Recurrent Pyogenic Cholangitis

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Outline

• Overview of recurrent pyogenic cholangitis (RPC).
• Anatomy of the biliary tree.
• Review of imaging modalities useful in the diagnosis of RPC.
• Review and evaluation of index patient.
Recurrent Pyogenic Cholangitis (RPC)

- Characterized by intrahepatic pigment stones, intrahepatic biliary duct dilatation and strictures, and recurrent bouts of cholangitis.
  - Typically does not involve the gallbladder.
  - Presents clinically with Charcot’s triad (fevers, jaundice, and right upper quadrant (RUQ) abdominal pain).

- Common problem in East and Southeast Asia.
  - Increasing prevalence in the US due to migration from endemic regions.

- Diagnosis is based on clinical presentation in conjunction with imaging findings.
Pathogenesis of RPC

• Pathogenesis is incompletely understood.
• Possible etiologic role for biliary helminths, enteric superinfection, and malnutrition.
  – Biliary helminths, such as *Ascaris lumbricoides and Clonorchis sinensis*, induce ductal injury and inflammatory response.
  – Secondary infection with enteric bacteria leads to pigment stone formation and recurrent cholangitis.
  – Enteric bacteria with beta-glucuronidase activity enter the biliary system via the portal vein and cause deconjugation of bilirubin glucuronide, leading to precipitation of bilirubin-pigment stones.
  – Low protein diet leads to decreased levels of an endogenously produced inhibitor of beta-glucuronidase activity (glucaro-1:4-lactone), leading to increased susceptibility to pigment stone formation.
Anatomy of the Biliary Tree

right hepatic duct

liver

left hepatic duct

common hepatic duct

common bile duct

pancreas

pancreatic duct

gallbladder

cystic duct

sphincter of Oddi

duodenum

http://usmlemd.files.wordpress.com/2008/10/biliary20tract.jpg
Menu of Imaging Modalities for RPC

- Ultrasonography (U/S)
- Computed Tomography (CT)
- Endoscopic Retrograde Cholangiopancreatography (ERCP)
- Magnetic Resonance Cholangiopancreatography (MRCP)
Ultrasonography in RPC

- First line investigation.
- Characteristic findings: dilatation of the biliary tree and intrahepatic calculi (seen in up to 90% of patients).
  - Calculi show variable calcification leading to variable echogenicity and acoustic shadowing.
- Advantages: Useful for diagnosis and follow-up, useful for needle-guided aspiration and core biopsies, no ionizing radiation, non-invasive.
- Disadvantages: Findings can be obscured by pneumobilia, operator dependent.
Patients 1 and 2: RPC with U/S

Patient 1: U/S.  
Legend: Acoustic shadowing, intrahepatic calculi in left hepatic duct.

Patient 2: U/S.  
Legend: Dilatation of common hepatic duct.

CT in RPC

• Most important non-invasive study for diagnosis.
• Characteristic findings: intrahepatic biliary duct dilatation, intrahepatic calculi, pneumobilia, hepatic parenchymal processes (e.g. atrophy, abscesses, bilomas), and cholangitis.
• Advantages: provides a complete picture of the disease process.
• Disadvantages: ionizing radiation, contrast allergy.
Patients 3 and 4: RPC with CT

Patient 3: C- axial CT of the abdomen.
Legend: Calcified calculi within intrahepatic biliary ducts.

Patient 4: C+ axial CT of the abdomen.
Legend: Dilatation of left hepatic duct with multiple filling defects.

ERCP in RPC

• Direct cholangiography.
• Characteristic findings: duct dilatation, calculi, intrahepatic strictures, and decreased arborization of the biliary tree.
• Advantages: Better spatial resolution than MRCP, allows for therapeutic intervention.
• Disadvantages: May not depict all of biliary tree, may precipitate septic shock.
Patients 5 and 6: RPC with ERCP

Patient 5: ERCP obtained with use of an occlusion balloon.
Legend: Multiple calculi within the left hepatic duct, ductal stricture.

Patient 6: ERCP.
Legend: Decreased arborization of right biliary ducts, indicating obstruction or stricture.
MRCP in RPC

- Performed using heavily T2-weighted images.
  - Bile-filled structures appear hyperintense against a hypointense background.
- Characteristic findings: calculi, intrahepatic biliary duct dilatation and strictures, and hepatic parenchymal processes (hepatic abscesses are hypointense on T1 and hyperintense on T2).
- Advantages: Allows visualization of ducts proximal to an obstruction, non-invasive, non-ionizing radiation, contrast not required, allows for 3D reconstruction of biliary tree.
- Disadvantages: Does not allow for immediate therapeutic intervention.
Patients 7 and 8: RPC with MRCP

**Patient 7**: Thick-slab reconstruction of a T2-weighted MRCP.
**Legend**: Obstructing calculus in left hepatic duct.


**Patient 8**: T2-weighted MRCP.
**Legend**: Multiple dark filling defects, indicating intrahepatic calculi.

Our Patient: Clinical Presentation

- 83 year old Chinese female with repeated episodes of RUQ abdominal pain, fevers, and nausea/vomiting.

- Laboratory values are unremarkable except for moderately elevated alkaline phosphatase.

- Past surgical history is remarkable for an open cholecystectomy approximately 50 years ago and a choledochoduodenostomy.
## ACR Appropriateness Criteria for RUQ Pain

**Clinical Condition:** Right Upper Quadrant Pain

**Variant 1:** Fever, elevated WBC, positive Murphy’s sign.

<table>
<thead>
<tr>
<th>Radiologic Procedure</th>
<th>Rating</th>
<th>Comments</th>
<th>RRL*</th>
</tr>
</thead>
<tbody>
<tr>
<td>US abdomen</td>
<td>9</td>
<td></td>
<td>O</td>
</tr>
<tr>
<td>CT abdomen with or without contrast</td>
<td>6</td>
<td>Generally should follow US of the RUQ based on US findings. With IV contrast preferred.</td>
<td>☹ ☹ ☹</td>
</tr>
<tr>
<td>Cholescintigraphy</td>
<td>6</td>
<td>Generally should follow US of the RUQ based on US findings.</td>
<td>☹ ☹</td>
</tr>
<tr>
<td>MRI abdomen with or without contrast</td>
<td>6</td>
<td>Generally should follow US of the RUQ based on US findings. See statement regarding contrast in text under “Anticipated Exceptions.”</td>
<td>O</td>
</tr>
</tbody>
</table>

**Rating Scale:** 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate

*Relative Radiation Level*
Our Patient: U/S Findings

• Summary of pertinent findings from U/S.
  – A large quantity of air was observed within the biliary system.
  – Limited visualization of the liver due to hyperechogenic air.
  – Further assessment by CT recommended.
**Our Patient: Initial CT Evaluation**

**Legend:**
- Stone in intrahepatic biliary duct,
- Cystic biliary dilatation,
- Pneumobilia,
- Spleen,
- Aorta,
- Stomach,
- Nodular, heterogeneous liver,
- Periductal enhancement, consistent with cholangitis.

*Our Patient: C+ axial CT of the abdomen in portal venous phase; PACS, BIDMC.*
Differential Diagnosis Based on CT Findings

• Simple obstructive cholangitis
  – Choledocholithiasis
  – Benign biliary stricture
  – Cholangiocarcinoma
  – External compression (hepatocellular carcinoma, pancreatitis)

• Primary sclerosing cholangitis
  – Ongoing inflammation leading to irregularly narrowed bile ducts; beaded appearance.

• Caroli disease
  – Congenital cystic dilatation of intrahepatic bile ducts.

• Recurrent pyogenic cholangitis
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• **Recurrent pyogenic cholangitis**
Our Patient: Calculi, Pneumobilia, and Biliary Duct Dilatation on CT

Legend:
Stone in intrahepatic biliary duct,
Cystic biliary dilatation, with air fluid levels,
Nodular, heterogeneous liver.

Our Patient: C+ axial CT of the abdomen in arterial phase; PACS, BIDMC.
Our Patient: Intrahepatic calculi on CT

Our patient: C+ sagittal reconstruction CT of the abdomen in portal venous phase; PACS, BIDMC.
Legend: Intrahepatic calculi.

Our patient: C+ coronal reconstruction CT of the abdomen in arterial phase; PACS, BIDMC.
Legend: Intrahepatic calculus.
Our patient also had a number of ERCPs to visualize her biliary anatomy and to remove obstructing calculi.
Our Patient: Multiple Filling Defects on ERCP

Legend:
Dilated common bile duct, Filling defects consistent with calculi in the left hepatic duct.
Our patient: Filling defect in the Right Hepatic Duct on ERCP

Legend:
Filling defect consistent with a calculus in the right hepatic duct.
Our Patient: Strictureing of Biliary Radicles on ERCP

Legend:
Strictures within intrahepatic biliary radicles.
Our Patient: Strictures within the Left Hepatic Duct on ERCP

Legend:
Dilated common bile duct, Stricturing within the left hepatic duct.

Our Patient: ERCP; PACS, BIDMC.
Our Patient: Heterogeneous Mass on CT

Legend: On follow-up CT scan, a heterogeneous, irregular mass was noted within the caudate lobe of the liver. The mass was found to be isodense on non-contrast CT and in the arterial phase (C+) CT but heterogeneously enhancing in the portal venous phase (C+) as shown in this image. This mass was concerning for cholangiocarcinoma.
Cholangiocarcinoma in Patients with RPC

- Incidence of cholangiocarcinoma in patients with RPC is 1.5-11%.
- Chronic bacterial infection leads to chronic proliferative cholangitis, which in turn leads to atypical epithelial hyperplasia and cholangiocarcinoma.
- Elevation of CA19-9 in most cases.
- Characteristic findings on CT
  - Predominately hypoattenuating with irregular margins.
  - Variable enhancement after contrast administration.
    - No enhancement, minimal peripheral enhancement, or central enhancement.
  - Delayed enhancement with increasing attenuation in ¾ of patients.
Our Patient: Biopsy of Biliary Mucosa via ERCP

- Biopsy of the irregular mass revealed biliary mucosa with chronic inflammation and granulation tissue.
- There was no evidence for cholangiocarcinoma.
- CA19-9 was within normal limits.

Our patient: Intraductal biopsy of mucosa at the takeoff of the right hepatic duct was performed with cold forceps via ERCP; PACS, BIDMC.
Summary

• Recurrent pyogenic cholangitis is a disease of the intrahepatic biliary system, characterized by intrahepatic pigment stones, strictures and dilation of the intrahepatic biliary tree, and recurrent cholangitis.

• Useful imaging modalities include U/S, CT, ERCP, and MRCP.

• We saw a patient with RPC who presented with RUQ pain and fevers, and we reviewed the imaging findings, which included multiple intrahepatic calculi on CT and ERCP, dilatation and stricturing of the intrahepatic biliary tree on CT and ERCP, and periductal enhancement on CT, consistent with cholangitis.
References


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