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An Acromegalic Patient with Sickle Cell Disease Undergoing Endoscopic Trans-Sphenoidal Surgery: Anesthetic Concerns for a Neuroanesthesiologist

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Abstract

Sickle cell disease (SCD) refers to a group of hemoglobinopathies that include mutations in the gene encoding the β subunit of hemoglobin. The glutamine in the heme portion of the hemoglobin molecule is abnormally substituted by valine. Sickle hemoglobin (hemoglobin type S), when gets deoxygenated, tends to polymerize and aggregate leading to vaso-occlusion and organ ischemia. Such patients are at increased risk of perioperative mortality and severe complications like vaso-occlusive crisis, acute chest syndrome, and congestive heart failure. We describe the perioperative management of a case of SCD with acromegaly scheduled for trans-sphenoidal removal of a functional pituitary adenoma. The acromegalic habitus, the cardiovascular effects of acromegaly, and the hormonal imbalances due to pituitary adenoma pose challenges in addition to the challenge of preventing complications of SCD making the anesthetic management more exigent.

Keywords
- acromegaly
- anesthesia
- sickle cell disease

Introduction

Sickle cell disease (SCD) is caused by a single-base substitution that leads to the production of mutant hemoglobin type S (HbS). Deoxygenation causes the hemoglobin (Hb) molecules to form insoluble tetramers that polymerize. This polymerization causes the deformation of the membrane of red blood cells, leading to their sickling and decreasing their ability to traverse the capillaries of the microcirculation. A rise in sickle hemoglobin levels (HbS) increases the risk of end-organ ischemia, perioperative mortality, and morbidity. We describe the anesthetic concerns and successful perioperative management of a case of SCD with acromegaly scheduled for trans-sphenoidal surgery (TSS).

Case Report

A 26-year-old male patient presented with an increase in the size of hands and feet, headaches, and a decrease in vision. He was diagnosed to have acromegaly due to a functional pituitary adenoma (size = 2.2*1.4 cm). At the age of 4 years, he presented with episodes of bone pain, fever, and weakness. Hb electrophoresis and genetic testing were done and he was diagnosed to have SCD. He had multiple episodes of bone pain and priapism in the past 2 years. The patient was on tablet hydroxyurea and folate for SCD.

The patient was planned for endoscopic TSS for the pituitary tumor. The patient had Hb of 7.2 g/dL and Hb electrophoresis showed an HbS level of 73.2%. Blood

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transfusions were given preoperatively to achieve Hb of 10.0 g/dL and reduce the HbS level to 28.8%. The patient's functional pituitary tumor added another layer of complexity to his anesthesia management necessitating an assessment of the patient's hormonal profile and cardiovascular condition. The preoperative echocardiography was normal and the hormonal profile showed raised growth hormone and insulin-like growth factor levels and decreased serum cortisol levels. Renal function tests, serum electrolytes, liver function tests, and coagulation profile of the patient were within normal limits. The patient underwent an ophthalmological examination for perimetry in view of pituitary lesion and to rule out complications of SCD like sickle cell retinopathy, vitreous hemorrhage, and retinal detachment. The patient was counselled about perioperative complications related to SCD, functional pituitary tumor, and the surgical procedure and advised tab alprazolam 0.25mg at the bed time to reduce his anxiety.

Standard fasting guidelines were followed. During the fasting period, normal saline was infused to prevent dehydration and subsequent sickling. The patient was anesthetized with fentanyl, propofol, atracurium, and sevoflurane as per standard anesthesia protocol. Patient was monitored continuously for electrocardiogram, invasive blood pressure, oxygen saturation), bispectral index, end-tidal carbon dioxide, pulse pressure variation (PPV), systolic pressure variation, temperature, urine output, and intermittent arterial blood gas (ABG). Compression stockings were put on the patient's legs. A forced air warmer was used to maintain normothermia strictly throughout the surgery. The surgery was completed in 3 hours with a blood loss of 200 mL. Normal saline was infused (PPV $<$ 13% intraoperatively) to maintain euvolemia throughout the perioperative period and a urine output of more than 0.5 mL/ kg/h was ensured. Intermittent ABGs were done to rule out acidosis, hypoxia, and hypernatremia. The intraoperative course remained uneventful and the trachea was extubated after completion of the surgery. Postoperatively adequate oxygenation, hydration, and analgesia were ensured. He was discharged on the fifth postoperative day.

Discussion

The principal anesthetic concern in the case of SCD is to prevent sickle cell crisis in perioperative period by ensuring adequate oxygenation, hydration, analgesia and preventing acidosis and thrombotic complications.

Prolonged fasting hours must be avoided as dehydration triggers HbS polymerization. Adequate intravascular volume has to be maintained perioperatively. Hypotonic fluids may decrease the red blood cell sickling and hence may be preferred; however, they cannot be used in intracranial neurosurgical procedures.

Hypoxia is an important trigger of the sickle cell crisis. In patients with normal oxygen saturation curves, pulse oximetry quite accurately represents the partial pressure of oxygen (PaO_2). However, patients with sickle Hb usually have significant rightward shift of the oxyhemoglobin curve; so, the estimate of PaO_2 based on pulse oximetry can be inaccurate. Also, the increased amounts of carboxyhemoglobin and methemoglobin present in HbS can lead to false overestimation of arterial saturation using a pulse oximeter.[1] Hence, it is important to do ABG confirmation. Hypothermia can lead to reflex vasoconstriction, increases the capillary transit time and red cell sludging and may lead to shunting of blood from the bone marrow.[2] It is a known precipitant of sickle cell crisis and hence be avoided at all costs. Preoperative blood transfusions are considered beneficial as literature collaborates the benefit of decreasing HbS in circulation and increasing the level of normal Hb tissue oxygen delivery over increased viscosity when the transfused Hb target is kept at or just below 10 g/dL.[3] In patients undergoing neurological or cardiac surgery, the preoperative exchange transfusions should be done targeting HbS less than 30%.[4] As SCD is a hypercoagulable disease associated with an increased risk for ischemic stroke and venous thromboembolism, adequate deep vein thrombosis prophylaxis should be instituted.[5]

SCD is related to various other neurosurgical conditions. Forty-three percent of patients with SCD and strokes have moyamoya like collaterals on imaging studies.[6] Sabbagh and Kedar had described pituitary adenomas in two patients of SCD that were managed medically.[7] Cerebrovascular complications occur in 13 to 17% of SCD patients with the causes being cerebral infarction and brain hemorrhagic complications.[8] Repeated vaso-occlusions and ischemia cause neovascularization in the brain, and these blood vessels being very friable, can rupture causing intracranial bleeding. Similarly, hematuria is caused by ischemic papillary necrosis and rupture of the renal capillaries and vitreous and retinal bleeding can occur as a result of proliferative and nonproliferative retinopathy.[9]

Conclusion

Patients with SCD are prone to have several systemic complications. The target hemoglobin level of 10 g/dL and HbS levels less than 30% should be ensured before surgery. Perioperative stress, dehydration, acidosis, hypothermia, and pain must be avoided. Additionally, the anesthetic management of the patient with acromegaly should include preparation for managing a difficult airway, optimization of cardiovascular status, provision of a bloodless surgical field, and an early and smooth recovery from anesthesia for postoperative neurological assessment.

Conflict of Interest
None declared.

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