EM CASE OF THE WEEK.

BROWARD HEALTH MEDICAL CENTER
DEPARTMENT OF EMERGENCY MEDICINE

BROWARD HEALTH
Care Warriors

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March 2017 | Vol 3 | Issue 30

Emergencies of Sickle Cell Disease

A 20-year-old African American male with past medical history significant for asthma & sickle cell thalassemia presents to the ED with bilateral shoulder pain for the last month that has been worsening in the last week. He has experienced these symptoms prior to this episode 2 years ago when he terminated his hydroxyurea after graduating from the pediatric hematology clinic and states this feels like “sickle pain” and he has ran out of his pain medication 1 week ago. He denies shortness of breath, fever, or recent travel, but admits to “getting over a recent cold” and intermittent smoking. Patient was afebrile at presentation with vitals within normal limits with the exception of mild tachycardia; however, a recheck of vitals reveal a fever of 101.5 F and worsening tachycardia to 122 bpm. On physical exam, patient is in no distress, has mild abdominal tenderness to palpation in RUQ, bilateral elbow pain 5/10 radiating to his shoulders and diminished breath sounds in all lung fields. He has no cough, but complains of worsening abdominal pain with deep inspiration, no rebound, guarding, rigidity, or hepatosplenomegaly. Remainder of physical exam is within normal limits without jaundice or scleral icterus. Which of the following is the most common cause of morbidity and mortality for this patient’s condition?

A. Hyper-coagulable state leading to pulmonary embolism

B. Avascular necrosis and subsequent infection

C. Priapism

D. Auto-splenectomy/ sequestration & infarct

E. Acute chest syndrome

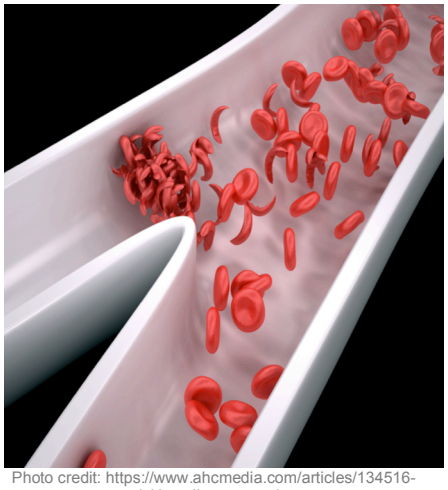

Photo credit: https://www.ahcmedia.com/articles/134516-sickle-cell-emergencies

In patients with SCD, the RBC becomes deformed and forms a characteristic sickle shape causing several complications including vaso-occlusive pain crisis. The pain is caused by small vessel occlusions causing infarction of bone and soft tissues.

Sickle Cell Disease is a very common inherited genetic disorder in the United States. Patients with this disorder, whether within the adult or pediatric populations, tend to frequent acute care centers for evaluation and treatment and require expert assessment to differentiate between urgent treatment for relief of symptoms vs. emergent intervention to minimize life threatening consequences.

EM Case of the Week is a weekly “pop quiz” for ED staff.

The goal is to educate all ED personnel by sharing common pearls and pitfalls involving the care of ED patients. We intend on providing better patient care through better education for our nurses and staff.

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March 2017 | Vol 3 | Issue 30

Acute Chest Syndrome

The correct answer is **E**. Acute Chest Syndrome is the leading cause of death in patients with Sickle Cell Disease (uptodate.com)

Acute Chest Syndromes is the most serious complication of SCD and can lead to fatality, therefore requiring prompt evaluation and management.

It is defined as *any new infiltrate on CXR* + 1 out of any of the following associated new findings:
-Fever >/= 101.4 F
-Cough
-Wheezing
-Tachypnea
-Chest Pain

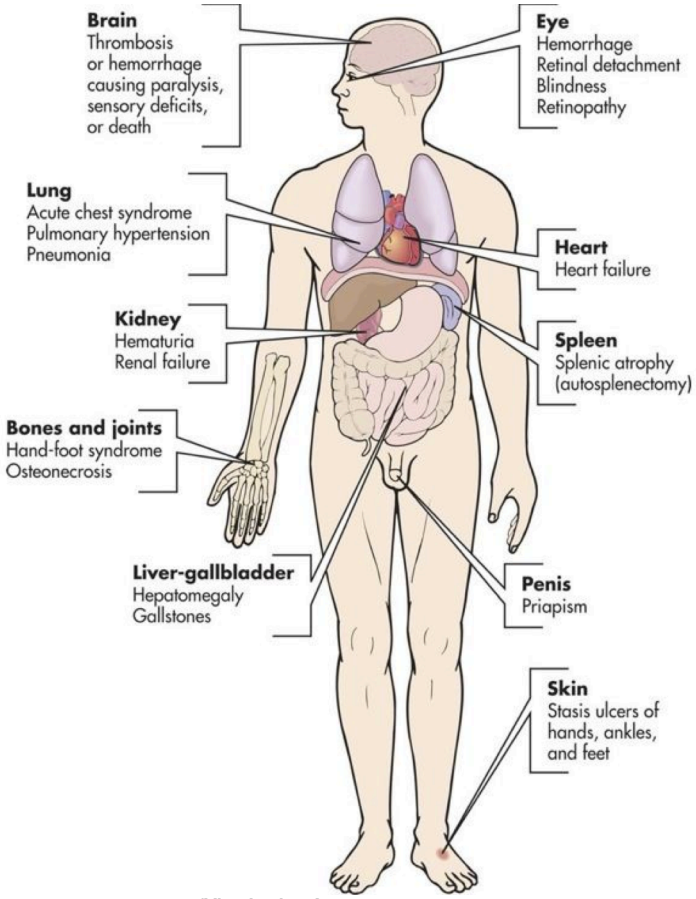
Although most commonly occurring in the pediatric population (ages 2-4yo), Acute Chest Syndrome is most lethal in the ADULT population as the underlying pathology is most critical in adult patients who develop Acute Chest (thrombosis in adults, as opposed to infectious in children).

