AMSER Case of the Month
July 2022

8-year-old female with fatigue, abdominal pain, and jaundice

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Patient Presentation

- **HPI:** 8-year-old female presenting to the pediatric ED with 1 week history of increasing fatigue and intermittent abdominal pain and 2 days of jaundice. Denies nausea/vomiting, recent illness or travel.

- **Past Medical/Surgical History:** Prior tympanostomy tubes, otherwise none.

- **Developmental History:** Normal development.

- **Health Maintenance:** Up to date on routine vaccinations.

- **Physical Exam:** Afebrile, tired appearance, icteric sclera, normal S1/S2, lungs clear to auscultation bilaterally, abdomen soft and nontender without palpable HSM, no peripheral edema, dark urine.
Pertinent Labs

• Hepatic Panel
  • AST - 2410 (H)
  • ALT - 2294 (H)
  • Alk Phos - 290
  • Total Bili - 7.9 (H)
  • Direct Bili - 4.5 (H)

• Coagulation Studies
  • Prothrombin Time - 15.7 (H)

• Additional Hepatic Testing
  • Gamma Glutamyl Transferase - 150 (H)
What Imaging Should We Order?
Select the applicable ACR Appropriateness Criteria

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<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
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<tr>
<td>US abdomen</td>
<td>Usually Appropriate</td>
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<td>CT abdomen with IV contrast</td>
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<td>MRI abdomen without and with IV contrast with MRCP</td>
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<td>MRI abdomen without IV contrast with MRCP</td>
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<td>ERCP</td>
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<td>US abdomen endoscopic</td>
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This imaging modality was ordered by the ER physician.
Findings (unlabeled)
Findings (labeled)

Centrilobular hyperechogenicity/periportal edema with decreased parenchymal echogenicity (classic “starry sky” appearance)

Mild gallbladder wall thickening (~3mm) without evidence of gallstones

Hepatomegaly (upper limit of normal = 12.8cm for patient’s age)
Findings (unlabeled)

Normal study done in an 8 year old boy for screening due to chronic Hep B infection.
Findings (labeled)

Patient

↑Centrilobular hyperechogenicity/periportal edema with decreased parenchymal echogenicity (classic “starry sky” appearance).

Normal study done in an 8 year old boy for screening due to chronic Hep B infection.
Findings (labeled)

Centrilobular hyperechogenicity/periportal edema of the liver on longitudinal gallbladder view

Borderline thickened gallbladder wall (~3mm) without visualized gallstones or sludge
Final Dx:

Acute hepatitis due to hepatocellular injury
Case Discussion

• Pathology:
  • Inflammation of the liver due to injury of hepatocytes – serum AST and ALT markedly elevated due to release from damaged hepatocytes.
  • Alkaline phosphatase would also be elevated in acute hepatitis, if due to cholestasis (i.e. extrahepatic or intrahepatic obstruction).

• Differential Diagnosis:
  • Viral infection, autoimmune etiology, toxic ingestion (i.e. acetaminophen toxicity), hepatic congestion due to poor hepatic venous outflow (i.e. Budd-Chiari Syndrome).

• Clinical Features:
  • Jaundice is evident due to systemic build-up of bilirubin.
  • +/- abdominal pain, nausea, vomiting.
  • Hepatic encephalopathy at presentation is indicative of acute liver failure.
Case Discussion

• Ultrasonographic Findings:
  • Decreased echogenicity of the liver parenchyma occurs due to hepatic edema (“sky”).
  • Relative accentuation in echogenicity of the fibrous walls of the portal veins (“stars”).
    • NOTE: Though the “starry-sky” sonographic appearance initially described by Kurtz, et al. (1980) is a classic finding of acute hepatitis, multiple studies have indicated poor sensitivity and specificity of this finding.
  • Hepatomegaly is most common sonographic finding, and typically measured as liver height >15.5cm at the midclavicular line in adults (reduced cutoff criteria in pediatric patients depending on age, see Konus, et al. [1990]).
  • Mild gallbladder wall thickening (≥ 3mm) without evidence of gallstones or dilatation is often appreciated.
Case Discussion

• Further Workup:
  • Toxicology panel/acetaminophen levels
  • Viral hepatitis serologies (Hep A IgM, HBsAg, anti-HBc, anti-HCV)
  • Autoimmune markers (ANA, anti-dsDNA, anti-smooth muscle antibodies, anti-liver/kidney microsomal antibodies, IgG 1/2/3/4)
  • Budd-Chiari syndrome - thrombus can be appreciated in the hepatic veins on U/S

• Treatment is etiology specific:
  • Acetaminophen toxicity - N-acetylcysteine
  • Viral hepatitis - disease-specific antiviral therapy
  • Autoimmune hepatitis - glucocorticoids
  • Budd-Chiari syndrome - TIPS, surgical decompression, or thrombolysis
Patient Follow-up

• This patient had an elevated IgG1 level of 1482 and an elevated anti-smooth muscle antibody titer of 1:80, suggestive of an autoimmune etiology.

• Patient improved clinically with only supportive care, so glucocorticoids were not provided.

• Patient was discharged after LFTs showed consistent downtrend and abdominal pain had resolved, with close GI follow-up scheduled.
References:


