Endoscopy in Primary Sclerosing Cholangitis

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Goals

- Role of ERCP in PSC
  - Initial Diagnosis
  - Complications
- Managing procedure risks
- Role of MRCP
Endoscopic Retrograde Cholangiopancreatography
ERCP – anatomy on cholangiogram

Gallbladder

Common Hepatic Duct

Cystic Duct

Common Bile Duct
Normal Intrahepatic Cholangiogram
PSC - pathogenesis

• Progressive inflammatory destruction and fibrosis of intra and extra hepatic bile ducts

• Primary injury to medium - large sized bile ducts (>100 μm in diameter)

• Smaller ducts captured on liver biopsy demonstrate nonspecific changes of obstruction or disappear (ductopenia)
**PSC – diagnostic criteria**

- **Chronic cholestatic liver enzyme elevation**
  
  2-3x ↑alk. Phos. / bilirubin vs. AST/ALT > 6 mo.

- **Compatible bile duct injury on ERCP (or MRCP)**

- **Exclusion of other causes of bile duct obstruction**

- **Liver biopsy plays limited role**
PSC – diagnostic criteria

Cholangiogram changes
- Strictures
- Saccular dilatation
- Beading
- Diverticulum

Location
- 10% intrahepatic ducts
- 15% extrahepatic ducts
- 75% combined
PSC – extrahepatic bile duct
PSC – intrahepatic ducts
PSC – intra and extrahepatic ducts
**PSC – special subgroups**

- Small Duct PSC
- PSC- Autoimmune Hepatitis Overlap
Small duct PSC

- 5% of PSC cases

- Diagnostic criteria
  - Chronic cholestatic liver disease
  - Liver biopsy compatible with PSC
  - Normal cholangiogram
  - Other causes of liver disease excluded
Small duct PSC

- Limited progression to large duct PSC (23%)
- Better prognosis
  - Less risk for cholangiocarcinoma
  - Longer interval to transplantation
**PSC - Autoimmune Hepatitis Overlap**

- Less than 10% of PSC patients
- ? More common in younger patients
- Symptomatic presentation
- Circulating autoantibodies more common (ANA, SMA, pANCA)
PSC - Autoimmune Hepatitis Overlap

• Diagnostic criteria
  • Cholangiogram compatible with PSC
  • Liver biopsy – more pronounced inflammatory portal tract infiltrate and interface hepatitis vs. PSC

• Responds to immunosuppressive therapy – symptoms, biopsy, and cholangiogram
**PSC – role of liver biopsy**

- Not required in all cases
- Confirm diagnosis and assess stage of fibrosis
- Required to exclude overlap with autoimmune hepatitis
- Required to diagnose small duct PSC
PSC - complications

- Stones
- Dominant stricture
- Cholangiocarcinoma
- Cholangitis
**PSC - complications**

- Often identified by change in clinical status
  - Abdominal pain
  - Fever
  - Jaundice
  - Rising alk. Phos., bilirubin
- Accelerate progression of disease
PSC + bile duct stones
PSC + dominant stricture

- Develops in 10-15% of patients
- Discrete narrowing in the extrahepatic bile duct
- 80% respond to endoscopic therapy
- Similar presentation for cholangiocarcinoma
- Response to endotherapy may differentiate
PSC + dominant stricture
**PSC + dominant stricture**

- Balloon dilatation may suffice.
- Long term stenting should be avoided
  - bacterial contamination
  - stone formation
- Exchange every 4-8 weeks until resolution
- Retrospective data suggests endoscopic therapy improves survival.
PSC + Cholangiocarcinoma

• Develops in 0.6-1.5% pts/year
• 10% lifetime risk
• No correlation with duration of disease or presence of cirrhosis
• Most commonly presents with change in status
  • pain, fever, rising liver enzymes, bilirubin, CA19-9
• Occasionally an incidental finding at ERCP or transplant
Cholangiocarcinoma - diagnosis

- CA19-9
- CA19-9/CEA index
- MRI / MRCP
- ERCP with tissue sampling
- Cholangioscopy with tissue sampling
- Positron Emission Tomography (PET scan)
PSC + Cholangiocarcinoma on ERCP
Cholangiocarcinoma - tissue sampling

- Negative cytology does not exclude tumor
- Special techniques may enhance accuracy
  - Immunocytochemistry for K-ras, \( p53 \)
  - Fluorescent In Situ Hybridization
  - Digital Image Analysis
**PSC + Cholangiocarcinoma - therapy**

- Stents placed across an obstructing tumor can improve the clinical status temporarily.
- Chemotherapy, radiation, and surgery alone or in combination do not improve prognosis.
- Chemotherapy + radiation prior to transplantation provides best opportunity for long term survival.
PSC + Cholangiocarcinomna - therapy
PSC + Cholangiocarcinoma - surveillance

• No clear evidence of benefit
• Serial CA 19-9 is a practical approach
• Serial ERCPs with tissue sampling increases risk of complications.
**ERCP in PSC - risks**

- Complications in up to 10% of procedures
- Pancreatitis, cholangitis, perforation, bleeding
- Minimize with
  - proper indication for procedure
  - pre-procedure antibiotic
  - avoid pancreatic duct injection
  - proper indication for therapy
MRCP - normal
MRCP in PSC

PRO

• No sedation, scope insertion, duct manipulation, contrast exposure, radiation
• Visualize liver parenchyma, and abdominal structures
• Applicable with altered anatomy
• Visualize ducts above obstruction

CON

• Less sensitive in defining bile duct abnormalities
• No tissue sampling
• No therapeutic options

PRO CON
PSC on MRCP
PSC on MRCP

Strictures, abnormal duct walls & peribiliary enhancement of active cholangitis
PSC with Cholangiocarcinoma
MRCP in PSC

• Overall best initial option for diagnosis
• May not avoid subsequent ERCP
• Compliments ERCP when evaluating dominant stricture or cholangiocarcinoma
PSC - Summary role of ERCP

• Required for diagnosis but role changing with refinement and experience with MRCP
• Required to fully evaluate change in clinical status
• Provides options for tissue sampling
• Provides treatment options for complications
• Risks significant but can be limited proper case selection, preparation, and proper indications for therapeutic intervention
Thank You !!!