Sickle Cell Crisis

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Sickle Cell Disease

- an autosomal recessive genetic disease of Hb synthesis
- result of a single–amino acid substitution in the β-globin chain of the Hb molecule, specifically, valine for glutamate

Sickle Cell **Crisis**

Rapid diagnosis and treatment are necessary to minimize morbidity and mortality.

Case 1

- a 16-year-old boy with a history of SCD presented to the ED with a 3-day history of fever, cough, and SOB.
- Five days prior, he had been evaluated and treated for severe pain in his legs and arms.
- He complained of persistent and worsening pain in both his lower extremities and pain in his chest, in spite of oral narcotic therapy.

Case 1

- His medical history included multiple, vasoocclusive, painful crises, including an episode of priapism, and he had received multiple blood transfusions over his lifetime.

Case 1

On examination

- temp 39.1°C, diaphoretic, and uncomfortable.
- HR of 80, BP 116/84 mm Hg, RR 26
- O2 Sat 89% and improved to 94% with 6 L/min via face mask.
- conjunctivae were icteric
- mucous membranes were moist

Case 1

- Cardiovascular II/VI systolic ejection murmur.
- labored respiration with suprasternal and intercostal retractions.
- decreased breath sounds in the right midzone and lower zone, and scattered crepitations on the right side.
- no lower extremity edema
- Abdominal examination Normal
- CNS Normal

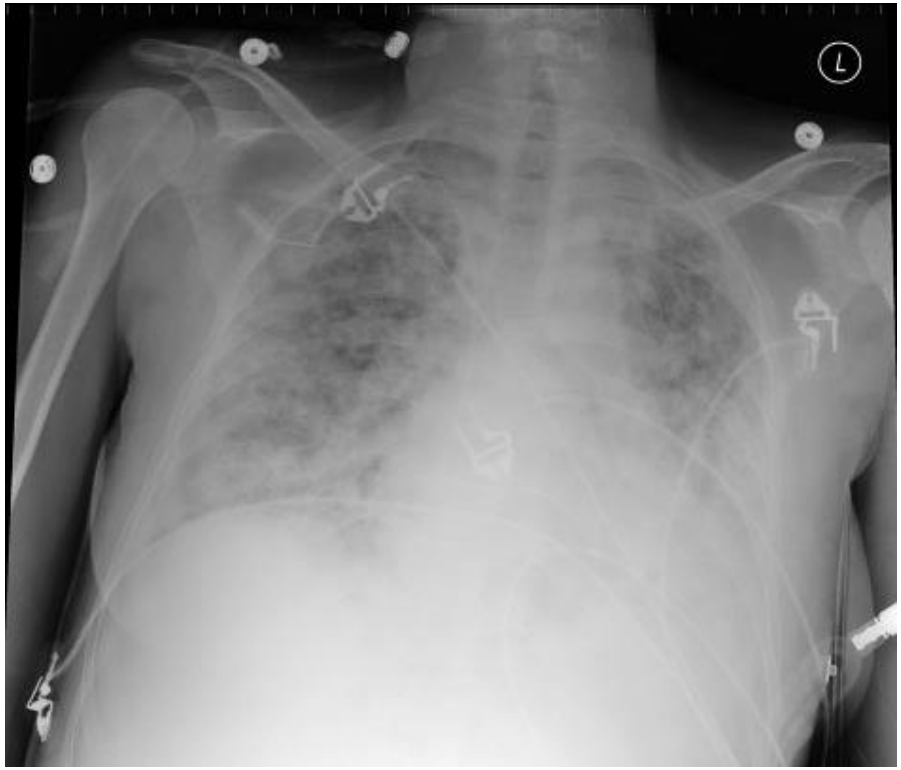
Case 1

- WBC of 17 500/µL
  - 62% neutrophils
  - 25% lymphocytes
  - 9% monocytes
  - 2% eosinophils
  - 1% basophils
  - 1% atypical lymphocytes.
- Hb was 8 g/dL
- reticulocyte 25%
- platelet 206 000/µL.

ABG on room air
- PO2 59 mm Hg
- PCO2 29 mm Hg
- pH 7.32
- HCO3 13 mmol

➢A chest x-ray right lower-lobe consolidation with a moderate right pleural effusion.

Case 1



Case 1

In the ED, he received

- antipyretics
- supplemental oxygen
- cefotaxime 2 g IV
- packed red blood cell transfusion was initiated after 20 mL/kg of normal saline was infused.

Case 1

Over the next hour while waiting for ICU bed:

the patient's oxygen saturation continued to worsen, and he was hypoxic even on supplemental oxygen of 12 L/min via nonrebreather mask.

Case 1

- He underwent emergency intubation using RSI
- A diagnostic pleural tap was performed which demonstrated an exudative fluid.
- The resulting Gram stain and culture were negative.

Case 1

What is it

DDX

- TRALI
- ACS
- Pneumonia

Acute Chest Syndrome

- Serious and life-threatening complications

- Leading cause of mortality and morbidity after the effective antimicrobials and the pneumococcal vaccine

Acute Chest Syndrome

- caused by a vasoocclusive crisis of pulmonary vasculature.

- not distinguishable from pneumonia

Acute Chest Syndrome

Dx

New infiltrate on CXR in combination with at least 1 clinical sign or symptom

- Chest pain
- Cough
- Wheezing
- Tachypnea
- Fever

- Fever and cough are common in children
- chest pain, sob, and chills are common in adults.

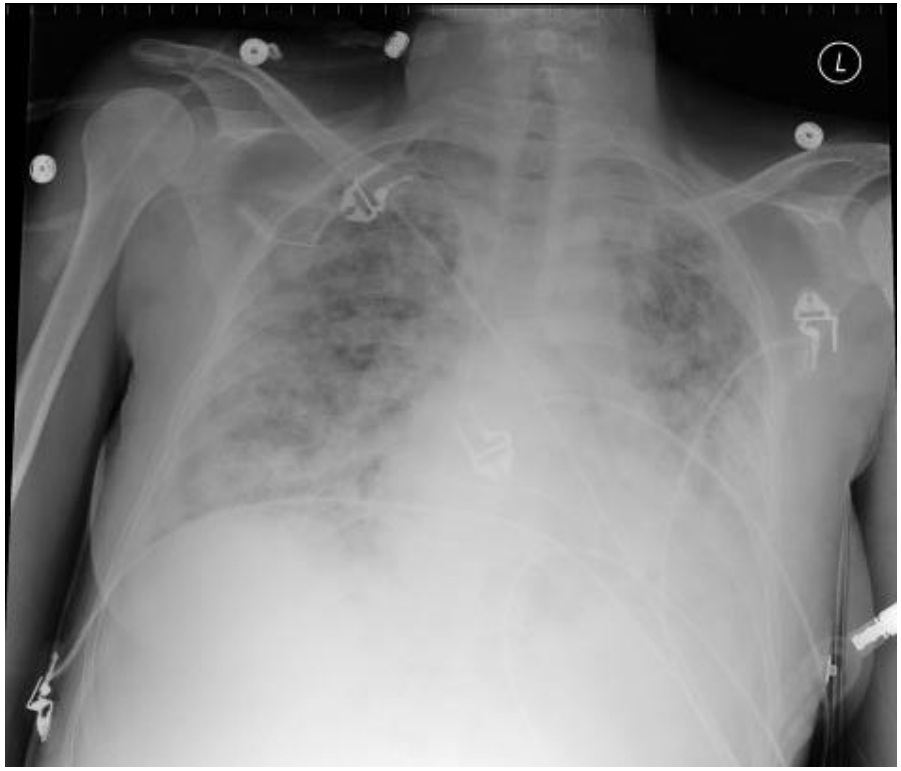
Acute Chest Syndrome

What is the most common physical examination?

