Liver Imaging: A Case of Cirrhosis and Budd-Chiari

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Agenda

- Patient presentation
- Menu of tests
- Interpretation of studies, part I
- Differential Diagnosis
- Anatomy
- Interpretation of studies, part II
- Diagnosis and outcome
Mr. P is a previously healthy 30-year-old man who presents to the ED

Week-long history of:
- Back pain
- Abdominal distension
- 15-lb weight gain

No significant PMH

ROS significant for insomnia
Menu of Tests

Imaging options to evaluate back pain and abdominal distension:

- **CT**
  - Test of choice
- **MRI**
  - If CT equivocal
- **Ultrasound**
  - If RUQ biliary etiology suggested
- **Plain films**
  - Seldom useful
- **Angiography**
  - No longer used for diagnosis; used for therapeutic interventions
Our Patient: Cirrhosis on Axial Abdominal CT

Star: Ascites

Arrow: Recanalized umbilical vein

Other features of cirrhosis: Enlarged spleen, Enlarged liver caudate and left lobes, atrophied right lobe, heterogeneous parenchyma
Radiographic Appearance of Cirrhosis on CT

- **Change in liver size**
  - Atrophy of right lobe
  - Enlargement of caudate and left lobe
    - Caudate:right lobe > 0.65 is 90% specific for cirrhosis

- **Change in liver contour**
  - Hobnail appearance

- **Associated changes**
  - Ascites
  - Splenomegaly
  - Collateral vessels
Differential Diagnosis

- Alcohol abuse
- Viral hepatitis
- Primary biliary cirrhosis
- Hemachromatosis
- Veno-occlusive disease
  - Budd-Chiari
  - CHF
- Drug toxicity
- Hereditary:
  - Wilson’s, alpha-1 anti-trypsin, metabolic disorders

Our patient had no history of alcohol abuse, no risk factors for hepatitis, and did not have any history of drug intake.
Our Patient: Axial CT Scan

Arrow: Inferior vena cava

The hepatic veins should be seen at this level and are not seen.
Companion Patient #1: Normal Axial Abdominal CT

Hepatic veins are seen at the same level

Yellow arrow: Right hepatic vein

http://radiographics.rsnaajnl.org/cgi/content/full/21/suppl_1/S133/F17A
Differential Diagnosis

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Liver Vascular Anatomy

- **Portal vein**
  - Right and left branches

- **Hepatic veins**
  - Right, left, and middle
  - Combine to form the IVC

http://www.moondragon.org/images2/hepaticanatomy.jpg
Our Patient: Coronal CT Scan

The portal vein appears patent on this coronal CT scan.

Arrow: Main Portal Vein
Menu of Tests, Part II

- Concern is for occlusion of the hepatic veins.
- Imaging options for evaluating hepatic vasculature:
  - Ultrasound with Doppler
  - MR angiogram/venogram
  - Conventional angiogram/venogram
Our Patient: Doppler US

Flow is seen only in the right hepatic vein; the left and middle hepatic veins are not visualized.
Clinical Diagnosis

- Imaging up to this point suggested veno-occlusive disease
  - Budd-Chiari syndrome most likely
    - Occlusion of hepatic veins
- Laboratory studies showed a CBC as follows: Hct 62% Plt 876
  - Suggested Polycythemia Vera
- LFT’s were elevated and rising
- Ammonia level was 88
Budd-Chiari Syndrome

- Thombosis of the hepatic veins and/or intrahepatic or suprahepatic IVC

- Etiology
  - Myeloproliferative disorders
  - Malignancy
  - Infection/liver lesions
  - OCP/pregnancy
  - Hypercoagulable states
    - Factor V Leiden, Prothrombin gene mutation, APLA, Protein C/S deficiency, ATIII deficiency, PNH, nephrotic syndrome
  - Behcet’s syndrome, other autoimmune disorders
  - Idiopathic
Polycythemia Vera

- Myeloproliferative disorder
  - Increased hematocrit, white blood cell count, and platelets

- Mechanism
  - Red blood cell production in the absence of erythropoietin stimulation
  - JAK2 mutation usually responsible

- Can cause clotting and thrombosis due to increased viscosity of blood
Budd-Chiari: Radiologic Appearance

- **Doppler US**
  - Lack of flow in hepatic veins

- **CT with IV contrast**
  - Thrombus may be visible within the hepatic venous system

- **MRV**
  - Reduced caliber or absence of hepatic veins

- **Venography**
  - “Spider web” appearance of the hepatic vasculature
  - Thrombus or occlusion of IVC or hepatic veins
Our Patient: Venogram
Performed during transjugular biopsy of the liver
Our Patient: Digital Subtraction Venogram

Arrow: “Spider-web” hepatic vasculature

Absence of normal hepatic veins with spider web vasculature, characteristic of Budd-Chiari
Budd-Chiari: Treatment

- Thrombolysis
  - Interventional radiology
  - Only if acute thrombus is present
- Angioplasty/Stenting
- Shunt
  - To relieve hepatic venous congestion
- Medical therapy
  - Treat underlying cause
  - Lactulose, beta-blocker, diuretic
- Liver transplant
Our Patient: Diagnosis

- **Diagnosis:**
  - Fulminant hepatic failure, acute on chronic
  - Budd-Chiari syndrome
  - Polycythemia vera

- **Tissue diagnosis:**
  - Liver biopsy
    - Compatible with cirrhosis and Budd-Chiari
  - Bone marrow biopsy
    - Myeloproliferation consistent with polycythemia vera
    - JAK2 mutation present

- **PMH revisited:**
  - Prior records indicated elevated hematocrit as early as 2004
  - Mother had essential thrombocytosis (elevated platelets)
Our Patient: Treatment

- Treatment for Budd-Chiari:
  - Lactulose, Nadolol, Lasix
  - Therapeutic paracentesis

- Treatment for polycythemia vera
  - Therapeutic phlebotomy
  - Hydroxyurea
  - Heparin for anti-coagulation

- Placed on liver transplant list
Thank you!

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The nurses, residents, fellows, and attendings who helped me care for Mr. P during my sub-internship.