Vesalius SCALpel™ : Spleen (see also: abdomen => spleen folios)

Anatomy

15-30% accessory spleens: 80% hilum, splenocolic ligament, omentum, pelvis, small bowel mesentery (scrotum male)
white zone: lymphatic nodules & germinal centers
red zone: thin walled sinuses, sinusoids, clear old, damaged RBCs
4-5 discrete vascular territories
marginal zone: interface
splenic cyst: true or post-traumatic (pseudocyst)(75%):
- 25% unknown cause
- LUQ pain, US, CT: symptomatic: aspirate (recurrence, bleeding), unroof, partial splenectomy
- congenital: epidermal, simple
- parasitic: echinococcal most common; liver > lung > spleen in frequency
- neoplastic: resect
- dermoid: 3 layers, rarest, congenital, resect
- neoplastic, echinococcal and dermoid resect; congenital and traumatic if symptomatic
- no percutaneous aspiration, can bleed, recur

Splenic artery aneurysm

3rd most common abdominal aneurysm after aorta, iliac
CT scan Dx
most near tail of pancreas, multiparous female, 5-10% incidence rupture
- indications for surgery:
  - > 2cm asymptomatic
  - all pregnant women or of childbearing age
    - rupture during pregnancy maternal and fetal mortality 80%
- if proximal can ligate proximally and distally, if distal may require splenectomy
- embolization option: splenic infarction very painful

Function

clear abnormal RBCs, platelets, cellular debris
- RBC life cycle normally ~120d, 20cc removed daily by spleen
  - RBC membrane defect: sperocytosis (splenectomy), elliptocytosis
  - sickle cell autosplenectomy; increased risk of infection
- should not see Howell-Jolly (residual nuclear chromatin), Heinz or Pappenheimer bodies with normally functioning spleen, cleared by spleen
  - if splenectomy is complete (no accessory) will see the above on smear; if not still have residual splenic tissue, accessory spleen
    - miss rate for accessory spleen lap-scope = open
- platelets 10d lifespan, 1/3 of platelets pooled in spleen, can rise to 80% with splenomegaly
- wbc 6h half life

abnormal function splenic antibodies (IgM) cause excess cell destruction, bind to platelets

opsonins
- properdin: initiates alternate pathway of complement activation
- tuftsin: binds granulocytes to promote phagocytosis

ITP

Primary or secondary
- Dx of exclusion:
  - SLE, antiphospholipid syndrome, immunodeficiency, lymphoproliferative, HIV, hepatitis C, heparin, drug related antibodies (quinidine), thyroid disease

peripheral smear to R/O pseudothrombocytopeniaa, inherited giant platelet syndrome & other hematological disorders

immune disorder with IgG antibodies, F:M 3:1
- IgG antiglobulin on platelets verifies
- can follow upper respiratory infection
- spleen is the major source of IgG in ITP, increased 5-6X
- antibodies bind to platelets which are destroyed by spleen
- platelets <50k, normal bone marrow
  - 30-50K bruising
  - 10-30K spontaneous ecchymosis
  - <10K internal bleeding

bleed: vaginal, mucosal, UGI, nose

usually sporadic, increasing with AIDS, SLE

spleen rarely palpable. if palpable consider Dx of hypersplenism

50% of cases children ~5yo, M=F

child good prognosis, 80% recovery without treatment < 6mo (usually within a few wks), do not do splenectomy, rare (<1%) intracranial hemorrhage

adults require Rx at presentation, 50% platelets < 10K

treatment
- initial trial of steroids, 1mg prednisone/kg/d
  - 3-6w, if responds wean, 50-75% success
  - initial response to steroids suggests good response to splenectomy
  - IV immune globulin for internal bleeding, platelets < 5K despite steroids, extensive progressive purpura
    - 80% respond, but common relapse
  - if no response, requirement for high dose (10-20mg/d) or recurrent drop platelets:
    - elective splenectomy, 85% success
- splenectomy
  - surgery 92% response v 30% medical Rx, surgery treatment of choice
  - predictors of response to surgery
    - young age, most common positive predictor
    - short interval diagnosis to surgery
    - initial response to steroids
    - HIV+

high pre-op platelets
40K platelets OK to proceed
don't give platelets unless untoward bleeding post op
look for accessory spleen: 30% in hematological disorder v 20%
plasmapheresis takes 4-5d for response, not beneficial in acute crisis
emergency splenectomy only for neurologic crisis (intracranial bleed, pl < 10K)
recurrence
accessory spleen, observe
search for other causes of thrombocytopenia: hemolytic disorder, thyroid, pregnancy, infection
moderate, 40-50K try cytoxan, steroids, plasma exchange
persistent: reoperate after CT, US, Tc scan looking for accessory spleen; gamma probe with indium labeled platelets

TTP

autoimmune response to endothelial cell antigen (arterioles, small capillaries)
disease of arteries with diffuse platelet trapping in small vessels (arterioles and capillaries)
platelet aggregation, hyaline deposits in/under endothelium
normal size spleen
pentad: fever, purpura (thrombocytopenia), hemolytic anemia, neurologic abnormalities, renal failure
profound thrombocytopenia, elevated WBC, elevated bilirubin (hemolysis), hematuria, protein casts
peak 20-30yo, F>M
may be initiated by viral or bacterial infection, pregnancy, drugs (BCP)
untreated 10% 1y survival
Rx steroids, plasmapheresis, rarely splenectomy

Hereditary spherocytosis

autosomal dominant, defect in RBC membrane structural protein spectrin
shortened RBC lifespan, osmotic fragility
anemia, reticulocytosis, jaundice, splenomegaly, pigment gallstones (94% by 13)
increased osmotic fragility, high sequestration, destruction by spleen
wait until at least age 4 before splenectomy, cholecystectomy at same time

