Full Case:

30 y/o with avascular necrosis of her right hip was admitted for a total hip arthroplasty. Her hematocrit was 22%, blood pressure was 130/90 mm Hg, and pulse was 107 beats per minute. She had a past medical history of recurrent joint and bone pain, jaundice, and abdominal pain requiring multiple hospitalizations for analgesia.

Questions:

What is sickle cell crisis?

Sickle cell crisis refers to the acute clinical picture generally caused by sickling of red blood cells in the microcirculation. Four main clinical types of crises have been described, as follows:

- Vascular occlusion crises with organ infarction and pain
- Hemolytic crises with hematologic features of sudden hemolysis (often associated with G6PD disease)
- Sequestration syndrome with sequestration of red blood cells in the liver and spleen causing their massive, sudden enlargement, and an acute fall in peripheral hematocrit
- Aplastic crises with bone marrow suppression

The most common crises in sickle cell disease (SCD) is vaso-occlusive crisis (VOC) which is characterized with intermittent, recurrent acute episodes of severe pain. It is generally felt that the central cause of pain is ongoing acute ischemia, vaso-occlusion, and infarction. The predominate pathophysiology and nociceptive pathways have not been elucidated. Patients in VOC present with fever, anemia, spine pain, limb pain, and abdominal pain. They are tachypneic and may have an enlarged liver and spleen in addition to abdominal tenderness. Serjent et al. reported that in 118 patients having 183 painful crises, the location was noted to be in the lumbar spine (49%), abdomen (32%), femoral shaft (30%), and the knee (21%). There is usually no identified triggering agent for most VOCs. The average rate of painful crises per patient year is 0.8%. There is a subset of patients (5.2%) that average 8 to 10 crises a year, which accounts for 33% or all
hospitalizations. Mortality positively correlates with increased pain rate in adults.


**Discuss the pathogenesis of sickle cell crisis.**

**Discuss preoperative exchange transfusion in this patient.**

Controversy exists as to whether these patients benefit from preoperative exchange transfusion. For high-risk surgical procedures, the exchange transfusion should be performed with coat-free packed red blood cells to reduce the HbS fraction to less than 40%. However, evidence indicates that most surgical procedures can be safely performed in patients with sickle cell disease (SCD) without aggressively transfusing. For example, for certain surgical procedures it has been shown that a packed red blood cell transfusion done simply to increase the hemoglobin (Hb) level to 10 g per dL is as effective in decreasing perioperative morbidity as is transfusions to reduce the HbS levels to less than 40%. Exchange transfusion is a treatment used in patients with SCD complicated by acute chest syndrome (ACS). It is beneficial to lower the hematocrit to lower than 35% with an exchange transfusion because an increased hemoglobin will increase blood viscosity and stasis in the circulation.
What are the indications for blood transfusion in SCD?

The indications for blood transfusion in SCD include the following:

Acute Conditions
- Vaso-occlusive crises (VOCs) with organ infarction and dysfunction
- Sequestration syndrome
- Acute hemolysis with exacerbation of anemia
- Aplastic crisis
- Acute chest syndrome (ACS)
- Acute multiorgan failure
- Severe unresponsive priapism
- Protracted painful crisis
- High-risk surgery (e.g., cardiovascular, thoracic)

Chronic Conditions
- Intractable leg ulcers
- Complicated pregnancy
- Stroke
- Recurrent painful syndrome
- Recurrent ACS
Discuss the anesthetic management for this patient. Discuss both regional and general anesthesia considerations.

A retrospective study reviewed 1,079 anesthetics and noted an apparent association between perioperative complications and the use of regional anesthesia. However, this study did not include obstetric procedures and the tendency that clinicians often use regional anesthesia for obstetric patients. Other studies did not find an advantage of regional anesthesia.

Theoretically, regional anesthesia may produce a compensatory vasoconstriction in the non-blocked area but this has not been supported in the literature. Yasler et al. found that epidural analgesia markedly improved oxygenation in children in vaso-occlusive crisis (VOCs). He also reported superior pain control through opioid sparing effects.

Therefore almost any anesthetic technique can be used if the following principles are considered:
- Good intravenous access for maintenance of intravascular volume
- Adequate oxygenation - denitrogenation before induction of general anesthesia
- Endotracheal intubation for general anesthesia to ensure a controlled airway and adequate ventilation
- Maintenance of adequate oxygen-carrying capacity through judicious use of red cell transfusions
- Maintenance of normothermia
- Opioids for analgesia carefully titrated for perioperative and postoperative analgesia
- Avoidance of tourniquet use
- Consider regional analgesia
. Avoidance of hypotension and venous stasis to prevent sickling of red blood cells
. Avoidance of hypoventilation of lungs to prevent acidosis
. Possible increased inspired oxygen concentration to ensure maintenance of normal to increased PaO2


Discuss how the comorbidities associated with SCD can affect your anesthetic.

Sickle cell anemia is a disease that can affect every end organ. Therefore evaluating the patient for end-organ dysfunction is paramount. This patient was anemic, tachycardic, and hypertensive. She may have dyspepsia, a hyper-reactive airway, chronic renal insufficiency, liver dysfunction, might be debilitated with cardiopulmonary complications. Patients with SCD often present with cardiomegaly, pulmonary hypertension, and heart failure. For this surgery, she will may require transfusions and may be difficult to cross match. In the postoperative period, pulmonary infarcts and infection are common. All these problems, in addition to the potential for acute
crisis and/or sequestration syndrome, pose a greater risk than normal for perioperative morbidity and mortality. The patients at greater risk include those with a homozygous state SCD, sickle disease (HbSC), and sickle thalassemia. In contrast, patients with sickle cell trait do not have an increased risk for intraoperative morbidity.


What is the treatment of sickle cell crisis?

The treatment of sickle cell crisis is to break the vicious cycle of sickling, ischemia, pain, and end-organ failure. The principles of treatment of painful sickle cell crisis include following:

. Bed rest
. Hydration
. Oxygen therapy
. Treatment of infection
. Analgesics (consider patient-controlled analgesia)
. Consider regional analgesia
. Transfusion to reduce the HbS concentration
. Incentive spirometry
. Maintenance of normothermia