- Rales is the most common physical examination
- Normal examination is the second most likely

Acute Chest Syndrome

- The radiographic findings can lag behind the clinical findings



Multilobar disease

Acute Chest Syndrome

Causes

Common causes

Pulmonary infection

Fat emboli

Rib infarction

Possible causes

- Thromboemboli
- In situ thrombosis
- Iatrogenic
  excessive hydration or narcotic use

Vichinsky EP, Neumayr LD, Earles AN, et al. Causes and outcomes of the acute chest syndrome in sickle cell disease. National Acute Chest Syndrome Study Group N Engl J Med 2000;342:1855–65.

Acute Chest Syndrome

What are the diagnostic testing in Acute Chest Syndrome?

- Serial CXR
- Ventilation perfusion imaging
- Serial CBC & Reticulocyte
- Blood gas
- Blood C/S

Acute Chest Syndrome

Therapeutic Modalities

Supportive measures

- Oxygen for hypoxia
- Appropriate hydration
- Appropriate pain control

Incentive spirometry

The incidence of infiltrates and atelectasis is reduced in patients who receive incentive spirometry

Antibiotics:

Third-generation cephalosporin + macrolides

Transfusion therapy

- Simple transfusion
- Exchange transfusion

Experimental therapy
Nitric oxide
Corticosteroids

Acute Chest Syndrome

Transfusion Therapy

Reports of dramatic improvement in clinical condition after initiation of transfusion.

Bodo I, Khoury H, Blinder M. Rapid resolution of the acute chest syndrome of sickle cell disease after automated red cell exchange. Blood 1997;90 Suppl 1:23b

Acute Chest Syndrome

Therapeutic Modalities

- *Mycoplasma pneumoniae* more commonly associated with acute chest syndrome.
- Aggressive broad antibiotic coverage is necessary.
- Treatment with an antibiotic from the macrolide class, as well as a third-generation cephalosporin,is recommended

Septicemia

- SCD pts have impaired immunologic function that is caused by splenic dysfunction.

- Impairment of splenic function can occur in infants as young as 3 months.

Septicemia

High risk for encapsulated organisms such as *S pneumoniae* and *H influenzae*.

Septicemia

Recommended antibiotic

- Third-generation cephalosporin; ceftriaxone, or cefotaxime

- Vancomycin should be added to protect against penicillin-resistant strains of *S pneumoniae* if suspected until culture results become available.

Septicemia

In summary

All SCD patients with fever must be managed with extreme caution because of the risk of overwhelming bacteremia which can rapidly lead to septic shock.

Stroke or CVA

- major complication of SCD
- is a leading cause of disability children and adults

Stroke or CVA

- Commonly caused by blockage of the intracranial internal carotid and middle cerebral arteries.

- Patients usually present with obvious signs such as
  - acute hemiparesis
  - aphasia or dysphasia
  - seizures
  - severe headaches
  - cranial nerve palsy
  - altered mental status

- The most common tends to be hemiparesis.

Stroke or CVA

- Presentation can be very subtle, such as a slight limp.

- Need a high index of suspicion for stroke in a patient with any new neurological finding on physical examination.

Stroke or CVA

Initial therapy is immediate 1.5- to 2-fold volume exchange transfusion in an ICU setting to reduce Hb S to less than 30% of total Hb.

Stroke or CVA

- After acute clearance of symptoms should be started on a long-term transfusion therapy.

- If not on a long-term transfusion program have an 80% chance of recurrent stroke within 3 years of the initial event

Stroke or CVA

- Long-term transfusion involves regularly scheduled blood transfusions aimed at reducing the percentage of Hb S and not at normalizing the Hb level.

Case 2

- A 44-year-old diabetic presented to the ED complaining of nonexertional sob and severe back pain for 12
- The patient reported malaise, fatigue, weakness that started 3 days before, chronic blurred vision, insomnia, and anxiety.
- The remainder of the review of systems was unremarkable.

Case 2

O/E

- HR 101 bpm
- RR 31/min
- Temp 37C
- BP 148/62 mm Hg
- O2 sat on RA 99%.
- The patient was awake, alert, and oriented
- He was motionless to avoid back pain.

