RUQ Pain in Pregnancy: A Case of a Choledochal Cyst

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Agenda

• Our Patient A.B.
• RUQ Anatomy and Differential Diagnosis of Acute RUQ pain
• Choledochal Cysts
• Menu of Tests and Images from Companion Patients
• Summary and Follow-up of our Patient
Our Patient: Initial Presentation

- A.B. is a 38 year old G2P1 at 23 weeks who presented with 2-3 days of RUQ pain that continued to worsen
  - Pain is dull, feels like pressure, is not worsened or ameliorated with eating or activity
  - No fever, chills, nausea, vomiting, abdominal trauma or sick contacts, no change in bowel habits
  - No loss of fluid, no vaginal bleeding, minimal contractions q2 min which subsided, + fetal movement
Our Patient: Past Medical History

• Obstetric Hx: SVD x1, no complications
  – Benign pre-natal course for current pregnancy
• Medical Hx: Denies
• Surgical Hx: ? Open removal of gallbladder/cyst at age 13 in China
• Medications: None
• Allergies: None
• Denies tobacco, alcohol and drug use
Our Patient: Physical Exam

- T 98.9, BP 103/68, HR 81, RR 20
- Gen: NAD, mildly uncomfortable
- Abd: Gravid, moderately distended
  - marked tenderness to palpation in RUQ and R-side, fullness appreciated but unable to palpate borders due to tenderness
  - No rebound or guarding
  - Fundus palpable 1 cm below the umbilicus
- Labs: ALT 6, AST 21, alk phos 47, Tbili 0.2
  - WBC 7.2, Hg 12, Hct 34.6, Plt 271
Before moving on with the case let’s review:

1) The anatomy of important structures in the right upper quadrant
2) The differential diagnosis of Acute RUQ pain
3) Preferable Imaging Modalities in Pregnancy
Anatomy of the Biliary Tree

http://gallstoneflush.com/images/biliary%20tract.JPG
Differential Diagnosis of Acute RUQ Pain

• Gallbladder Disease:
  – Cholecystitis
  – Cholangitis
  – Choledocholithiasis
• Hepatitis
• Hepatomegaly
• Retroperitoneal appendicitis

• Malignancy:
  – Hepatocellular carcinoma
  – Cholangiocarcinoma
  – Liver metastases
  – Gastric cancer
  – Metastatic cancer
  – Lymphoma
Imaging Modalities in Pregnancy

• Preferable to Avoid Ionizing Radiation
  – Plain films, CT, ERCP, nuclear medicine

• Common Tests:
  – Ultrasound: sound waves
    • Pros: Inexpensive, good for identifying fluid
    • Cons: Requires skilled technologist, limited view
  – MRI: electromagnetic radio waves
    • Pros: use up to 1.5 Tesla, good soft tissue differentiation
    • Cons: should not use gadolinium as it crosses the placenta, expensive

• Special Tests:
  – MRCP: special MRI for imaging the bile and pancreatic ducts
Now it is time to review our patient’s imaging.

The first step: RUQ ultrasound

Let’s look at a normal ultrasound first.
Normal RUQ Ultrasound

RUQ ultrasound

- Left hepatic duct
- Right hepatic duct
- Liver
- Gallbladder
There is a very large anechoic structure found inferior to the liver.
Our Patient: RUQ Ultrasound

• 13.3 x 10.8 x 12.7 cm cystic structure with layering echogenic material, which most likely represents a markedly distended gallbladder

• Gallbladder neck and presumed dilated cystic duct are markedly tortuous, containing multiple echogenic foci, compatible with gallstones

• Common bile duct cannot be imaged due to tenderness limiting examination

• There is no peripheral intrahepatic biliary dilation but evaluation of the central biliary tree is limited

• No gallbladder wall thickening or definite pericholecystic fluid

• Normal hepatopetal flow is seen in the main portal vein.
To better define this abnormal fluid collection we should look at our patient’s MRI ...
Our Patient: MRI of Abdomen/Pelvis

What Do You See?

CORONAL SSFSE (HASTE) MRI

SAGITTAL SSFSE (HASTE) MRI
Our Patient: MRI of Abdomen/Pelvis

CORONAL SSFSE (HASTE) MRI
* Liver, * Cystic structure, * Fetus

SAGITTAL SSFSE (HASTE) MRI
PACS, BIDMC

* Liver, * Cystic structure, * Fetus
Our Patient: MRI Findings

- Massively dilated common bile duct which measures up to 11 x 13 cm in transaxial diameter
- Massive dilation of the central intrahepatic bile ducts with numerous filling defects within the ducts consistent with stones
- Pancreatic head is seen to be splayed about the distended CBD
- Duodenum is displaced laterally and posteriorly to the dilated duct
- No definite obstructing stone or mass is identified, but the distortion of the duodenum and pancreatic anatomy limits definitive evaluation
- Gallbladder is collapsed and displaced anterior to the dilated CBD
- Intrauterine pregnancy identified, no gross abnormalities visualized

