Biliary Cyst

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PGY-4
April 19, 2012
Case Presentation

- 56 year old male referred from an outside institution for evaluation of type I choledochal cyst. Pt initially presented in 2008 with a history of chronic abdominal pain associated with weight loss (15 lbs) over 1 ½ years and mild elevation of LFTs.
- Extensive workup including MRCP, ERCP and EUS showed a dilated bile duct measuring up to 1.5cm, without intrahepatic duct dilation or evidence of stone or masses.
- PMH: HTN, Asthma
- PSH: none
- Social Hx: Etoh (3-4+ beers/daily +/- vodka x 40 yrs), denies smoking/drug use
- FamilyHx: noncontributatory
Physical Exam and Labs

- T98.5 BP 112/71 HR 65
- Wt 155lbs
- HEENT no adenopathy
- Skin nonjaundice
- Chest clear
- RRR
- Abdomen – soft, nondistended, vague abdominal discomfort on palpation, no appreciable mass
- Wbc 4.7
- INR 1.13
- Tbil 0.9
- AlkP 241
- AST 165
- ALT 103
- Lipase 36
- CEA 0.6
- CA 19-9 93
Diagnosis

- Type I bile duct cyst
Operation

- Cholecystectomy
- Biliary cyst excision
- Roux-en-Y hepaticojejunostomy
Bile duct cyst
Hepaticojejunostomy
Anastomosis
Pathology

- Gallbladder – chronic cholecystitis
- Bile duct cyst – widest diameter 2cm
  - Smooth mucosa, no evidence of malignancy
Post-Op Course

- POD#3 – started on diet
- POD#9 – discharged home
Biliary Cyst
Epidemiology

- Account for 1% of all benign biliary disease
- Western population 1/100,000-150,000
- Asian population 1/1000
- More than half the cases are in Japan
- Female:Male 3-4:1
- Usually diagnosed in childhood
  - 60% before 10 years old
  - 20% adults
Classification

- 1959 - Alonso-Lej proposed first classification system
- 1977 - Modified by Todani et al.
Distribution of Bile Duct Cysts

Type I: 79.4%
Type II: 2.7%
Type III: 3.7%
Type IV: 13.6%
Type V: 0.6%

Pathogenesis

- Congenital vs acquired
- **Anomalous pancreaticobiliary duct junction** (Babbitt 1969)
  - Long common channel
  - Pancreaticobiliary reflux
  - High amylase level in bile
  - Found in 50-80% of pt
- Distal ductal stenosis
- Sphincter dysfunction
- Aganglionosis of distal duct
- Hereditary?
Signs and Symptoms

- Classic triad – 20%
  - Abdominal pain
  - Abdominal mass
  - Jaundice
- Cholangitis
- Pancreatitis
- Weight loss
- Asymptomatic/incidental finding
Associated Hepatobiliary Pathology

- Cholangitis
- Pancreatitis
- Intrahepatic abscesses
- Cirrhosis
- Portal hypertension
- Cystolithasis/hepaticolithasis
- Cholecystitis
- **Malignancy**
  - Cholangiocarcinoma 20-30x increase incidence
Associated Malignancy

- Overall cancer risk 10-15%
- Earlier presentation (10-15 years) compared to general population
- Risk increases with age

Associated Malignancy

- Location
  - Extrahepatic duct 50-62%
  - Gallbladder 38-46%
  - Intrahepatic duct 2.5%
  - Liver and pancreas 0.7%
Malignancy After Cyst Excision

- Excision of the biliary cyst does not eliminate risk of malignancy in the biliary tract
  - Enteric anastomosis
  - Intrahepatic duct
- Lee et al (2011) – South Korea
  - 0.6% (with excision) vs 9.9% (without excision)
- Long term surveillance is required for all patients after cyst excision
Workup

- Labs – LFTs, INR
- Abdominal Ultrasound
- CT scan
- EUS
- MRCP
- ERCP
- Percutaneous transhepatic cholangiography
Treatment

- Historically cystenterostomy (to duodenum and jejunum) was performed to relieved symptoms
  - Complications - ascending cholangitis, strictures, calculi, malignant transformation.
  - Found high incidence malignancies despite adequate drainage
  - Many required reoperation
- Currently the recommended treatment is complete extrahepatic cyst excision with roux-en-y hepticojejunostomy
- Patients with prior cystenterostomy is also recommended to have revision with cyst excision.
Technical Pearls

- Excision of cyst is often difficult due to fibrosis and adhesions to surrounding structures from recurrent inflammation
- Intramural saline injection
- Partial cystectomy
  - Leave the posterior wall
  - Need to excise mucosal layer
  - Lifelong surveillance
Cyst excision and roux-en-y hepaticojejunostomy

Extrahepatic cyst excision with roux-en-y hepaticojejunostomy +/- partial hepatectomy

Excision of biliary diverticulum only

Roux-en-y cholangiojejunostomy with transhepatic stent placement +/- hepatectomy Liver transplant

Endoscopic sphincterotomy
Summary

- Biliary cyst is a rare finding in the Western hemisphere compared to Asia.
- It is most commonly found in children and female population.
- Most common symptom is abdominal pain and cholangitis
- Associated with very high incidence of malignant transformation
- Treatment is extrahepatic cyst excision with roux-en-y hepaticojejunostomy
- All patients need lifelong surveillance because of the risk of malignancy
References

- Kamisawa et al. Pancreaticobiliary maljunction. Clinical gastroenterology and hepatology. 2009. 7:S84-S88