Anesthetic Management of a Parturient with MoyaMoya and Sickle Cell Dz

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Introduction

Moyamoya disease was first described in Japan in the 1960's and it has since been found in individuals in the United States , Europe, Australia , and Africa; incidence in the USA is 0.086/100,000 people. It is characterized by chronic progressive stenosis of the terminal portion of the internal carotid arteries and the formation of collateral circulation of abnormal net-like blood vessels. The name "Moyamoya" means "puff of smoke" in Japanese and describes the look of the tangle of tiny vessels formed to compensate for the blockage. Studies show that pregnancy is associated with a 34% increased risk of stroke, due to underlying physiological changes of vasodilation, hypervolemia, hyper coagulopathy.

We present a case series of two patients with Sickle Cell dz (SCD) and MoyaMoya syndrome who had different baseline symptoms, similar anesthetic management but very different outcomes.

These comorbities make delivery planning complex and OB Anesthesia input was critical.

Case Report 1

23 G4P1021 with SCD and MoyaMoya dz underwent IOL /TOLAC at 38.6wks. Her SCD history includes multiple vaso-occlusive crisis and acute chest syndromes with recent crisis 2 weeks before IOL. She intermittently received exchange transfusions and was on folate with mild opioid regimen. Her MoyaMoya was incidentally discovered on imaging as a child, but had no basesline neuro deficits.

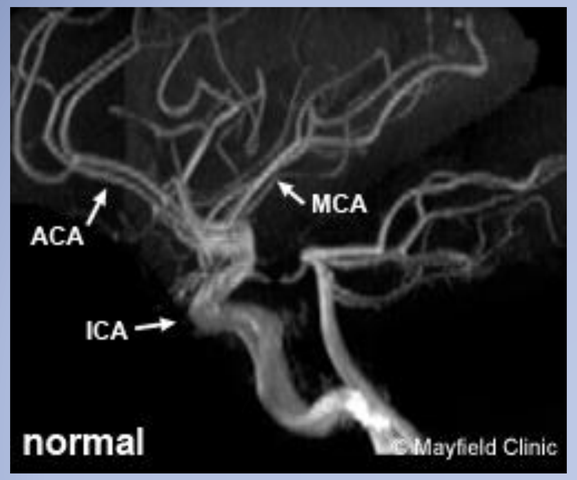

FIGURE 1:
Normal cerebral angiogram demonstrating regular blood flow through cerebral blood vessels.

FIGURE 2
To compensate for the narrowing arteries, the brain creates collateral blood vessels in an attempt to deliver oxygen-rich blood to deprived areas of the brain. These tiny collateral vessels, when seen on an angiogram, have a hazy, filmy appearance.

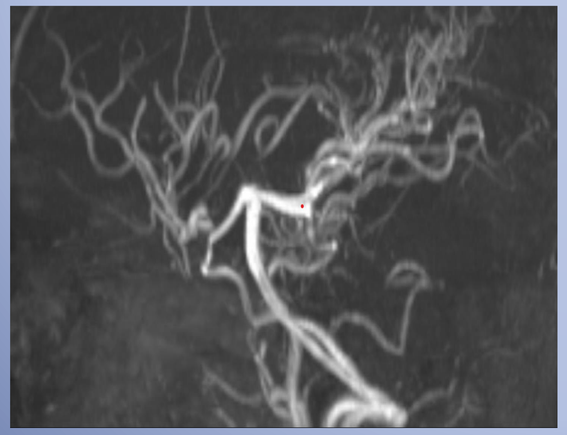

Figure 3 : MRI head angiogram for Case #2 stenosis of right CA with numerous collaterals to right MCA

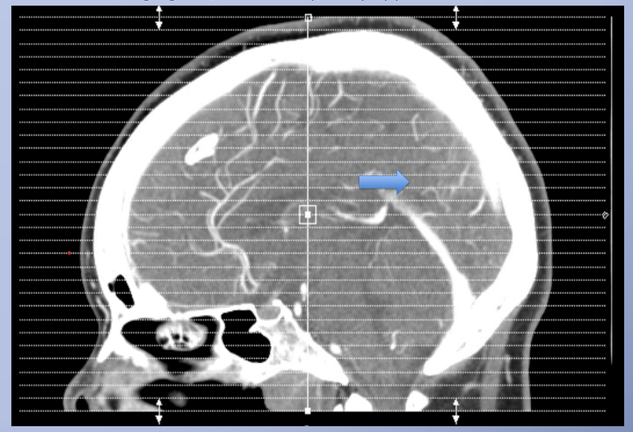

Figure 4: CT angiogram with contrast for head and neck Case #1 Showing tufts of blood vessel

Case 1 Cont'd

Anesthetic management included an early labor epidural. Pt progressed to cervical exam 6/50/-1 but was delivered via urgent c-section for Cat II tracing. She had a QBL 1468mL (postpartum hemorrhage) with a Hgb nadir 10->7.1. Otherwise, the pt recovered well and was discharged on PPD #4. She presented 7 days later from OSH with diffuse intracranial hemorrhage.

Case Report 2

22yo G1P0 with SCD and MoyaMoya dz underwent IOL at 37wks for gestational HTN. Pt PMH includes multiple CVA/TIAs since age 5yrs, DVTs, and complex partial seizures. Pt had undergone two encephalo duro arterio synangiosis (EDAS) procedures for her moyamoya (left EDAS in 2013, and right EDAS in 2017) and got monthly exchange transfusions for the previous 3 years to keep her level of Hb SS < 30%. We placed an early epidural (DPE) for labor analgesia. Pt progressed to cervical exam 7/60/-2, but developed symptoms of choriamniotis with fetal decelerations that led to an urgent c-section. Lower transverse c-section was performed under epidural anesthesia with QBL 968mL and no immediate complications. The pt did receive 2units PRBCs for acute on chronic anemia and to prevent vaso-occlusive crisis. Pain was well controlled with duramorph, morphine PCA, and adjuncts.

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Discussion

Moyamoya disease runs in families and thus thought to be the result of inherited genetic abnormalities. It primarily affects children, but it can also occur in adults. In children, the first symptom of Moyamoya disease is often stroke, or recurrent transient ischemic attacks (TIA, commonly referred to as “mini-strokes”), frequently accompanied by muscular weakness or paralysis affecting one side of the body, or seizures. Adults most often experience a ischemic stroke due to recurring blood clots in the affected brain vessels.

Pregnant patients have higher risk of stroke due to their physiologic changes. (**figure 5**) The risk further increases with concurrent SCDs patients with Moyamoya that have very fragile small vessels in the brain, and strokes can occur when the sickled RBCs lodge in these vessels. Surgical treatment of Moya Moya disease is intracranial bypass grafting after which successful labor and delivery has been reported.

In Case 1, our patient had few baseline symptoms concerning her MoyaMoya but a devastating outcome. There was controvery amongst the mulltidisciplinary teams concerning her mode of delivery. In the past, women were counseled to have a C/S but more recent literature suggested the larger blood loss and increased VTE risk is potentially just as dangerous as a short period of Valsava.

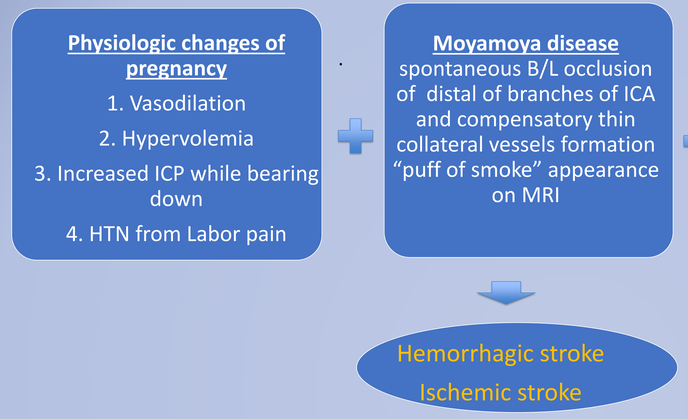

Figure 5
Increase risk of stroke in pregnant patient with MoyaMoya disease

Case #1: Baseline Symptoms	Goals of Care	Management	Post-Partum Course
• Mx SCD crisis/ acute chest • Intermittent exchange transfusions	• Hemodynamic stability • Avoid hypo/HTN • Avoid hypocapnia • Avoid hypothermia • Avoid hypoxemia	• Early epidural analgesia • Invasive monitoring (A-line) for hemodynamic control • Failed IOL->C-section	• ımmediate post-partum course uncomplicated • Presented to OSH with severe HA followed by diffuse cerebral Hemorrhage.
Case #2 • IOL for HTN • TIA/ CVAs, DVT	• Avoid pain and bearing down during labor. • Avoid hypertension. • Avoid hypocapnia	• Early epidural placement • A line placement • IOL failed-> C section.	• 2 PRBCs simple transfusion • Antibitoics for chorio • Discharged PPD #6 • Continues monthly exchange transfusions

References

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Discussion

Our second patient was a 22 G1P0 admitted for IOL 2/2 gHTN. Her pregnancy was complicated by a history of sickle cell disease and Moyamoya syndrome as well, but she had more baseline neurological symptoms that prompted two EDAS procedures. Both patients had failed vaginal attempts that ended in c-sections. More important than the mode of delivery, is the prior surgical correction in the second case compared to the first. Children usually respond to revascularization surgery best, but the majority usually have no further strokes or related problems after surgery. Without treatment, MoyaMoya dz can be fatal as the result of intracerebral hemorrhage. In case 1 her physiologic changes of pregnancy and fluid shifts with attempted IOL followed by c-section strained her already friable and narrowed cerebral blood vessels that led to her death.

For any delivery, there are certain things that must be in place for safety strategic planning amongst a multidisciplinary team to address baseline symptoms, mode of delivery, hemodynamic monitoring, anticoagulation, and blood /exchange transfusions. Our team included MFM, OB anesthesiology, hematology, neurology and neurosurgery.