

A case study of MOYÀ MOYÀ disease disguised as Preeclampsia during pregnancy:A rare but critical challenge

Dr. ISHVARI SURESHKUMAR YADAV,Junior Resident , Department of obstetrics & gynaecology GGMC & Sir JJ Group of Hospitals

Dr. PREETI F LEWIS , HOU and Associate professor,Department of obstetrics & gynaecology GGMC & Sir JJ Group of Hospitals



INTRODUCTION

Moyamoya disease (MMD) is a rare cerebrovascular disease. Characterized by progressive stenosis or occlusion of the bilateral internal carotid arteries leading to the development of fragile collaterals. Disruption of the vessels can lead to both ischemic and hemorrhagic strokes Presenting symptoms includes Ischemic stroke,TIA, Intracranial hemorrhage, seizures, headache, cognitive or psychiatric changesThe prevalence of MMD is the highest in Japan, with an incidence of 3.14 cases per 100,000 individuals, The MMD affects women of reproductive age The women with MMD Has 54% prevalence of HDP, As increase in blood pressure occure to maintain cerebral blood flow.Currently , There is no treatment to reverse the primary disease and stroke prevention is main aim of treatment Medical management with antiplatet drugs (aspirin and dipyridamole) decrease the risk of microemboli as risk of thrombosis is increased during pregnancy[3]. Calcium channel blockers and magnesium sulphate have vasodilatory properties and can be used as antihypertensive drugs in pregnancy with MMD[1]. Surgical revascularization is performed to improve cerebral blood flow , However, the efficacy of this approach and long-term outcomes, particular in patients with hemorrhagic symptoms, remain unclear.There are no consistent guidelines for managing pregnancy and delivery in women with MMD, and more than 70% of pregnant women with MMD undergo cesarean delivery to avoid the risk of intracerebral hemorrhage or cerebral infarction, but uncomplicated vaginal delivery with either vaccum or forceps has also been frequently reported.

HISTORY

27 yr old Indian female, MS 10 yr , G4P3L3 All uncomplicated FTND with 8MGA , Unregistered, started experiencing Intermittent Headache, Generalized, Insidious onset, then She Visited Private Practitioner ,Found to have Raised BP She started on T Lobet 100 mg TDS Home BP Charting advised, Found to have raised BP Antihypertensive increased to T lobet 100 mg TDS and Nicardia10R TDS After few days , She started complaining of headache , blurring of vision and Pedal edema BP Recorded to 200/120 mmHg with Brisk DTR, Proteinuria of 2+ with impaired consciousness with Thrombocytopenia Provisionally Diagnosed as Impending Eclampsia As a Initial management Injectable Antihypertensive and Inj MgSo4 loading dose given Injectable Antihypertensive given repeatedly at fixed interval with BP monitoring Patient then referred to Higher centre for further managementPatient came to us, was examined BP found raised upto 170/110mmHg, With T Nicardia 30 XL BP was stabilized Blood and Blood Products transfusion done FHS monitoring being done , found to be decreased Emergency LSCS taken ivo Fetal Distress with impending preeclampsia Immediately Post Section , Patient was complaining of severe thunderclap headache. Patients continue to be drowsy , hypoglycemic with BP ranging from 200/130 to 180/120 mmHg NCCT done-S/O Right FrontoParietal lobe SAH with post ictal edema with its mass effect Pt started on T Nimodipine , Atorvastatin, Prazopressin, Temla 40 with D5 PintsCT Brain Angiography done- S/o Acute Non Hemorrhagic Infarct in Fronto Parietal Temporal region with ICA Occlusion with Resultant Collaterals S/o Moya Moya Disease Vangiography done- S/o Right supraclinoid ICA occlusion with limbic arch and pial pial collaterals s/o Moya Moya disease secondary to CNS vasculitis Pt started on T Aspirin 75mg OD

DISCUSSION

Moyamoya disease is characterised by presence of luminal thrombosis, hyperplasia of smooth muscle cells, thinned media and weakend arterial wall. In response to occlusion of distal internal carotid artery fragile collateral blood vessels and small microaneurysms develop which are due to changes in circulatory pattern and sheer stresses at bifurcation at the base of the brain MMD during pregnancy and delivery it theorized that the physiologic changes associated with pregnancy including increased blood flow, hypercoagulability, and hyperventilation during delivery, can worsen symptoms of MMD. Due to the lack of standard recommendations for its management, we present a case of MMD in pregnancy managed by a coordinated multidisciplinary team Patient with MMD have High incidence of HPD , Careful monitoring of their blood pressure is essential to prevent intracranial complications during the pregnancy and parturition. It is probably best to maintain a normal blood pressure throughout pregnancy and labor and postpartum period with a systolic blood pressure (SBP) between 100 to 160 mmHg and a diastolic blood pressure (DBP) below 110 mmHg, to facilitate delivery in order to reduce blood pressure and prevent intracranial hemorrhage

MANAGEMENT

MEDICAL MANAGEMENT:

Aspirin/ Dipyridamole
Nifedipine
calcium Channel Blocker
MgSo4

SURGICAL MANAGEMENT:

Extracranial- Intracranial revascularization
Direct- A)STA - MCA
B) Encephalomyosynangiosis
Indirect
A)EDAS
Encephalomyoarteriosynangiosis
B)Pial synangiosis.

CONCLUSION

This is a unique case of a SAH secondary to Moyamoya disease, Presented as preeclampsia. Patient was successfully managed and had stabilised with no neurological sequelae prior to discharge. Future pregnancies will require multidisciplinary team involvement to create a thorough antepartum, intrapartum, and postpartum Patient is advised to avoid contraceptive OC pills.

What are the warning signs?

When there are symptoms, they may include:

