PBLD – Table #9

A Patient with Sickle Cell Disease, Moyamoya Syndrome and a Basilar Artery Aneurysm Presenting for Dental Extractions

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Educational Objectives

By the conclusion of this learning module, participants will be able to:

- Discuss preoperative evaluation, anesthetic implications and post-operative considerations associated with sickle cell disease
- Describe management goals and strategies in a patient with moyamoya syndrome
- Discuss managements strategies in a patient with an intracranial aneurysm
- Construct a plan for perioperative management of a patient with sickle-cell disease, moyamoya syndrome and an intracranial aneurysm

Case History:

A 17 year old, 63 kg male with a history of sickle cell disease (SCD) – HbSS presents as an add-on late in the day for extraction of impacted wisdom teeth for severe jaw pain. He had an ischemic stroke at two years of age – and has residual right-sided weakness. The patient has not had recurrent stroke since, but has been on a regimen of monthly transfusions to keep his HbS less than 30%. He was diagnosed with moyamoya syndrome at 8 years old and has had gradual progression of the disease. On neuroimaging two years ago, he was diagnosed with a basilar artery aneurysm. He arrives without a pre-anesthetic evaluation, three hours prior to surgery for preoperative intravenous hydration, as coordinated by his dentist. Because of his chronic progressive vasculopathy, he is developmentally delayed and suffers from bouts of agitation.

Questions:

Should this case proceed? If not what preparation needs to be made prior to the procedure?

What are the perioperative concerns of SCD?

What is the association of moyamoya syndrome and intracranial aneurysms in relation to SCD? What are perioperative risks associated with moyamoya syndrome? What are the perioperative risks associated with a cerebral aneurysm?

Case History and Physical Examination (continued):

He has had no acute pain crises in the past year. He has had two episodes of acute chest syndrome. He has also had complications of iron overload secondary to transfusions. He suffers from splenomegaly, cholelithiasis and dactylitis. His moyamoya syndrome had progressed since he was 8 years old, with most significant stenosis occurring between ages 9-13 years. He has recurrent migraine headaches, for which he takes valproic acid. In 2011, he had an MRI/that revealed an aneurysm of the basilar artery between the left posterior cerebral artery and the left superior cerebellar artery. He has reactive airway disease and is exposed to second-hand smoke at home. He is in the 12^{th} grade and is in special education classes. His medications include valproic acid, albuterol, fluticasone, metoclopramide and ranitidine (for nausea associated with migraines). His BP is 138/56, HR 62, SpO2 100% on room air. He has a withdrawn affect. His cardiac and lung examination are normal. His right upper and lower extremity strength are 4+/5.

Questions:

Do we need further information about his medical treatment? What preoperative studies would you order?

Case History & Preoperative Studies:

Patient was last transfused two weeks ago. His hematocrit prior to transfusion was 28%. His most recent HbS was 35%. BUN was 11 and Cr was 0.5. Urine dipstick was negative for hematuria, proteinuria and leukocytes. A transthoracic echocardiogram (TTE) from seven months prior showed normal left ventricular (LV) function, mild LV dilation and an estimated right ventricular pressure of 26 mmHg.

Questions:

What risks and possible complications should be discussed with this patient and his family?

What are the systemic manifestations of sickle cell disease? (Hematologic? Skeletal? Cardiac? Pulmonary? Neurologic? Genitourinary involvement?)

Why might a chest radiograph be useful perioperatively in a patient with SCD?

Why are SCD patients prone to clinical dehydration? What is the role of preoperative hydration in a patient with SCD?

What is the role of transfusion – either exchange transfusion or simple red blood cell transfusion preoperatively in patients with SCD? Is there a goal hemoglobin or hematocrit for SCD patients? What about a goal HbS percentage? What about SCD patients at high risk for cerebrovascular accidents (CVAs)?

Should we involve other health care members in this patient's perioperative care? If so, who would we involve?

What are medical treatment options for patients with moymoya syndrome? What are surgical treatment options for patients with sickle cell disease and concomitant moyamoya syndrome?

Case Progression:

You speak to the neurosurgeon who saw the patient in clinic seven months prior. He believes the aneurysm does not warrant surgical intervention and will follow the patient annually with surveillance. You also notify the hematology team of the patient's procedure.

Questions:

What is your anesthetic plan? What clinical factors involved in red cell sickling need to be addressed? What are neuroanesthetic aims in regards to moyamoya syndrome and a cerebral aneurysm? How might they conflict with the perioperative management of SCD?

Would you give premedication to this patient?

What are your anesthetic hemodynamic goals for this patient? Will you keep the patient's blood pressure below, at or above baseline? What are your temperature goals? CO2 goals? Intravascular volume goals?

