Rapid Fire: Sickle Cell Disease

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KEYWORDS

• Sickle cell disease • Acute chest syndrome • Aplastic crisis • Splenic sequestration • Sickle cell anemia

KEY POINTS

- Sickle cell emergencies are most commonly related to acute anemia or vasoocclusive crisis.
- The cause of acute anemia must be differentiated between hemolytic, aplastic, or sequestration to determine treatment.
- Acute chest syndrome is the leading cause of morbidity and mortality in sickle cell disease. Early recognition and treatment are key.
- Sickle cell vasoocclusive diseases have many other presentations that require knowledge of presentation patterns and specific treatments.

CASE: FLULIKE SYMPTOMS

Pertinent history: A 29-year-old man with a history of sickle cell disease (SCD) presents to the emergency department (ED) with concerns over flulike symptoms. He works as a school teacher, and over the past week several of his students have developed influenza. Three days ago he developed some increasing soreness in his arms and legs. He first thought this was his sickle cell pain and treated it with his oxycodone that he is prescribed. However, over the past 2 days he has developed increasing cough productive of green sputum and chills. He reports pain in his chest, which he attributes to coughing spells. The chest pain causes him to have difficulty taking a deep breath. Today he developed shortness of breath at rest and decided to seek emergency care. He denies a sore throat, rhinorrhea, headache, or abdominal pain.

PHM: SCD.

Medications: hydroxyurea, oxycodone.

SH: occasional marijuana use, last 1 week ago, denies other drugs/tobacco/alcohol use.

Pertinent physical examination: Blood pressure 172/84, pulse 115, temperature 100.7°F, RR 26, peripheral capillary oxygen saturation 89%

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General: awake and alert; noted mild respiratory distress
HEENT: equal, round, pupils reactive to light; tympanic membranes normal bilaterally; nasal turbinates mildly edematous; normal oropharynx
Neck: supple, no JVD
Cardiovascular: tachycardic with regular rate; no murmur appreciated
Pulmonary: rales to right lower lung field; splinting with deep inspiration
Abdominal: abdomen soft and nondistended; bowel sounds normal; no hernias or masses appreciated
Neurologic: alert and oriented × 3; nonfocal throughout
Musculoskeletal: compartments soft, though pain from palpation in all 4 extremities; radial/ulnar and dorsal pedis pulses 3+ bilateral; normal strength and sensation × 4

Diagnostic findings	
WBC	15
HgB	9
Platelets	300
Reticulocyte count	3%
Na	138
K	3.9
Cl	100
CO_2	24
Glucose	128
ALT	28
AST	30
Bilirubin	1.1

Abbreviations: ALT, alanine aminotransferase; AST, aspartate aminotransferase; Cl, chloride; CO_2, carbon dioxide; HgB, hemoglobin; K, potassium; Na, sodium; WBC, white blood cell.

Chest radiograph (CXR): new right lower lobe infiltrate as compared with previous CXR; no pneumothorax; cardio-mediastinal silhouette normal in appearance.

Electrocardiogram: sinus tachycardia without acute ST depression or elevation.

Plan: aggressive pain management, gentle hydration, antibiotics, oxygen.

Patient's Course: The patient presented with concerns over catching the flu from his students. He has a history of SCD, and the provider was concerned about the possibility of worsening underlying pathology, such as acute chest syndrome. Vital signs demonstrated fever, tachycardia, and hypoxia. The patient had tachypnea as well, which was thought to be due to splinting from pain. After multiple rapid doses of intravenous (IV) hydromorphone, the patient's pain improved. He was placed on 3 L of oxygen by nasal cannula with improvement of hypoxia. His respiratory splinting decreased and his tachypnea resolved. Respiratory therapy was instructed to start the patient on incentive spirometry every hour. D5 1/4NS was administered at 200 mL/h. His hemoglobin (HgB) returned at 9 mg/dL, which the patient reported as close to his baseline hemoglobulin of 10 mg/dL. Rapid influenza swab was obtained and read as negative. Ceftriaxone and azithromycin were administered because of new infiltrate on CXR. Arterial blood gas (ABG) was obtained showing the following: