Vesalius SCALpel™ : Liver

Physiology

alk phos from ductal membrane, increased in obstruction
fasting glucose from gluconeogenesis from alanine and lactate
    not from glycolysis
Kupfer cells/RES (liver, spleen, lung) clear debris & bacteria; stain cytokeratin
most coagulation proteins from liver, VonWillibrand's from vascular endothelium
fat soluble vitamins require bile salts for absorption
    vit D hydroxylated in liver, to kidney to active form
    XS vit A hepatotoxic
AST/SGOT, ALT/SGPT, LDH measure hepatocyte function
alk phos, 5'nucleotidase, leucine aminotransferase, GGT measure excretory capacity
    increased in obstruction
need > 20% of liver to support life

Abscess

pyogenic (80%): fever, RUQ pain, jaundice, pruritis, (palpable liver?), WBC, sepsis
    biliary more common than appendicitis, diverticulitis sources
    iatrogenic: stent, manipulation
    30% of unknown etiology
    40% one organism, 40% polymicrobial, 20% sterile
    systemic source: subacute bacterial endocarditis, catheter (staph, strep)
    abdominal source: Gm+ and neg. aerobes and anaerobes: klebsiella, strep, e. coli,
        staph, pseudomonas, anaerobes (bacteroides)
    Rx: antibiotics and percutaneous drainage (85% successful)
amebic (20%): pain, abdominal tenderness, (hepatomegaly), diarrhea
    5% of patients with intestinal amebiasis develop amebic liver abscess
        ingest to cecum, venous to liver, lung
    anchovy paste liver abscess, usually solitary, R lobe
    Dx: stool for ova and parasites, serology
    Rx: 75% cure w drugs: flagyl, chloroquine
        aspirate, drain
        surgery for rupture, lo mort, 25%
fungal:
    immunosupressed, prone to liver/spleen candida abscess
    Rx fungicidal drugs

Simple cyst

most common cystic disease of liver, 5% of pop, 50% single, asymptomatic
    symptomatic F>M, age <50: unroof only if absolutely necessary
    can percutaneously aspirate, sclerose w alcohol (but not if bile stained cyst fluid: can
        enter and sclerose biliary system)

Polycyctic liver disease

autosomal dominant, associated with polycystic kidney disease
insidious abdominal distention, early satiety, respiratory compromise, portal hypertension (end stage)
non-op Rx unless very symptomatic
distorted anatomy, may come to transplant
polycystic liver 30% intracranial aneurysm (aneur also associated with FMD)

Traumatic retention cyst/pseudocyst: often resolve, rarely require surgery

Echinococcus (hydatid)

20% ductal communication
jaundice, cholangitis, enzyme elevation, (pre-op ERCP?)
mediterranian, south America, Pacific
intermediate hosts sheep, elk, caribou
to liver via portal system
peritcyst (50% calcified), ectocyst, endocyst
slow progress, enlargement causes pain, mass, jaundice
CT multiloculated, complex, indistinct margins
serologic indirect hemagglutination 90% positive (Cassoni skin test obsolete)
Rx: medical (abendazole, proziquantel) alone only 30% cure
surgery: evacuate, cidal agent into cyst, < 10% recurrence
spill of contents: anaphylactic reaction, spread of daughter scolices
if bile in cyst, no scolicidal instillation, damage ductal system

Cystadenoma

multiloculated, septated, 80% women
can become malignant, require resection

Solid benign lesions: hemangioma, focal nodular hyperplasia, adenoma, bile duct hamartoma

hemangioma:

differentiate from focal nodular hyperplasia and adenoma
spiral CT: relative hypoattenuation, early enhancement periphery, then central complete isoattenuating fill at 3-60min in 50-80%
MRI also highly accurate, 90%, best test
most common benign solid benign liver lesion
young, female, 90% solitary
no malignant potential, rare rupture (risk for trauma with large)
asymptomatic unless enlarge causing compression and pain
observe 90%, very large resect (embolization, radiation little effect)

Kassabach-Merritt syndrome: platelet trapping in large hemangioma
child: 50% associated with cutaneous lesion
most <4cm, asymptomatic until reach 10cm
<3% spontaneous rupture

adenoma

associated with BCP, benign, 30% incidence of hemorrhage, rupture (increased risk >5cm, rapid growth)
pure hepatocytes, no ducts or reticuloendothelial cells, rare malig
difficult to differentiate from FNH: both rapid enhancement and washout on CT/MRI; FNH characterized by central scar
hyperintense T2 weighted images
lack of nucleotide uptake because of absence of Kupfer cells (v FNH)
heterogeneous if bleed within adenoma (10-15%)
incidence related to duration of use of BCP > 2y, may regress w withdrawal of exogenous hormone stimulation
may enlarge and have increased tendency to rupture during pregnancy
surgery may be necessary for symptoms (pain, bleeding), failure to regress
may progress to hepatocellular carcinoma

focal nodular hyperplasia (FNH)

young women, incidental finding, ? local reaction to injury
most single, 15% multiple
well circumscribed, non-encapsulated within normal liver
birth control pills trophic but not causative
contain bile ducts, hot spot on Tc99 scan due to increased Kupffer cells activity
CT/MRI: marked early enhancemen and washout, central scar in each nodule, needle bx if uncertain
Rx: observe, don't grow or bleed, no malignant potential,
prove by bx if necessary
resect for unclear Dx or symptoms
if resected, do not recur

bile duct hamartoma

common multiple small firm gray-white subcapsular nodules

Budd-Chiari

hepatic vein thrombosis or obstruction
50% associated with polycythemia or myeloproliferative disease
oral contraceptives lesser risk
IVC web most common cause in Asia
vague abdominal symptoms and onset of ascites most common presentation

may progress to fulminant liver failure
Rx palliation, relieve hepatic congestion: surgery or interventional radiology
lifelong anticoagulation for hypercoagulable state

