An Unusual Cause of Pain in the Right Hypochondrium
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ABSTRACT
Choledochal cyst is an uncommon anomaly of the biliary system. It manifests by cystic dilation of the extra or intrahepatic biliary tract or both. Three cases of choledochal cysts are reported, who had presented with pain in right hypochondrium. All the three patients presented in one Surgical Unit of a Teaching Hospital within a span of three months. The first patient was initially managed as obstructive jaundice, second as acute pancreatitis and third patient as a case of pancreatic pseudocyst. However, after investigations, the final diagnosis in all three of them was the same i.e. choledochal cyst which was managed by Roux-en-Y hepatico-jejunostomy.


INTRODUCTION
Although cystic disease of the biliary tree has been described since 1723, much about its etiology, pathophysiology, natural course and optimal treatment remains under debate. Choledochal cysts are usually a surgical problem of infancy or childhood, however, in approximately 20% of cases they are recognized in adults. Early diagnosis and proper treatment are very important, because the cysts are associated with a risk of biliary tract malignancy.1 External drainage alone has no definitive role in the surgical management of choledochal cysts. In general, all bile duct cysts should be excised and bile flow re-established by mucosa-to-mucosa biliary-enteric anastomosis.2 The three cases that we report here were all treated by complete excision of the cyst and a Roux-en-Y hepatico-jejunostomy.

CASE REPORT
Case 1: A 40 years old lady from Kotli, AJK presented in E.R. of Benazir Bhutto Hospital (BBH), Rawalpindi in July, 2011 with two and a half years history of pain in the right hypochondrium associated with vomiting off and on. Symptoms had exacerbated during the last 2 months. She also had symptoms suggestive of obstructive jaundice and cholangitis. On examination, she was pale, jaundiced and had scratch marks all over the body. Abdominal examination revealed a 7 x 8 cms fusiform palpable mass in right (RHC) that moved with respiration.

Initial baseline investigations showed a raised WBC count (18.4 x 10^3/L), t-bilirubin (4.1 mg%) and ALP (1014 U/L). Serum amylase was not raised. Abdominal ultrasound showed a large cystic lesion at porta hepatis, separate from gallbladder. Contrast enhanced CT scan abdomen showed an 8.1 x 6.4 cms thick walled cystic area at the confluence of hepatic ducts and porta hepatis extending for 10 cms in craniocaudal direction and distally tapered down in CBD. Intra and extra hepatic biliary channels and right and left hepatic ducts were moderately dilated. Choledochal cyst type-I was the final diagnosis.

Patient was operated in July 2011 and underwent complete excision of the cyst with Roux-en-Y hepatico-jejunostomy. Operative findings were a type-I choledochal cyst extending from porta hepatis down to the duodenum. Histopathology of the specimen was consistent with the operative finding. Patient had a smooth postoperative course and he was discharged in a stable condition and was regularly followed-up without any complaints.

Case 2: A 30 years old female was referred from a private hospital in Gujar Khan in September, 2011 with a 2 years history of RHC pain associated with vomiting off and on. For the last one week, she had acute exacerbation of symptoms. Laboratory investigation in emergency reported very high serum amylase level and was admitted with the initial diagnosis of acute pancreatitis. Ultrasound, however, showed three cystic areas in the right lobe of liver measuring 5 x 5, 2.5 x 3.5 and 1.5 x 2 cms. Thick walled gallbladder contained multiple small calculi. Sonologist suspected liver abscess. For clarification, contrast enhanced CT scan abdomen was done which revealed multifocal saccular, fusiform dilatation of the proximal CBD (5.5 cms maximum). Cysts appeared infected and liver size was within upper limit of normal. The final diagnosis based on the CT scan was type IV (a) choledochal cysts.

Patient was operated in BBH, Rawalpindi in October 2011. Complete excision of the cyst followed by Roux-en-Y hepatico-jejunostomy was performed. A 5 cm fusiform choledochal cyst extending from porta hepatis
to first part of duodenum was removed. Histopathology was consistent with choledochal cyst. Patient had a smooth postoperative recovery and was discharged in a stable condition. Regular follow-ups showed a complete recovery.

