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

Variant 1: Jaundice. No known predisposing conditions. Initial imaging.		
Procedure	Appropriateness Category	Relative Radiation Level
US abdomen	Usually Appropriate	0
CT abdomen with IV contrast	Usually Appropriate	***
MRI abdomen without and with IV contrast with MRCP	Usually Appropriate	0
MRI abdomen without IV contrast with MRCP	May Be Appropriate	0
CT abdomen without and with IV contrast	Usually Not Appropriate	****
CT abdomen without IV contrast	Usually Not Appropriate	***
ERCP	Usually Not Appropriate	ବହତ
US abdomen endoscopic	Usually Not Appropriate	0

This imaging modality was ordered by the ER physician



Findings (unlabeled)





Centrilobular hyperechogenicity/periportal edema with decreased parenchymal echogenicity (classic "starry sky" appearance) Findings (labeled)



Findings (unlabeled)





Findings (labeled)



Findings (unlabeled)





Findings (labeled)





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, antiliver/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.



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