

Location: Office

Vital signs:

T.38.9° C (102F), HR 96/min, RR 21/min and BP 118/65 mm Hg.

C.C: Chest pain

HPI:

A 7-year-old African American child who has a history of sickle cell disease is brought to the office by his mother with a one-day history of right arm pain, chest pain, fever, non-productive cough, and mild shortness of breath. He describe the left sided chest pain, 8-9 /10, constant, aggravated by deep breath, but not relieved by anything. Child denies having had any kind of injury. Patient is on prophylactic penicillin, and had received polyvalent pneumococcal vaccine. His past medical history is significant for sickle cell anemia.

Developmental history: Age Appropriate per Denver Developmental growth scale.

Family History: Sickle cell disease in maternal cousin. No other major medical problems in the family.

Social history: Lives with mom, dad and older sister. No smokers at home. All: None. Meds: Penicillin VK and folic acid. How do you approach this patient?

Order physical exam:

General

HEENT/Neck

Lungs

Heart

Abdomen

Extremities

Skin

Results:

The 7-year-old child is in moderate to severe pain, and mild respiratory distress. Pallor is present. Severe local tenderness is noted over the right arm, ribs and sternum. Heart sounds are normal. No murmurs/rubs/gallops. There are decreased breath sounds, and increased tactile vocal fremitus (TVF) present over the right middle lobe.

Order review:

Pulse oximetry, stat

Results:

88% on room air

Order review:

Admit the patient to floor/ward

Oxygen, inhalation (to keep pulse ox greater than 93%)

IV access

CBC with diff, stat

BMP, stat

Blood cultures

Urinalysis and urine cultures

Sputum gram stain and culture

Chest X-ray, PA, and lateral, stat

EKG, 12 lead

Type and screen for blood

Vitals Q 4 hours

Pulse oximetry, continuous (or) Q1-2 hours depending on the severity of hypoxia

Regular diet

IV hypotonic fluids D5 1/4 NSS or D5W NS, bolus and then continuous

IV Morphine, continuous
IV Cefuroxime and IV Azithromycin
Incentive spirometry
Albuterol nebulization, Q4 hours
Consider blood transfusion if hypoxia does not improve on nasal canula

Next:

Advance the clock to see the next available result
Reexamine the patient in 1 hour and take interim history and focused lung exam

Order:

Repeat hemoglobin and hematocrit in 4 hours and again in 8 hours if stable
Patient will say that he is feeling much better
Stop IV fluids next day
Repeat CBC with diff, and BMP next day

*Usually the case will end if you do this much.

Discharge:

Observe the patient for 48-hours, stop IV antibiotics once the cultures are negative and discharge him to home on oral antibiotics
Patient education
High calorie diet

Discussion:

Acute and chronic pulmonary complications are the most common cause of death in patients with sickle cell anemia. Recurrent microvascular obstruction leading to pulmonary hypertension and pulmonary fibrosis is the underlying pathology. The primary acute pulmonary diseases seen in patients with SCD include:

1. Acute chest syndrome
2. Infection or pneumonia
3. Infarction due to in-situ thrombosis
4. Fat embolism from bone marrow infarction

Differentiation of pneumonia from acute chest syndrome is difficult, as the sputum and blood cultures are often nondiagnostic.

Microvascular occlusion resulting in bone marrow infarction can cause pulmonary fat embolism (PFE). Sickle cell disease (SCD) patients who have PFE manifest altered mental status, thrombocytopenia, petechial rash, falling hematocrit, and severe hypoxemia. The therapeutic options for PFE include exchange transfusion, plasma infusions, and glucocorticoids.

Acute chest syndrome is the most common form of acute pulmonary complication in patients with SCD. Intravascular sickling resulting in macrovascular or microvascular infarction within the pulmonary vasculature is assumed to be the most frequent cause of ACS in patients with SCD. Previous studies have shown that upto 50% of ACS is the result of bacterial infection. ACS should be suspected if the patient presents with chest pain/extremity pain, fever of $>38.5^{\circ}\text{C}$, tachypnea, wheezing, or cough. On examination, local tenderness over the ribs or sternum and signs of consolidation are present. Leukocytosis, thrombocytopenia or thrombocytosis, falling hemoglobin concentration, elevations in lactate dehydrogenase and bilirubin levels, presence of a new pulmonary infiltrate, involving at least one complete lung segment (not atelectasis) are the other common findings.

The acute management of ACS is primarily a supportive care:

1. Patient should be given supplemental oxygen to prevent further sickling
2. Pain should be controlled with judicious use of narcotics
3. Dehydration should be corrected with volume replacement
4. Incentive spirometry can be used to prevent atelectasis. Some SCD patients may manifest airway hyperreactivity, and these should be given bronchodilators.
5. Antibiotics to cover *S. pneumoniae*, *H. influenzae* type b, *Mycoplasma pneumoniae*, and

Chlamydia pneumoniae.

6. Exchange transfusion is indicated for patients with hypoxia with new infiltrates.

Chronic therapy:

1. Penicillin prophylaxis with penicillin V potassium 125 mg BID is recommended in children aged 4 months to at least 3 years.
2. Polyvalent pneumococcal vaccine for both adults and children.
3. Hydroxyurea for recurrent vasoocclusive crisis.

Primary Diagnosis:

Sickle cell Anemia

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