Sickle cell disease

HbA replaced by HbS (valine substitution for glutamic acid 6th position on beta chain of Hb)
with decreased O_2, RBC elongates and distorts causing increased viscosity, stasis, crenation, clotting, worsening hypoxemia, cycle
mortality from recurrent infection, renal failure, heart failure
spleen autoinfarcts, rarely need splenectomy
Howell-Jolly bodies seen
splenic infarction may result in splenic abscess

Thallasemia

autosomal dominant defect in Hb synthesis
- presents early in life
- persistence of HbF (fetal) and decreased HbA

accumulation of intracellular material causes sturctureal abnormality RBC
need recurrent transfusion, can't maintain Hb > 10
- need for splenectomy
- high risk OPSS

Primary hypersplenism : rarely responds to steroids

Secondary hypersplenism

- portal hypertension may result in splenic enlargement, anemia, leucopenia, thrombocytopenia
- treat portal hypertension, no need for splenectomy

Splenic vein thrombosis

- acute or chronic pancreatitis, pancreatic tumor
- splenic enlargement, trapping
- normal liver (thrombosis may propogate into portal vein)
- isolated gastric varices, no esophageal varices
  - gastric varices lower incidence of bleeding than esophageal, not amenable to banding
  - splenectomy cures gastric varices and hypersplenism

Acquired immune hemolytic anemia: medical Rx 1st

Hairy cell leukemia: high recurrence after splenectomy; now treated with alpha2 interferon

Portal vein thrombosis

- hypercoagulable state, stasis, (long splenic v stump?)
- abdominal pain 1-2w post splenectomy, may result in dead gut
- anticoagulate, thrombolytic: heparin to coumidin X 6mo

Felty's syndrome

- rheumatoid arthritis, neutropenia, recurrent leg infection/ulcer, splenomegaly
- splenectomy may be beneficial if medical Rx fails

Sarcoidosis

- lung and liver disease, 20% incidence splenomegaly
- splenectomy may be beneficial

Gaucher's

disorder of lipid metabolism
splenectomy may be beneficial

Myeloid metaplasia

progressive marrow fibrosis
peripheral extramedullary hematopoeisis
immature precursors in peripheral blood
highest incidence of portal vein thrombosis
splenectomy

Splenic abscess

chills, fever, LUQ tenderness, splenomegaly
contiguous spread, hematogenous spread, immunocompromise, (sickle infarct)
splenic enterococcus abscess may seed diseased mitral valve
staph, salmonella, e.coli, enterococcus (ICU pts), fungus
salmonella increased in ICU pts, sickle cell disease, typhoid, immunocompromised
poultry, turtle sources
Dx: US, CT
percutaneous drainage 20-30% success for unilocular
splenectomy may be necessary for multiple

Trauma

penetrating LUQ with intraabdominal bleeding requires surgrery
blunt
FAST exam has replace peritoneal lavage, go to CT scan if positive or suspicious for injury
CT grading more accurate in pediatric than adult
grade
I < 10% of surface, < 1cm deep
II non-expanding subcapsular hematoma 10-50% of surface, non-expanding
intraparenchymal hematoma < 2cm, bleeding capsular tear or parenchymal
laceration 1-3cm deep without trabecular vessel involvement
III expanding subcapsular or intraparenchymal hematoma, bleeding subcapsular
hematoma > 50% of surface, intraparenchymal hematoma > 2cm,
parenchymal laceration > 3cm deep or trabecular vessel involvement
IV ruptured intraparenchymal hematoma with active bleeding, laceration involving
segmental or hilar vessels resulting in major (> 25% of volume)
devascularization
V completely shattered or avulsed, hilar laceration with total devascularization
non-op management (grade I-III)

stable patient, grade I-III, ability to do serial exams (even on vent), <2U blood loss related to spleen (v pelvic, femur fx)
low failure rate (80% of blunt trauma, 90% success)
incidence of missed injuries ~2%
failure: hemodynamic decompensation, new or increased abdominal pain (other visceral injury), dropping Hct
contrast blush on angio indicates active bleeding, poor prognosis, to OR
24h ICU observation, 3-4d bed rest, minimal activity 1-2w, no contact sports 3 mo no need for CT or US f/u
indications for splenectomy: hemodynamic instability, peritoneal signs, ongoing blood loss

splenorrhaphy (those who are candidates with isolated splenic injury don't go to the operating room anymore)
blood loss <500cc, minimal associated injuries, no hilar involvement, minimal-moderate splenic disruption, normal coag, no associated injuries
suture, cautery, surgical, hemostatic glue, partial splenectomy, mesh wrap

Spontaneous rupture

malaria most common cause worldwide, mono in US
sarcoid, leukemia, delayed rupture from blunt

Overwhelming post-splenectomy sepsis (OPSS)

avoid splenectomy < 4
highest risk 1st 2y post splenectomy (60% of adult cases, 80% of child)
the earlier the infection, the higher the mortality
risk varies with indication for splenectomy
accessory spleen not enough to confer immunity
greatest risk children, less common in adults
hematologic disease higher risk than trauma
highest risk thallasemia, lymphoma, Hodgkins, don't do as well
strep pneumonia (70%), h. flu, neisseria (encapsulated BT)
pneumococcal vaccine (covers 73% of strains, 40% of strains penicillin resistant), h. flu, n. meningitis vaccines
give early, 2w before elective, otherwise prior to discharge, revaccinate @5y
prophylactic antibiotics 6mo-1y, penn, amoxicillin, erythromycin
child with febrile illness after splenectomy take to ER