Discussion:

As most of us in healthcare have experienced, the frequency with which we see sickle cell patients can be desensitizing to our clinical judgment and sometimes even make us question the patient's compliance with simple steps they can take to evade and minimize their own morbidity and symptoms; however, despite this fallacy, vigilance needs be employed to prevent the serious and life threatening complications that such patients are susceptible to.

In fact this propensity for such patients to visit us so frequently should heighten our suspicion rule out the serious complications of the Sickle Cell phenotype and furthermore to treat them with the kindness, care, and extra caution needed to prevent them from spiraling down into such life-threatening problems when they present with pain crisis rather than becoming a routine therapy.

Needless to say Sickle Cell is a painful condition that these patients are born into and must deal with chronically life-long, they deserve dignity, respect, and our empathy no matter how frequently they present.



(Via: Jordan Anne https://s-media-cache-ak0.pinimg.com/564x/32/51/39/32513920e9228b82c253ab99b638b6d9.jpg)

Treatment:

Patients with the Sickle Cell genotype have varying phenotypic expression causing a wide range of presentations. Therefore treatment must be targeted at the level of their particular presentation and underlying condition causing the crisis in addition to the standard of care for a pain crisis.

The following is a broad, general overview and what to look for when examining the various end-organ sites where sickle cell complications routinely occur. These organs are where we require a high index of suspicion with prompt activation of high-level triage to exclude end organ damage prior to the baseline standard of care for a sickle pain crisis.

For a list of educational lectures, grand rounds, workshops, and didactics please visit ***BrowardER.com*** and **click** on the ***"Conference"*** **link**.

All are welcome to attend!

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- **Stroke.** A stroke can occur if sickled cells blocks blood flow to any area of the brain. *Look out for:* seizures, weakness, numbness, sudden speech difficulty, and loss of consciousness.
- **Acute chest syndrome.** This life-threatening complication of sickle cell anemia can be caused by pneumonia or by sickled cells blocking causing dead space in the lungs. *Look out for:* chest pain, abdominal pain, fever, and difficulty breathing, tachypnea, new infiltrate on CXR, wheezing, crackles.
- **Pulmonary hypertension.** People with sickle cell anemia can also develop pulmonary hypertension more commonly in adults than children. *Look out for:* shortness of breath, haziness on chest films, non-reassuring vitals.
- **Visceral damage.** Sickled cells blocking circulation causes hypoxia that immediately deprives an organ of blood and thus oxygen in a patient whose blood is already chronically low in oxygen saturation. Chronic deprivation of oxygen-rich blood can damage nerves and organs commonly the kidneys, liver and spleen and is cumulative. *Look out for:* otherwise unexplained blood chemistry, non-specific inflammatory markers, and LFT abnormalities and visceral pain disproportionate to exam.
- **Blindness.** Capillaries that supply the eyes can get occluded by sickled cells. Over time, this can damage the retina irreversibly leading to blindness. *Look out for:* visual changes, a blurry or narrowing visual field, headache/ dizziness, diplopia.
- **Skin ulcers.** Sickle cell anemia can cause open sores or ulcers, on any area of the skin but commonly the legs. *Look out for:* bed sores and stages of ulceration, these patients spend a lot of time fatigued and bed-ridden due to pain which further increases their chances of bed-sores.
- **Gallstones.** The breakdown of red blood cells produces bilirubin, a high level of which can lead to gallstones and prominently a chronic state of high biliary sludge. *Look out for:* acute RUQ abdominal pain particularly after a fatty meal since the fat content leads to contraction of the gallbladder exacerbating the symptom.
- **Priapism.** Men with sickle cell anemia may experience painful, long-lasting erections when sickled cells block the venous plexus of the penis which if untreated can damage the penis permanently. *Look out for:* priapism in patients with history of sickle cell and ensure this is treated more emergently than in patients without SCD as more invasive treatment, surgical management, and overall resistance to medical management tends to be higher in these patients.

Take Home Points

- There are many potential complications of sickle cell disease, but most start primarily as an occlusive crisis that leads to infarct of bone or tissue, with the site of occlusion and time before treatment determining the severity of complication.
- Although the sites can include any organ most notably the brain, spleen, lung, intestines, femoral head, and reproductive organs, complications in the lung are the most serious as this is the most common and immediately vital to life.
- In acute cases of pain crisis, we must take extra care to make sure acute chest syndrome is ruled out first and more serious complications are not present before simple treatment with fluids and pain control for the acute pain crisis.
- Standard of care includes maintenance IV fluids, pain control tailored to the patient, low threshold for antibiotics with any signs of infection, PRBC or exchange transfusions as needed for more serious cases, and of course treatment of the underlying stressor which initiated the pain crisis.

ABOUT THE AUTHOR

This month's case was written by Farhad Evoghlian. Farhad is a 4th year medical student from NSU-COM. He did his emergency medicine rotation at BHMC in December 2016. Farhad plans on pursuing a career in Pediatric Emergency Medicine after graduation.

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