MAC versus general anesthesia? If general anesthesia, an inhalation agent or total intravenous anesthetic? What would you use for analgesia? Would you place an arterial line?

Intraoperative Care:

The patient had an uneventful induction of anesthesia and placement of an oral endotracheal tube. After starting a total-intravenous anesthesia with a propofol infusion and a bolus of fentanyl, the patient's blood pressure is 76/48. Will you give vasoactive medications? Which ones would you use?

During a lull in the wisdom teeth extraction, the fellow you are working with asks about anesthetics concerns in the hypothetical situation that this patient is going for cerebral aneurysm clipping. Would you reduce brain volume with mannitol and furosemide? Why or why not? Would you cool the patient for cerebral protection?

You plan for the patient's post-operative recovery. Where would you send this patient? If he has an uneventful recovery in the PACU after several hours, would you send him home?

Postoperative Care:

You extubate the patient in the operating room. He is hemodynamically stable. What is the number one cause of perioperative mortality in patients with SCD? What is the role of oxygenation post-operatively? What about the role of pain-control post-operatively? What is your hydration plan post-operatively?

Discussion:

Sickle cell disease (SCD) is a multisystem inherited disorder resulting from a mutation in the β-globin gene that codes for a variant of hemoglobin A called hemoglobin S. A non-polar valine substitutes the negatively charged glutamic acid in the sixth position of the chain. Hemoglobin S is highly unstable in the oxygenated state. It breaks down, releases iron intracellularly and produces oxidant damage to the red cell membrane. Hemolysis then releases iron into the blood stream, consuming free nitric oxide and leaving vascular endothelium vulnerable to free radical damage causing the progressive, chronic, inflammatory vasculopathy that characterizes SCD. Secondly, deoxygenated hemoglobin S is insoluble, and precipitates out of solution into a gel. This precipitation deforms the red cell into a "sickle" shape. The sickled red blood cells along with vasoconstriction can cause ischemia, vaso-occlusion and infarction.

About 0.2% of African-Americans are homozygous, HbSS and have sickle cell anemia (SCA). 8% are heterozygous, HbAS and have sickle cell trait (SCT). Patients with SCA have a median age of death in the late 40's and early 50's, have progressive end organ damage and acute exacerbations. The two common acute exacerbations are from acute pain crises and acute chest syndrome.

Acute chest syndrome (ACS) is the primary cause of death in patients with SCD. Patients have a new pulmonary infiltrate on chest x-ray that is not from atelectasis. Symptoms include fever greater than 38.5°C, chest pain, hypoxemia and wheezing. It occurs postoperatively in 10% of children and usually 2-3 days after surgery.

Another manifestation of SCD is cerebrovascular accident (CVA), affecting about 10% of children with the disease. These are usually watershed injuries, although there is also an increased incidence of subarachnoid hemorrhages (SAH). Children at risk of CVA are transfused to keep their HbS % less than 30%. This lowers their risk of stroke significantly. The risk of recurrence is significantly higher in SCD patients who developed moyamoya collaterals.

Moyamoya *disease* is a disease of unknown etiology with an angiographic pattern consisting of progressive stenosis of the internal carotid arteries, with the formation of collateral vessels giving a "hazy puff of smoke" appearance. Moyamoya *syndrome* is characterized by the same angiographic changes in known pathologic states such as SCD, neurofibromatosis or Down syndrome. The proposed pathogenesis in SCD includes chronic inflammation causing impaired cerebral autoregulation, sickled erythrocytes causing vascular injury, vascular endothelial hyperplasia and finally stenosis.

SCD is also associated with intracranial aneurysms. Sickled erythrocytes adhere to vascular endothelium, causing arterial endothelial damage. Dysfunctional vasoconstriction and autoregulation cause hemodynamic stress at these points in the arterial wall, creating aneurysms and also sometimes hemorrhage. Patients with SCD have a higher incidence of posterior circulation aneurysms as compared to the rest of the population. SCD patients also have other pathologic changes of cerebral vasculature, such as ectatic or tortuous vessels – which can be friable and predispose to subarachnoid hemorrhages.

Patients with SCD are at risk for both nonspecific perioperative complications and SCD complications. Sickle cell complications range from 0% to 19%. Acute sickle exacerbations occur around 15% of the time. Total perioperative mortality is around 1.1%. Anesthesiologists must consider the disease process as a progressive, chronic vasculopathy, rather than random exacerbations of acute sickle crises. Several anesthetic issues arise in SCD patients, particularly those complicated by moyamoya syndrome and cerebral aneurysms. Caring for a patient with any one of these conditions is challenging. The combination of the three in one patient makes this anesthetic particularly difficult.

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