A 16-year-old female with known homozygous sickle cell anemia presented to the emergency department (ED) with fever, cough, and pain. Five days prior to presentation she was in her normal state of health when she developed a non-productive cough. One day prior to admission she developed pain consistent with prior vaso-occlusive crises in her legs, back, and abdomen. Her medical history was significant for acute chest syndrome and osteomyelitis. On examination she was febrile and mildly tachycardic with normal blood pressure, respiratory rate, and oxygen saturation. Her physical examination was significant for decreased breath sounds and crackles at her right lung base. No hepatosplenomegaly or point tenderness was appreciated on her abdominal exam. A chest X-ray done in the ED showed a right lower-lobe opacity. Her hemoglobin was found to be 6.6 g/dL, which was slightly below her baseline of 7 g/dL to 8 g/dL. Complete metabolic panel done at the time was unremarkable. She was given intravenous fluids, antibiotics, and pain medication and admitted to the hematology service for further management.

Hospital Day 1: The patient was started on continuous morphine with patient-controlled analgesia for pain, and antibiotics were continued. A respiratory viral panel sent from the ED was positive for parainfluenza. Repeat liver function tests (LFTs) were done secondary to patient reports of right upper quadrant pain and were again found to be unremarkable.

Hospital Day 2: A routine morning complete blood count showed marked elevation in the patient’s white blood count to 62,000. Her hemoglobin dropped to 4.3 g/dL and her reticulocyte count was 4.3% (reference range, 0.5%-1.5%). Due to concern for sepsis, blood cultures were redrawn and antibiotic coverage was expanded. Packed red blood cells (pRBCs) were ordered, but matched units were not immediately available due to the patient’s history of multiple antibodies. The patient became tachycardic and hypotensive and was transferred to the pediatric intensive care unit. The patient stabilized after receiving a simple blood transfusion, and an exchange transfusion was planned for the following morning when a sufficient quantity of cross-matched blood would be available.

Hospital Day 3: Prior to exchange transfusion, the patient again became hypotensive and repeat hemoglobin was 3 g/dL with 3.1% retic count. The one available unit of matched pRBCs was given stat. She was electively intubated secondary to worsening acute chest
syndrome (see Figure 1, page 448) and concern for inability to maintain a clear airway while receiving sedation for central line placement. After intubation, the patient became acutely bradycardic with poor perfusion and progressed to cardiac arrest. Due to the patient’s life-threatening condition, unmatched pRBCs were infused as cross-matched blood was still not available. Spontaneous rhythm returned after resuscitation and the patient demonstrated improved perfusion. Significant abdominal distension was observed during the resuscitation attempts, and exam revealed a palpable liver edge 14 cm to 15 cm below the costal margin vertically and to the right mid-clavicular line horizontally. An abdominal radiograph was obtained (see Figure 2). LFTs done at that time showed a total bilirubin of 3.7 mg/dL, aspartate aminotransferase of 59 IU/L, and alanine aminotransferase of 21 IU/L. Over the course of the patient’s decompensation and resuscitation, 5 units of unmatched pRBCs were given, with a resulting hemoglobin level of 10 g/dL.

**Diagnosis:**

**Acute Hepatic Sequestration**

**DISCUSSION**

The patient described in this case study was diagnosed clinically with acute hepatic sequestration (AHS) given her: 1) precipitous drop in hemoglobin, 2) acute hepatic enlargement, and 3) relative stability of LFTs. These three markers are the hallmarks of AHS. Liver biopsy was not done due to high risk of hemorrhage and death, but AHS is thought to be caused by obstruction of sinusoidal flow by masses of sickled erythrocytes. This patient’s cardiac arrest is likely attributed to a combination of 1) reduced cardiac output secondary to impaired venous return as a result of significant hepatomegaly and positive pressure ventilation, and 2) severe anemia.

To the best of our knowledge, there are only 12 reported cases in the published literature of AHS, with only four of those reporting AHS in patients younger than 21 years. More commonly in pediatrics we monitor for splenic sequestration, which is thought to have a similar pathophysiology to AHS; nevertheless, this case report demonstrates that AHS is not solely an adult complication of sickle cell disease. In the few pediatric and adult documented cases, both vaso-occlusive pain crisis and infection appear to be common preceding symptoms. Our patient’s blood cultures were never positive for any pathogen, but she was treated early with broad-spectrum antibiotics. Of note, a respiratory viral panel was found to be positive for parainfluenza. This likely contributed to her acute chest syndrome and caused an overall increase in sickling, but the degree
that it affected her AHS is not completely known. Lastly, the patient was at high risk for a complication of AHS known as reverse sequestration (or auto-transfusion). As the liver begins to release the sequestered blood, hyperviscosity syndrome may result with volume overload, heart failure, and intracerebral hemorrhage. She did not suffer these complications and at this point has not had any further recurrence of hepatic sequestration. Management of AHS is similar to that of splenic sequestration (ie, exchange transfusion to reduce or eliminate sickled RBCs, adequate fluid resuscitation, and pain control).

CONCLUSION

AHS is a rare clinical manifestation of sickle cell disease in the pediatric population, but it is a potentially lethal complication. About 10% of patients admitted for sickle cell disease will have hepatic complications. Although there are fewer than five reported pediatric cases documented in the literature, the consequences of failing to recognize rare occurrences of AHS can be fatal. A thorough abdominal exam and early recognition of hepatomegaly is crucial in every patient presenting with sickle cell complications.

REFERENCES