Cirrhosis

ascites
spironolactone, lasix
peritoneal-venous shunt
contraindications: variceal bleed (increases vascular volume),
bacterial peritonitis
uncontrolled coagulopathy, CHF

Liver transplant

90% 1y survival, 70% 5y
no liver transplant for metastatic colon cancer

Hepatocellular carcinoma (HCC)

most common primary cancer worldwide
sub-Saharan Africa, Asia 100/100,000 v US 3/100,000
Japan 60% hep C association, Asia hep B
US: older (>50), M:F 8:1, cirrhosis, environmental facrors associated
risk factors: alcohol (cirrhosis), viral hepatitis B, C (increasing in US)(hep A not a risk factor), hemochromatosis (200X)
multiphase CT or MRI with gadolinium: initial low attenuation, bright enhancement with contrast, hypodense on delayed images
alphafetoprotein positive 90% marked elevation (> 400)
no beneficial chemotherapy, frequently (70%) not resectable
extrahepatic nodal disease makes tumor unresectable
highly vascular; US, CT non-specific; MRI shows relationship to vessels
fibrolamellar variant: women, better prognosis, less AFP elevation (10%)
only Rx resection, 30% resectable, 30% 5y survival if resected for cure
no beneficial chemo or radio Rx
transplant candidate: critical location makes unresectable, <5cm, no extrahepatic spread, portal vein open, no nodes
non-operative candidate: radiofrequency ablation (RFA); cryo
RFA: hi freq alternating current causes ions to oscillate generating friction and local heat resulting in coagulation necrosis
vessels < 3mm destroyed or thrombosed
lesion < 2.5cm single electrode
cryo: cycles of rapid cooling produce intracellular ice crystals which destroy organelles and membranes; slow cooling freezes extracellular fluid resulting in loss of intracellular fluid and ions leading to protein denatruation and membrane disruption; increased thermal conductivity of previously

frozen liver enhances effect of subsequent cycles; peripheral lesions <3cm ideal for cryo

Liver transplant for HCC & Cirrhosis

Milan criteria
- one tumor < 5cm
- up to 3 nodules < 3cm

UCSF criteria
- one tumor </= 6.5cm, total size < 8cm

4y survival 70%
consider preop Tace, PEI, RFI

Hepatoblastoma

most common hepatic tumor in children, <3yo
enlarged liver, abdominal swelling, 90% elevated AFP; jaundice rare
normal hepatic function, avascular defect on nuclear scan
rapid progression, prognosis related to histology
surgery only hope for cure; possible benefit of chemo, radioRx

Angiosarcoma

least common liver malignancy, most common mesechymal liver tumor
related to exposure to thorium dioxide, vinyl chloride
adults, rapid progression, no therapy
child: large, bilateral, unresectable at presentation, no effective Rx

Liver metastases

most common liver cancer in the US
80% colorectal > intestine, kidney, adrenal, breast, gastric, ovarian, melanoma
25% of colon cancers have liver mets @ Dx, 25% subsequently develop liver mets
other liver mets rarely isolated
- resection of neuroendocrine mets may help control symptoms
- extrahepatic mets and advanced cirrhosis contraindications to resection

CT/MRI: hypo, iso or hyperdense depending on vascularity
- colorectal relatively avascular
- intraop US most accurate

only 25% of patients eligible for resection are ever referred
number of mets inversely related to survival: 2-4 mets resected w 1cm margin 20%
- increase in 5y survival, isolated colon met(s): proven benefit 25-40% 5y survival
- staged resection for more than one lesion
  - allows regeneration between resections
- portal vein embolization to reduce tumor size

unresected hepatic mets 3-24mo survival

may consider resecting limited (solitary pulmonary) non-nodal extrahepatic mets
alternate Rx: radiofrequency ablation (RFA), hepatic artery infusion chemo

Wilson's disease

Cu deposition
Rx: D-penicillamine, chelates Cu
end stage may require transplant

Alpha-1 antitrypsin deficiency

lung and liver damage

Primary biliary cirrhosis

middle aged women
progressive bile duct destruction by cytotoxic T cells
antimitochondrial antibodies
pruritis without jaundice, indolent
rising bili related to disease progression
may lead to transplant

Primary sclerosing cholangitis

50-66% have associated IBD
fibrotic stricture any part of biliary tree
increased incidence cholangiocarcinoma
rapid progression look for cancer
more variable progression than primary biliary cirrhosis
may also require transplant

References:

Kim R et al. Consensus and controversy in the management of hepatocellular carcinoma. JACS, 201(1), July '07: 108-123.

Aljiffry M et al. Evidence based approach to cholangiocarcinoma: a systematic review of the current literature. JACS, 208(1), Jan. '09: 134-147.

Hammill C, Wong L. Intrahepatic cholangiocarcinoma: a malignancy of increasing importance. JACS, 207(4), Oct. '08: 594-603.

Bockhorn M et al. The role of surgery in caroli's disease. JACS, 202(6), June '06: 928-932.