daily obstetrician consultations had been done.

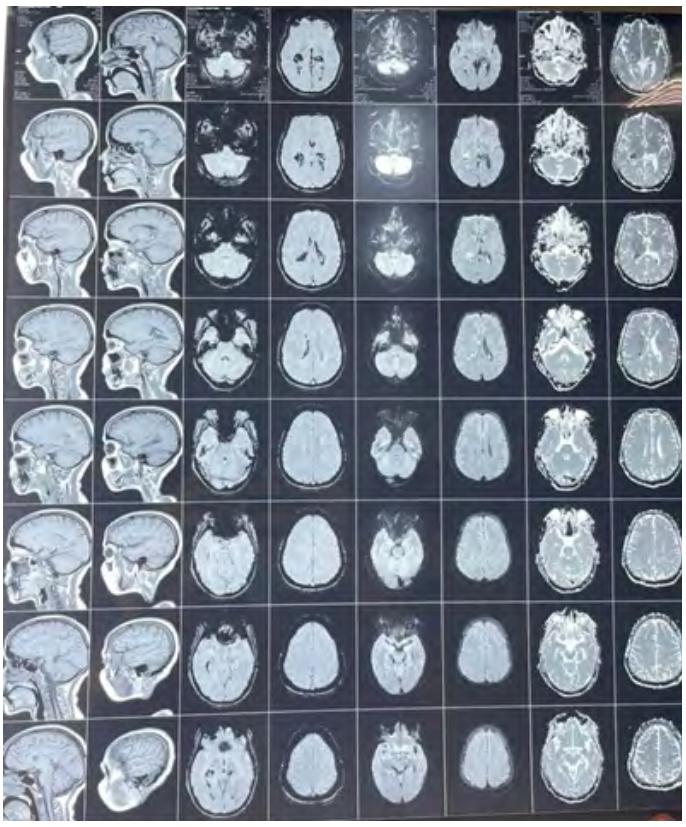
The hospital course had been complicated by an episode of fever for 3 days after 1 week of admission, but no source of infection in blood, respiratory system, or urinary tract had been found.

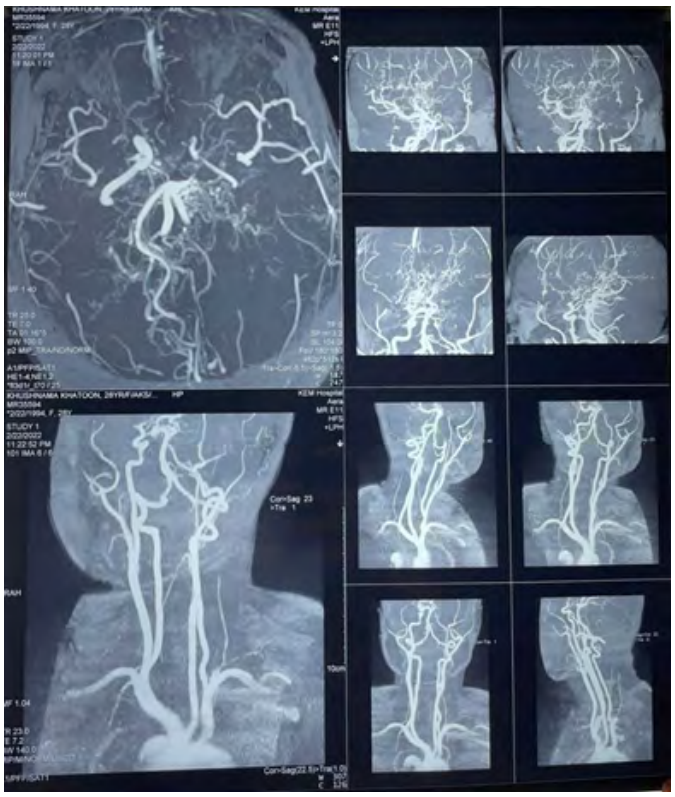
Neuromedicine, neurosurgery, and interventional neuroradiology referral had been done. A unanimous decision to perform DSA after delivery had been taken to prevent teratogenic exposure. Advice to continue Levetiracetam 500 mg twice daily had been taken with blood pressure monitoring and adequate hydration to prevent hypotension.

The patient had been discharged at 2 weeks and advised to follow up at Antenatal care OPD weekly, Neurology OPD to monitor bleeding and take further decision on antiplatelet therapy.









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