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Acute Splenic Sequestration Crises with Splenic Artery Thrombosis following Laparoscopic Cholecystectomy in Sickle Beta Thalassemia

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Background	Sickle beta thalassemia (Hb S/β) is a benign hemoglobinopathy; however, its clinical presentation di ers depending on the inheritance of either hemoglobin sickle-β0 thalassemia (HbSβ0) or hemoglobin sickle-β+ thalassemia (HbSβ+). Compared to HbSβ0, in which there is zero production of beta-globin, patients with HbSβ+ exhibit a milder disease presentation due to reduced production of Hemoglobin A (HbA). ere have been eight reported cases of patients with HbSβ+ presenting with acute splenic sequestration crisis (ASSC) in the literature. Yet, none of these cases demonstrated laparoscopic surgery as the main catalytic factor.
Summary	A 47-year-old female with a history of repaired mitral valve prolapse, cholelithiasis, and sickle-beta thalassemia (HbSβ+) presented with left upper quadrant abdominal pain eight days status-post-laparoscopic cholecystectomy with intraoperative cholangiogram. Admission labs revealed an elevated total bilirubin of 1.3 mg/dL, alkaline phosphatase of 143 unit/L, leukocyte count of 24,000/cm³, and a platelet count of 582,000/µl. Hemoglobin and hematocrit were 9.3 gm/dL and 29.9%, respectively. A computed tomography scan of the abdomen demonstrated global splenic infarction secondary to thrombosis of the splenic artery. It was determined that no surgical intervention was required. e patient was treated with a heparin drip and subsequently transitioned to low molecular weight heparin (LMWH) on discharge on hospital day 3.
Conclusion	Hypoxic conditions of surgery can contribute to a sickling crisis, ultimately leading to splenic infarction in sickle beta-thalassemia patients. Based on prior case reports, ASSC may be triggered by infection, high altitude exposure, and systemic in ammatory response in patients with HbSβ+. We suggest surgery be considered a potential precipitating factor for ASSC in patients with HbSβ+ due to the high incidence of hypoxemic events in the peri- and postoperative periods. Such patients should receive up-to-date hemoglobin electrophoresis preoperatively to determine their risk of sickling in the postoperative period.
Key Words	sickle beta-thalassemia; acute splenic sequestration crises
Abbreviations	Hb S/β : sickle-beta thalassemia, ASCC: acute splenic sequestration crises, HbSβ0: hemoglobin sickle-β0 thalassemia, HbSβ+: hemoglobin sickle-β+ thalassemia, HbA: hemoglobin A, LMWH: low-molecular-weight heparin, ASA: aspirin, SCD: sickle cell disease, SCT: sickle cell trait

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Case Description

While the heterogeneous disorder of sickle beta-thalassemia (Hb S/β) has a variable clinical picture, it is considered a relatively benign hemoglobinopathy. However, high altitude exposure and multiple comorbid conditions have been documented in the literature as causes of splenic sequestration and infarction in this population. Acute splenic sequestration crisis (ASSC) manifesting in adults with Hb S/β has only been described in a handful of case reports. We report a case of a middle-aged female living at low altitude with Hb S/β and a surgical history of recent laparoscopic cholecystectomy. Subsequent hospitalization demonstrated splenic artery thrombosis and splenic infarction likely caused by a hypoxemic event triggering ASSC.

A 47-year-old-female with a past medical history of mitral valve prolapse repair, symptomatic cholelithiasis status post-laparoscopic cholecystectomy with intraoperative cholangiogram and Hb S/β presented to the emergency department on postoperative day (POD) 8 with a complaint of left upper quadrant abdominal pain. She reported persistent abdominal pain and constipation since surgery; however, the pain was increasing in severity. A review of systems demonstrated nausea without vomiting. Her vital signs were within normal limits at the time of initial exam, with an oral temperature of 36.9°C, blood pressure of 112/72 mm Hg, heart rate of 90 bpm, and a body mass index of 32.5 kg/m^2. A physical exam of the abdomen revealed a soft abdomen with moderate tenderness to palpation in the epigastric region and left upper quadrant.

ere was no rigidity, guarding, or peritoneal signs on exam, and the 3cMo9t-l8o43112.551it3cMo9t 0 019112.551it

have utilized a continuous pulse-oximeter to elucidate that hypoxemia rates of surgical patients in the operating room and immediate postoperative period are approximately 7.9%. Of patients who did experience hypoxemic events, 70% of these events persisted longer than two minutes.[4,13] Additionally, a 2010 study concluded that rates of intraoperative hypoxemia and body mass index were directly proportional.[14,15] Our patient's body mass index of 32.5 kg/m^2 classi ed her as obese and rendered her more susceptible to hypoxemia during surgery. Our patient had been NPO for nine hours before surgery and had no documented preoperative episodes of hypoxemia. e surgery lasted two hours and 18 minutes due to a di cult intraoperative cholangiogram and pneumoperitoneum was set at 15 mmHg during the case. She subsequently experienced a hypoxemic event in the immediate postoperative period with an oxygen desaturation to 90% requiring 6L of supplemental O2. We postulate that our patient's splenic infarction was triggered by hypoxic conditions during the perioperative period of her laparoscopic surgery. Her leukocytosis, thrombocytosis, and computed tomography imaging moreover supports this hypothesis. While our patient had not had any previous sickling episodes or history of deep vein thrombosis, she had several additional prothrombotic risk factors including previous tobacco use as a former smoker and obesity. Furthermore, the hemoglobin electrophoresis completed on hospital day one showed that her HbS was 67.3%, a notable increase from her previous HbS of 55.4% just one year prior. e only limitation to this observation is a lack of documented hemoglobin electrophoresis immediately prior to the operation. As this was the rst occurrence of ASSC in our patient, she did not undergo splenectomy. Following the recommendations outlined in the literature, we suggest appropriate immunizations after the rst episode of ASSC and future elective splenectomy for Hb S/β patients due to the high risk of recurrence and associated complications.

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