Case 2

O/E

- Normal S1 and S2
- Chest Normal
- Strength was 4/5 in all 4 extremities.
- Deep tendon reflexes were normoactive.
- Normal flexor plantar response was obtained, and no meningismus

Case 2

- WBC 11.2 × 109/L (with no abnormalities in differential count)
- hg of 9.4 g/dL
- hct of 26.3%
- mcv of 76.7 Femtoliters (fL)
- Mch 27.3 pg
- platelets of 144 × 109/L.

Case 2

- Blood glucose 267 mg/dL

- AST of 79 U/L
- ALT 30 of U/L
- ALK 475 U/L
- total bilirubin level of 2.3 mg/dL
- direct bilirubin level of 0.8 mg/dL

Case 2

- ESR 54 mm/h
- ECG Normal
- MRI of the lumbar spine was Normal

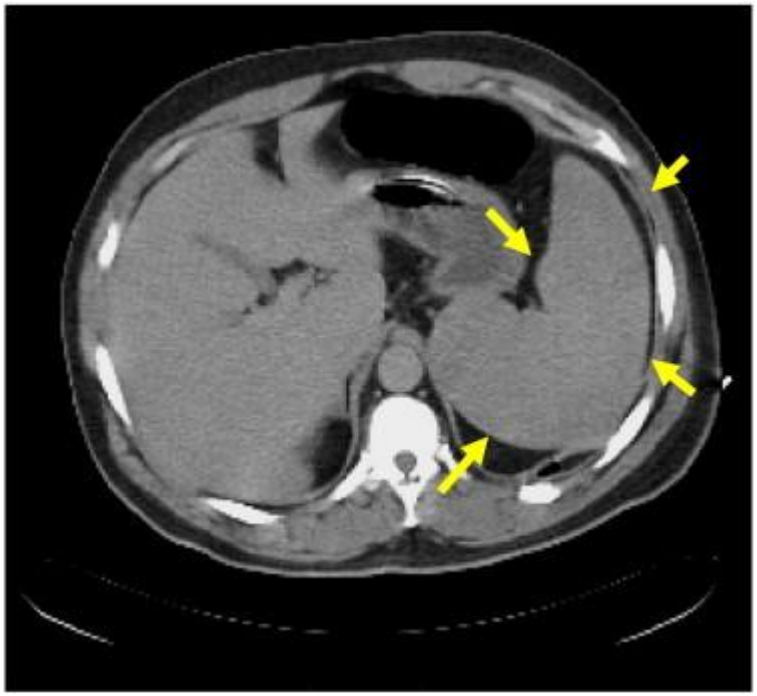
Case 2

- Despite aggressive narcotic treatment of back pain, the pain continued to increase

- 1 hour later hypotension of 90/50 mm Hg.

