Indications for Splenectomy in Patients with Hematologic Disorders

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Case presentation

12 yom with three day history of abdominal pain, fevers and AMS

PMHx: Sickle cell requiring transfusions q2 weeks; CVA, developmental delay, seizure

PSHx: denies

Meds: miralax, folic acid, exjade, hydroxyurea, oxcarbazepine, keppra, clobazam

Allergies: Ceftriaxone
Case presentation

Vitals: 101 116/76 149 30 99%

PE:
- Lethargic, nonverbal
- Tachypneic, tachycardic
- Abd: rigid, splenomegaly, diffusely tender

Labs:
- Chem: 139/3.6/111/18/15/0.24/119
- CBC 22>7.8/22.9<154 BASELINE:6.7/20.9
- LA 2.5->0.8
CTAP
Case Presentation

U/S: hepatomegaly, cholelithiasis, enlarged spleen with no flow

CTAP: Enlarged spleen 14.2 cm with poor enhancement, cholelithiasis, hepatomegaly 14 cm, ascites
Plan

IVF resuscitation
Foley insertion
Preparation of RBC
OR for exploratory laparotomy, splenectomy
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Operative Course

Culture of fibropurulent ascites

Omentectomy, distal pancreatectomy, splenectomy, diaphragm repair

EBL 50

Transfused 300 cc pRBC
Post Op Course

POD 0: Chest tube insertion

POD 1:
- Febrile, tachycardic
- 1 unit pRBC transfused

POD 2:
- Extubated,
- mucous plug of RUL

POD 3-
- CT removed
- Rx for pneumonia begun
Post Op Course

POD 5 -10
- Bowel function
- Seizure activity
- NGT removed and PO feeds initiated

POD 11
- DC JP and patient

Plan for vaccinations and antibiotic rx in PCP office

All Cx negative; path necrotic spleen
Questions?

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Indications for splenectomy

- Erythrocyte disorders
- Platelet disorders
- Bone marrow disorders
- Cysts, abscess
- Cancers
Splenectomy

Wait until age 5 in children in possible
Vaccinate prior if possible
Before you operate

Get hematology team on board

Oxygenate, hydrate, prepare transfusion

http://www.downstatesurgery.org/files/cases/sc.pdf
Membrane defects

Spherocytosis
- Ankyrin, spectrin, band 3 protein defects -> increase susceptibility to destruction
- anemia, when severe do splenectomy (children in hb 6-8 range)
- CURRATIVE

Eliptocytosis

ovalocytosis
Sickle Cell Disease

Autosomal recessive substitution of valine for glutamine in B chain

More susceptible to oxidative stress leading to deformed RBC

Spleen goes from splenomegaly to atrophy within first decade of life

Unless..

◦ Splenic sequestration
Splenic Sequestration

Splenic sequestration: red blood cells become entrapped in spleen causing splenomegaly and anemia of 2 gram drop

Acute vs chronic
- Incidence 7-30%
- 2nd most common cause of death in first decade
- Abdominal pain, distension, pallor, weakness, tachycardia
Sickle Cell Disease

Indications for splenectomy
- Persistent splenomegaly
- Sequestration
- Infarction
- Splenic abscess
- Developmental delay and delayed growth
Splenectomy vs. Conservative management for acute sequestration crises in people with sickle cell disease

Cochrane review

Limited to case studies, no RCT trials

Outweigh risks vs. benefits.
Overwhelming post splenectomy infection

Typically encapsulated organisms

Usually within first 3-4 years

In trauma less then 1%, in hematologic disorders 15-20%

High mortality
Thalassemia

Autosomal recessive resulting in defective alpha and beta globulin subunits
  ◦ Abnormal shaped RBC destroyed by the spleen

May require chronic transfusion and iron chelators

Splenectomy:
  ◦ Painful hypersplenism
  ◦ Transfusions>250ml/kg/year
Autoimmune Hemolytic Anemia

Antibodies to RBC antigens
- Shortened red blood cell survival
- Positive Combs test

Rx: STEROIDS to decrease autoantibody formation
- Splenectomy in warm reactive antibodies only if:
  - Failed high dose steroids
    - 20% cured
    - 50% have decrease in steroid requirement
Immune/Idiopathic thrombocytopenic Purpura ITP

Most common indication for elective splenectomy

Autoantibodies to platelets and removal in the RES

Treatment based on bleeding and platelet count

- Corticosteroids works 20%, high remission rate 60-80%
- IV Ig, immunosuppressives (rituximab, cyclophosphamide, azathioprine)
- Failure of medical management or persistent platelet count <10000-> splenectomy
  - Long term response 60%
Thrombotic Thrombocytopenic Purpura

Deficiency of metalloproteinase that cleaves von Willebrands resulting in platelet clumping

Treatment
- Plasma exchange
- Splenectomy 8-20% relapse
Other platelet and WBC disorders

- Essential thrombocythemia
- Polycythemia vera
- Primary myelofibrosis
- CML
- CLL
- HD, NHL – for staging
Conclusion

Hematologic disorder
- Splenomegaly
- Anemia requiring excessive transfusions
- Warm AIHA
references

Cameron  11th edition Edited by  John L Cameron, Andrew M Cameron. 2014.


Shirley Owusu-Ofori, Tracey Remmington. Splenectomy versus conservative management for acute sequestration crises in people with sickle cell disease. Cochrane review. 2015. Issue 9, Art No CD003425


Questions