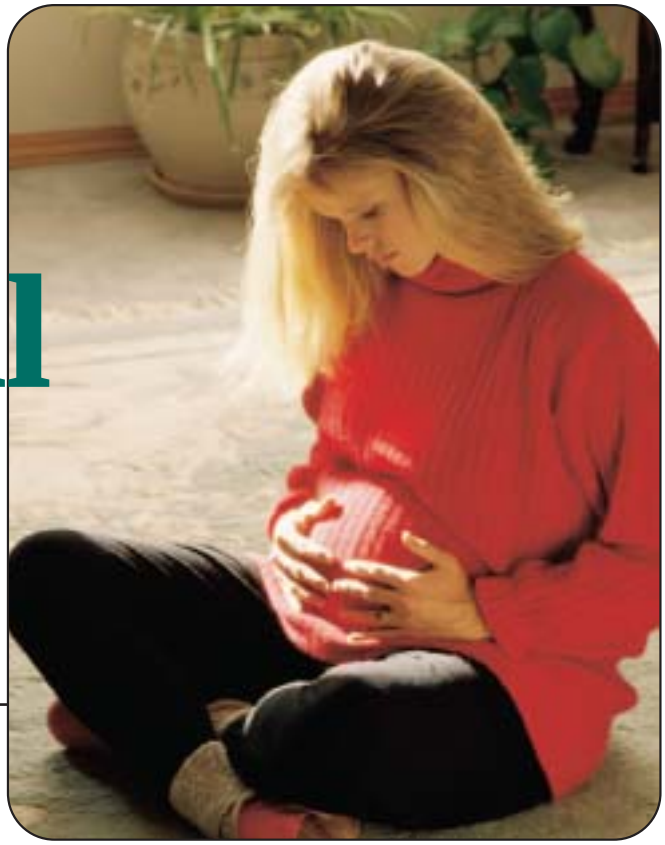
The Pregnant Patient with Sickle-Cell Disease



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The pulmonary consultant was asked to see a 31-year-old pregnant woman — 35 weeks gestation with dyspnea, productive cough and desaturation. The patient had a history of sickle-cell disease (SCD), with approximately four crises in the past year. She had presented three days earlier complaining of a productive cough of two weeks duration with yellowish-white phlegm. Despite a recent course of oral azithromycin, her symptoms persisted. One day prior to admission, she developed bilateral knee pain. She was admitted with a diagnosis of sickle-cell crisis and treated accordingly. Her cough and dyspnea continued to progress and she began to desaturate to 83% on room air while in hospital.

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Physical examination demonstrated a tachypneic 31-year-old woman breathing at a rate of 20 breaths per minute. Her blood pressure was 150/80mmHg and she had a pulse of 80 bpm. She was not cyanosed. Auscultation revealed bilateral end expiratory wheezing. Her jugular venous pressure was slightly elevated and she had normal heart sounds. Her extremities were normal on exam.

The patient's laboratory tests revealed hemoglobin of 70 g/L and a white blood cell (WBC) count of 23. Her reticulocyte count was elevated (contributing to the elevated WBC count). Her chest X-ray was unremarkable.

She had been initially treated with oxygen, hydration, pain control and broad spectrum antibiotics consisting of ticarcillin/clavulanic acid.

Questions

1) What is the differential diagnosis?
2) How would you proceed?

In this particular case, the differential diagnosis includes all of the pulmonary complications of SCD, in addition to causes secondary to preganacy.

Due to the history of presentation, it was believed her symptoms were most compatible with an acute chest syndrome (ACS) of SCD. Although pneumonia was pos-

Pregnancy-related causes of dyspnea

- Physiologic dyspnea of pregnancy
- Cardiac pulmonary edema
- Thromboembolic disease
- Asthma exacerbations (in known asthmatics)

Pulmonary complications of sickle-cell disease

- Infections/pneumonia
- Pulmonary infarct secondary to *in-situ* thrombosis
- Fat emboli
- Acute chest syndrome
- Chronic pulmonary disease

sible, it was believed to be unlikely due to the normal chest X-ray. Thromboembolic disease of pregnancy was more difficult to differentiate from ACS. The pulmonary findings of a pulmonary embolus (PE) can resemble an ACS. Similar abnormalities can also be seen in a ventilation-perfusion scan with both a PE and ACS. A leg duplex was performed and found to be normal. Thromboembolic disease of pregnancy was, therefore, believed to be unlikely. The patient was diagnosed with ACS.

ACS is the most common pulmonary complication of SCD. It is the leading cause of death among SCD patinets, accounting for 25% of all deaths. Patients usually present with fever, pleuritic chest pain, dyspnea, leukocytosis and new infiltrates on chest X-ray. Although the pathogenesis is not completely understood, there are numerous causes. The most common cause is pulmonary infarction. The infectious causes include bacterial, chlamydia, mycoplasma, legionella and viral.

Treatment is supportive for the ACS patient, with oxygen, pain control and hydration. Broad spectrum antibiotics, such as a macrolide, should be given. Bronchodilator treatments should be commenced and blood transfusions may also be indicated. D_x

References:
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2. Vichinsky EP, Neumayr LD, Earles AN, et al: Causes and outcomes of the acute chest syndrome in sickle-cell disease. N Engl J Med 2000; 342:1855-65.

Causes of acute chest syndrome

- Pulmonary infarction secondary to *in-situ* thrombosis
- Fat emboli from bone infarcts
- Infectious etiology