Case 2

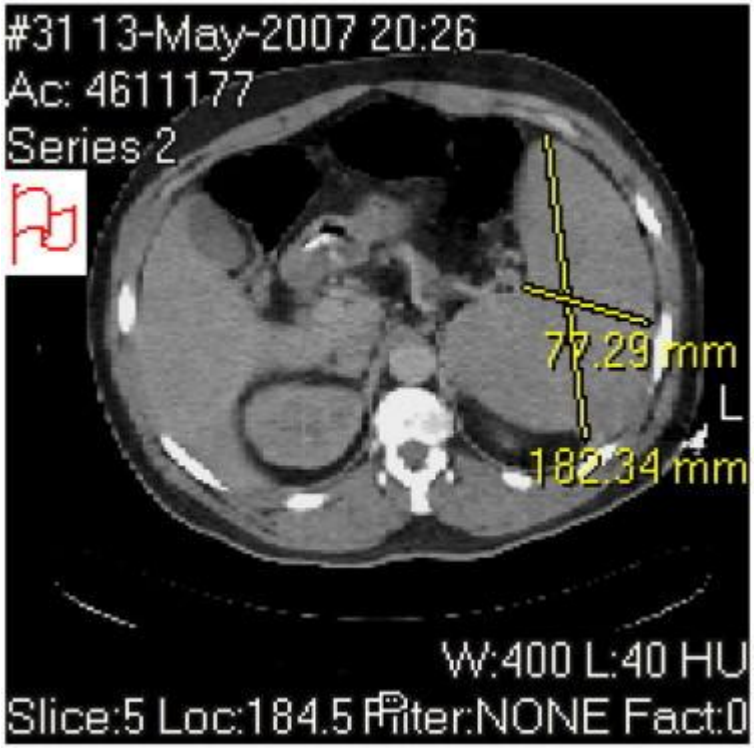
1st CT



CT abdomen was ordered, revealing an enlarged spleen

Case 2

2nd CT



The new CT scan of the abdomen revealed an increasing splenomegaly compared with the previous one

Case 2

- Despite transfusion therapy with 4 u prbc, the patient's hg progressively dropped to a level of less than 4 mg/dL over the course of 3 hours, with thrombocytopenia (<50 × 109/L).
- immediately transferred to an ICU.
- altered mental status and worsening gas exchange.
- Airway protection with intubation and mechanical ventilation were initiated.

Case 2

- As the patient was rapidly deteriorating, an emergent splenectomy was performed,
- the patient recovered every organ function and, 6 months later, has resumed his normal activities.

Acute splenic sequestration crisis (ASSC)

- sudden impounding of red blood cells by the spleen
- characterized by
  - rapid fall in hemoglobin concentration
  - rise in reticulocyte count
  - and splenomegaly
- requires prompt recognition and treatment.
- In the adult patient, ASSC is extremely rare.

Acute splenic sequestration crisis (ASSC)

- Hypotension caused by large volumes of blood (mainly sickled cells) entrapped in the spleen.
- Hb levels may fall acutely more than 2 g/dL less than the patient's normal value, causing circulatory compromise

Acute splenic sequestration crisis (ASSC)

- Prompt diagnosis and therapy with red blood cell transfusions are therefore crucial to prevent hypovolemic shock.
- Surgical splenectomy may be indicated in certain patients to prevent recurrences

Aplastic Crisis

- caused by temporary cessation of red cell production with a corresponding decrease in the reticulocyte count.
- approximately 80%, are thought to be caused by human parvovirus B19

Aplastic Crisis

- Diagnosis is made by comparing baseline blood and reticulocyte counts to those obtained during the acute illness.

Aplastic Crisis

- Therapy is initiated if the patient is symptomatic, that is, tachypnea, tachycardia, or hypoxia.
- Therapy includes simple blood transfusion to raise serum Hb back to the patient's baseline and to prevent heart failure secondary to severe anemia.

Aplastic Crisis

Because parvovirus B19 is contagious, affected persons should be isolated from

- pregnant women
- immunocompromised patients
- chronic illness pts

Priapism

- painful prolonged erection of the penis
- caused by sickling of the red blood cells producing venous stasis in the erectile tissue of the penis.
- The resulting stasis causes ischemia, hypoxia, and pain.

Priapism

- Initial treatment involves intravenous hydration and analgesia.

- Episodes refractory to this initial management include direct irrigation of the corporeal bodies of the penis

Osteomyelitis

- most commonly caused by *Salmonella* species or *Staphylococcus aureus*
- Bone pain or joint pain with localized swelling and decreased range of motion, along with fever, should alert the physician to the possibility of osteomyelitis.

Osteomyelitis

- Increased white blood cell count and elevated ESR are common laboratory findings.
- Patients with suspected infections with these organisms should be started on the appropriate broad-spectrum antibiotic and have diagnostic imaging performed to confirm the diagnosis.

VasoOcclusive pain Crises

most common symptoms of SCD

- severe pain
- localized ischemia.

Triggers

- dehydration, fever, cold exposure, and emotional stress

Therapy

- intravenous/oral hydration
- pain management
- It is useful to assess pain in a standard manner using pain measurement scales

Sickle Cell Crisis

Summary

- Acute Chest Syndrome
- Septicemia
- Stroke or CVA
- Acute splenic sequestration crisis (ASSC)
- Aplastic Crisis
- VasoOcclusive pain Crises
- Osteomyelitis