Splenic Infarct in a Sickle Cell Trait Patient

Grand Rounds
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Case - History

• CC: L flank pain
• HPI: 70 yo F w SOB and worsening L flank pain of 3 days duration. C/o fever, weakness, N/V/D; vomitus – nonbilious/nonbloody.
• PMH: HTN, sickle cell trait (recently diagnosed)
Meds: HCTZ, Diovan, Ranitidine, Calcium, Folic Acid

SH: Pt had taken a flight from Louisiana 7 days prior to presentation.

ROS: Exercise tolerance significantly diminished – can barely walk to bathroom.
Case - PE

- T 102.7, HR 100, BP 190/70, RR 40, SaO2 99%
- AAOx3, mod distress
- Dry oral mucosa
- Tachycardic, no murmurs
- Tachypneic, labored, diminished bilat bases
- Soft, left flank tenderness, no rebound or guarding
- L CVA tenderness
Case - Labs

Ca 8.4

38.9 7.7 112
20.6 80%

137 100 22
3.2 26 1.1
171

5.9 49 60
3.3 19 4.0

33.1 58
2.7

Trop 0.03
Lipase 6.0
Case - Operative Course

- Taken to the OR for emergent splenectomy.
- Operative finding notable for omentum adhesed to LUQ; splenomegaly with patchy areas of necrosis.
- EBL 2 L.
- Transfused 6 units PRBC.
Case - Normal Spleen
Case - Histology
Case - Postoperative Course

- Tachypnea - resolved after drainage of L pleural effusion.
- Received vaccinations for H. flu, pneumococcus, and meningococcus.
- Received ASA for Plt count > 1M.
- Discharged to rehab in stable condition.
Sickle cell disease

- Homozygous for Hb S allele (Hb SS)
- Change in AA composition of the β globin chain of hemoglobin (glutamic acid replaced by valine)
- AA change secondary to nucleotide substitution at codon 6 of the hemoglobin gene (GAG to GTG)
Sickle cell trait - Epidemiology

- Inheritance of one Hb S allele (Hb AS)
  - Hb A 60%; Hb S 40%
- Appx 8% of African Americans
- Est 300 million worldwide
- Est 3 million in the US
- Normal lifespans
Sickle cell trait - Associated Complications

- Increased rates of UTI
- Gross hematuria
- Splenic infarction with altitude hypoxia or exercise
- Exertional rhabdomyolysis
- Renal failure
- Glaucoma
- Recurrent hyphema
- Sudden death
Sickle cell trait - Pathophysiology

- Subclinical tissue infarction from microvascular obstruction
- Sickling occurs at 15 torr PO2
### Splenic Infarction in Sickle Cell Trait

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<thead>
<tr>
<th>Title</th>
<th>Year</th>
<th>Authors</th>
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<tbody>
<tr>
<td>Altitude-related specific infarction in sickle cell trait--case reports of a father and son</td>
<td>1985</td>
<td>Goldberg, et al.</td>
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<td>Sickle cell trait in a white Jewish family presenting as splenic infarction at high altitude</td>
<td>1988</td>
<td>Shalev, et al.</td>
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<td>Splenic infarct in a white man with sickle cell trait</td>
<td>1982</td>
<td>Cox</td>
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<td>Splenic infarction in a white man with sickle cell trait</td>
<td>1979</td>
<td>Diep, et al.</td>
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<tr>
<td>Splenic syndrome in sickle cell trait: four case presentations and a review of the literature</td>
<td>1999</td>
<td>Franklin, et al.</td>
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<td>Splenic syndrome at mountain altitudes in sickle cell trait</td>
<td>1985</td>
<td>Lane, et al.</td>
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Splenic Infarction in Sickle Cell Trait

- Spleen most consistently injured organ in sickle cell trait patients.
- S/Sx included LUQ pain, N/V, L pleural effusion, L lung atelectasis, fever.
- Elevated LDH, leukocytosis.
- CT scan consistent with contained hemorrhage.
- Usually self-limited, resolving in 10-21 days.
- Rarely requires surgery.
Splenectomy - Indications

• Autoimmune and idiopathic
  – Idiopathic thrombocytopenic purpura
  – Thrombotic thrombocytopenic purpura
  – Idiopathic autoimmune hemolytic anemia
  – Felty’s syndrome
  – Sarcoidosis

• Cell membrane disorders
  – Hereditary elliptocytosis
  – Hereditary pyropoikilocytosis
  – Hereditary hydrocytosis
  – Hereditary xerocytosis
• Genetic deficiencies
  – Thalassemia
  – Sickle cell anemia
  – Gaucher’s disease
  – Pyruvate kinase deficiency
  – Glucose-6-phosphate dehydrogenase deficiency

• Disorders of WBC origin
  – Hodgkin’s lymphoma
  – Non-Hodgkin’s lymphoma
  – Chronic myelogenous leukemia
  – Hairy cell leukemia

• Disorders of bone marrow
  – Myelofibrosis of myeloid metaplasia
  – Myeloproliferative disorders
Overwhelming Post-splenectomy Infection (OPSI)

- Lifetime risk: <1% to 5%
- Mortality: approaches 50%
- Caused by encapsulated organisms
  - Pneumococcus - 50%-90%
  - Meningococcus, H.flu type B, Grp A strep
- Vaccinations
  - Ideally >2 weeks prior to elective splenectomy
  - Just prior to discharge for non-elective splenectomy
Take Aways

• Sickle cell trait patients have subclinical disease which can manifest in extreme environmental conditions.

• Splenic infarction in these patients are usually self-limited; indications for surgery include sepsis, hemodynamic compromise.

• Splenectomy requires knowledge of surrounding anatomy.