DIAGNOSIS: Type I or IVb Choledochal Cyst
Choledochal Cysts: The Basics

- Rare, congenital dilatations of the biliary tract
  - Intrahepatic and extrahepatic
- Risk Factors:
  - Female predominance
  - more common in Asia
- Complications:
  - Recurrent cholangitis, Choledocholithiasis
  - Biliary stricture, Recurrent acute pancreatitis
  - Malignant transformation: 15% risk of developing cholangiocarcinoma
# Todani Classification for Choledochal Cysts

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
<th>Incidence</th>
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<tbody>
<tr>
<td>Type I</td>
<td>Fusiform or cystic dilations of the extrahepatic biliary tree</td>
<td>&gt;50% of choledochal cysts</td>
</tr>
<tr>
<td>Type II</td>
<td>Saccular diverticulum of the extrahepatic biliary tree</td>
<td>5% of choledochal cysts</td>
</tr>
<tr>
<td>Type III</td>
<td>Bile duct dilatation within the duodenal wall (choledochocoele)</td>
<td>5% of choledochal cysts</td>
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<table>
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<tr>
<th>Type IVa</th>
<th>Type IVb</th>
<th>Type V</th>
</tr>
</thead>
<tbody>
<tr>
<td>Multiple cysts present, intra and extrahepatic (Carolī’s disease)</td>
<td>Multiple cysts present, extrahepatic only</td>
<td>Intrahepatic biliary cysts only</td>
</tr>
<tr>
<td>5-10% of choledochal cysts</td>
<td>5-10% of choledochal cysts</td>
<td>1% of choledochal cysts</td>
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</table>
Imaging to Determine Management of Choledochal Cysts

• Type I, II and IV cysts will be visible on RUQ US
  – Types III (duodenal) and V (intrahepatic) will not

• HIDA scan to determine continuity with biliary tract
  – Excellent for extrahepatic cysts, difficult for intrahepatic cysts

• CT scans and MRI provide better intrahepatic visualization
  – Better for surrounding structures, evaluation of malignancy

• TREATMENT: Surgery
  • Cyst excision, Roux-en-Y hepaticojejunostomy
  • Others: Cholecystectomy, Intrahepatic cyst resection
Let’s look at some companion patients for more examples of choledochal cysts.
Companion Patient #1 – RUQ US

- Neonate born to 19 yo G2P1
- Identified Type I Choledochal Cyst in utero
- Thought to be ovarian cyst early on in pregnancy

RUQ ultrasound

Cyst, * Dilated intrahepatic ducts
Companion Patient #2 – RUQ US

- Choledochal cyst identified on RUQ US
- Polypoid mass at proximal region of cyst
- Pathology confirmed cholangiocarcinoma

RUQ ultrasound

Lee HK, Park SJ et al
Companion Patient #3 - MRCP

- 28 yo female
- Recurrent episodes of RUQ pain
- MRCP images best defined the type and extent of the choledochal cyst compared to US and CT images
- Surgery performed
- Patient doing well

NB: MRCP images do not require contrast as bile serves as a natural contrast material.
Companion Patient #4 - MRI

- 19 yo G1P0 at 22 wks
- Presented with RUQ pain
- Found Type I choledochal cyst filled with stones
- Underwent CCY, Roux-en-Y hepaticojejunostomy, and cyst excision
- Healthy baby born at 40 weeks gestation
- MRI used for diagnosis

This **NORMAL** HIDA scan shows radiotracer in the liver after 5 minutes (left image) and radiotracer in the gallbladder and duodenum after 45 minutes (right image).
Companion Patient #6: ERCP

- 50 yo male with abnormal liver function tests and abnormal anechoic structure found on RUQ US
- ERCP for CBD stent placement

- NB: ERCP is an invasive procedure, seldom used today for diagnosis of choledochal cysts.
Let’s get back to our patient ...
Our Patient: Intermittent History

- Recovered from acute episode of RUQ pain
- Decided to hold off on surgery until postpartum, if possible
- Healthy baby boy delivered at 37 weeks
- Imaging for surgical planning: CT and MRCP
  - Identified Type I vs. IVb choledochal cyst
  - Planned surgery: cyst excision, roux-en-y hepaticojejunostomy, intrahepatic stone removal, liver biopsy
Our Patient: Imaging for Surgical Planning

CT

MRCP

Liver

C+ COR CT

* Choledochal Cyst

Liver

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Our Patient’s Surgery: Roux-en-Y Hepaticojejunostomy

- Percutaneous transhepatic stents
- Hepaticojejunostomy
- Duodenum
- Jejunum
- Jejunojejunostomy

Our Patient - Follow Up

- Recovered slowly from surgery
- Baby boy is doing well
- Followed closely by hepatology and surgery

- Two weeks post-op CT
  - Dilated intrahepatic ducts *
  - Pneumobilia *
Summary

• Imaging is essential for diagnosis of RUQ pain and surgical planning
• Imaging modalities should be chosen carefully in pregnancy to avoid harm to the fetus
  – Good choices include ultrasound, MRI and MRCP
• Other RUQ imaging options include: CT, HIDA scan, ERCP
• Choledochal cysts are rare but have serious complications and should be removed surgically
Acknowledgements

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References

- Lieberman, G. “Primary Care Radiology: Radiologic Assessment of Abdominal Pain.” Primary Care Radiology Module. Eradiology.bidmc.harvard.edu