Case 3: A 30 years old male, resident of Mianwali was admitted through E.R. in September, 2011 in BBH, Rawalpindi with 8 months history of RHC pain associated with episodes of vomiting off and on. His history dated back to January, 2011 when he first suffered a severe episode of RHC pain associated with repeated vomiting. Initial work-up based on ultrasound abdomen and a CT scan showed a 4 cm well defined cystic lesion near the head of pancreas. With the diagnosis of pseudocyst of pancreas, the patient was operated in a private hospital in Mianwali, where cystogastrostomy was done in February 2011. However, in the following months, there was a persistence of the initial symptoms without any relief from symptomatic treatment. He finally presented in BBH, Rawalpindi. Contrast enhanced CT scan of abdomen (Figure 1) showed a 4 cm diameter and 9 cm long cystic tubular structure, extending from porta hepatitis to the head of pancreas lying anterior to portal vein and causing compression of second part of duodenum but no obstruction. ERCP showed a markedly dilated CBD throughout its course without intrahepatic dilatation. The final diagnosis was choledochal cyst type-I.

The patient was operated in September 2011. Complete excision of cyst with Roux-en-Y hepatico-jejunostomy was done. Patient had a smooth postoperative course and was discharged in stable condition. Regular follow-ups showed a complete recovery.

**DISCUSSION**

Choledochal cysts are rare congenital entities characterized by single or multiple dilatations of the intra and/or extrahepatic biliary tree. Although the incidence in the western population is 1 in 100,000-150,000 live births, it is remarkably higher in Asian countries, particularly Japan, where they can be found in up to 1 in 1,000 live births. There is also an unexplained female: male preponderance, commonly reported as 1.5:1 in the paediatric population and in up to 4.9:1 in the adult population.

Multiple etiologic theories have been proposed for the origin of choledochal cysts, however, exact etiology remains unknown. The most widely accepted theory relates the cyst to an anomalous pancreaticobiliary ductal union (APBDU) leading to a long common channel, allowing pancreatic juice to reflux into the biliary system causing ductal ectasia and ultimately dilatation. Pancreaticobiliary reflux can result in various pathological conditions including choledocholithiasis, cholangitis, gallstones, acute pancreatitis, bile duct and gallbladder cancer and pancreatic ductal carcinoma.

The clinical presentation of this disease is not specific. Adults are usually asymptomatic or present with vague symptoms, generally in the right upper abdominal quadrant. The classical triad of clinical symptoms, described by Alonso-Lej et al., involves right hypochondrial pain, palpable abdominal mass and jaundice. This is most frequently seen before the age of 10 years. In contrast to children, choledochal cysts in adults are associated with a higher incidence of cyst related complications. These include cystolithiasis, hepaticolithiasis, calculous cholecystitis, pancreatitis, cholangiocarcinoma and cirrhosis with portal hypertension.

Diagnostic studies focus on the imaging of the cyst. Often the cyst is found on an ultrasound scan or a CT
scan done for generalized symptoms. Endoscopic retrograde cholangiopancreatography (ERCP) is the most effective examination method for evaluating the anatomy of insertion of pancreatic duct prior to surgery. Endoscopic ultrasonography (EUS) allows definite diagnosis of pancreaticobiliary maljunction. MRI, MRCP or MDCT (multidetector computed tomography) can suggest the correct pre-operative diagnosis most of the time.

Years ago, the primary treatment of cysts was simple internal drainage by cystenterostomy or partial cyst excision. However, several serious clinical outcomes, including stomal stenosis, cholestasis, cholangiolithiasis and even cholangiocarcinoma, led to poor prognosis and even secondary surgical operation. At present, total cyst excision with Roux-en-Y hepaticojejunostomy is considered to be a safer and more ideal treatment for patients with congenital biliary duct cysts. This should be performed as early as possible and the optimal treatment time is the infant period as there is a small but real cancer risk over the long-term if the cyst remains in place. For types, Todani classification, incidence and treatment of choledochal cysts, see Table I.

REFERENCES


