Perioperative Pain Management of a Patient with Shwachman-Diamond Syndrome

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Introduction

Shwachman-Diamond Syndrome (SDS) is a rare disorder that affects multiple organ systems in the body. Patients with SDS often have recurrent upper respiratory infections, decreased FRC, and hepatic dysfunction. Oxygen desaturations and respiratory collapse can occur if caution is not used when administering opioids. Other pain modalities, such as acetaminophen, are not recommended as they can mask the signs and symptoms of an acute infection. For these reasons, patients with SDS pose a unique challenge to the anesthesiologist in the perioperative period.

Syndrome Characteristics

- Autosomal recessive inheritance
  - Mutation of 7q11
- Pancreatic exocrine insufficiency
- Malnutrition of fat-soluble vitamins
- Hematologic dysfunction
  - Neutropenia
  - Thrombocytopenia
  - Anemia
- Skeletal abnormalities
  - Rib cage abnormalities
  - Femoral head dystosis
  - Kyphoscoliosis
  - Syndactyly
- Hepatic dysfunction
  - Hepatomegaly
  - Elevated hepatic enzymes

Case Discussion

A 15-year old male with SDS, presented with severe abdominal pain, ascites, and dyspnea with rales. He had recently developed graft-versus-host disease (GVHD) following bone marrow transplantation. Although he had significant liver failure, pulmonary insufficiency, and renal dysfunction, his main complaint was intractable abdominal pain. He was scheduled for a flexible bronchoscopy and bone marrow aspiration biopsy. His platelet count was 145 K/μL and creatinine was 0.6 mg/dL.

Induction was accomplished by IV rapid-sequence using propofol and succinylcholine. Dexametomidine and sevoflurane were administered for maintenance of anesthesia. The patient was not given any opioids intraoperatively for concern that extubation criteria would not be met at the end of the case. The procedure was uneventful, and the patient was extubated without any complications. He was initially given IV morphine boluses for pain in PACU but was soon converted to a hydromorphone PCA due to inadequate pain control.

Pain Management

As there was concern for his compromised pulmonary status, opioid-sparing agents such as IV ketorolac and acetaminophen were considered. In the setting of thrombocytopenia, liver failure, and his immunocompromised state, those modalities were not appropriate.

An ultra-low dose ketamine infusion was initiated as an opioid adjuvant, and it did provide modest pain relief. Due to continued pain, the decision was made to initiate IV methadone therapy due to its long duration of action and lack of active metabolites. The patient began to experience adequate pain relief and was eventually transitioned to oral methadone. He was discharged home with the plan for a slow methadone taper with oral hydromorphone for breakthrough pain. A multi-modal approach to perioperative pain control can be beneficial in this challenging patient population.

References