

Case Report:

Choledochal Cyst Coexisting with Gallbladder Carcinoma - An Uncommon Case Report

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Abstract:

Choledochal Cyst is a relatively rare condition. Even rarer is a choledochal cyst in association with a gallbladder carcinoma. This study reports a rare case of choledochal cyst coexisting with gallbladder carcinoma in a Bangladeshi patient. A 35 year old lady presented at IBN Sina Medical College Hospital, Kallyanpur, Dhaka with the history of recurrent right upper quadrant abdominal pain from childhood, which became severe for last 4 days before admission. The pain was colicky in nature and radiated to the back. Episodes were associated with low-grade fever, anorexia as well as vomiting. The preoperative diagnosis was made by abdominal ultrasound and MRCP. Exploratory laparotomy, enbloc cholecystectomy with excision of the choledochal cyst and roux-en-Y hepaticojejunostomy was also done. Post operative recovery was uneventful. Patient was followed up for six months and no obvious complication was noticed. Early suspicion of this rare pancreato-biliary disease is important because surgical treatment is the only way to avoid the complications of the disease.

Keywords: Adult; choledochal cyst; gallbladder cancer; surgery

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Introduction:

Choledochal Cyst is a relatively rare condition. Even rarer is a choledochal cyst in association with a gallbladder carcinoma. Choledochal cyst is a pathologic condition characterized by varying degrees of congenital dilatation of the biliary system, including the common, intrahepatic and extrahepatic bile ducts.¹ It is a rare congenital anomaly which is, for unknown reasons, quite common in Asian Population and very rare in Africans.^{2,3} At least 60% of patients are diagnosed during the first decade of life, but 20% remain undiagnosed until adulthood.^{2,4} Due to the recent improvement in noninvasive hepatobiliary imaging technique majority of cases of choledochal cysts are now detected in adult in most developed countries.^{4,5,6}

These cysts are clinically important because of their attendant complications of recurrent cholangitis, biliary stricture, choledocholithiasis, recurrent acute pancreatitis, and malignant transformation.⁷ The risk

of malignant transformation is well-documented to be age-related and believed to be the result of chronic inflammation.⁸ The malignant transformation usually arises from the cyst or along the bile duct, especially when there is anomalous pancreaticobiliary junction.^{9,10} Very rarely, the cancer can occur in the gallbladder or in the head of the pancreas. The frequent occurrences of cancers in adult choledochal cyst make surgical management more challenging.⁶ We presented a case of adult choledochal cyst coexisting with gallbladder cancer highlighting the clinical presentation and the management option for such patients in an economic poor resource setting.

Case Report

A 35 years old lady hailing from Feni, rural Bangladesh got admitted to this hospital with the complaints of recurrent right upper quadrant abdominal pain from her childhood, which became severe for the last 4 days. The pain was colicky in nature and radiated to the back. Episodes were associated with low-grade

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fever, anorexia and vomiting.

Physical examination revealed acutely ill woman anaemic, anicteric with right hypochondrial tenderness and a positive Murphy's sign.

She is non-diabetic, normotensive and non-asthmatic. She has not any positive family history relevant to this disease.

Laboratory investigation reveals that Complete Blood Count showed leukocytosis with predominant neutrophilia (78%). Hb% was 8.6 mg/dl later corrected by blood transfusion. SGPT (203 U/L) and alkaline phosphatase (421 U/L) were elevated. Prothrombin time was 26.6 s on admission later corrected. HBsAg was negative.

On Abdominal ultrasound findings were fusiform dilatation of the extrahepatic biliary duct and common bile duct (about 82.6x53.6mm), the pancreas and other intra-abdominal organs were essentially normal, and there were no ascitic fluid, right sided mild pleural effusion.

At laparotomy, there was a markedly dilated CBD, extending from retroduodenal part up to the common hepatic duct. The gall bladder wall was thick at neck region, cystic lymph node was enlarged. Cholecystectomy and complete resection of the choledochal cyst (supraduodenal to the confluence) was done in enbloc. Billiary continuity was established by an end-to-side hepaticojejunostomy with Roux-en-Y loop.

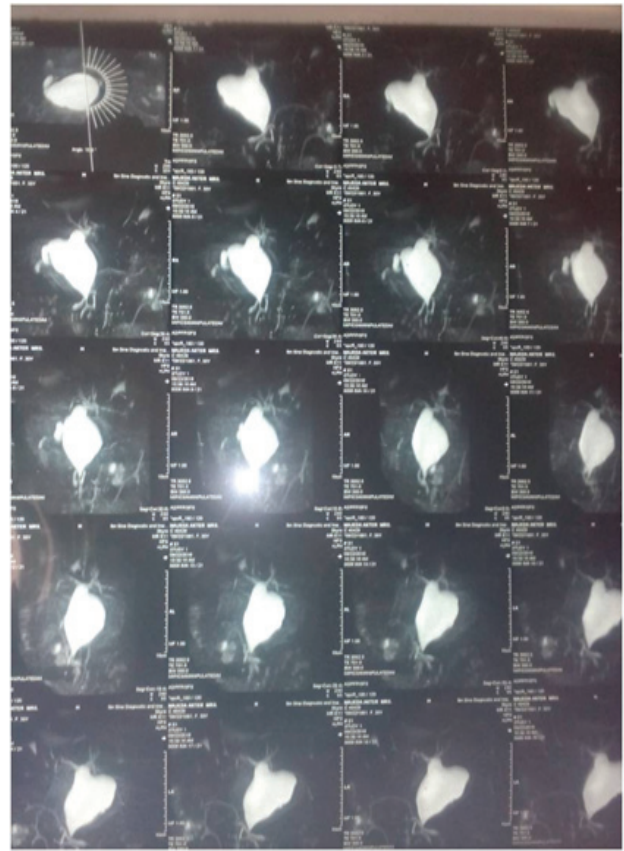
Postoperatively she had an uneventful recovery from the anaesthesia. There were no other complications of anastomosis and wound. Stitches were removed on the 10th POD and the wound was healthy.

Histopathology shows Adenocarcinoma of the gallbladder grade 1 with lymph node metastasis. No malignant transformation in the cyst wall.

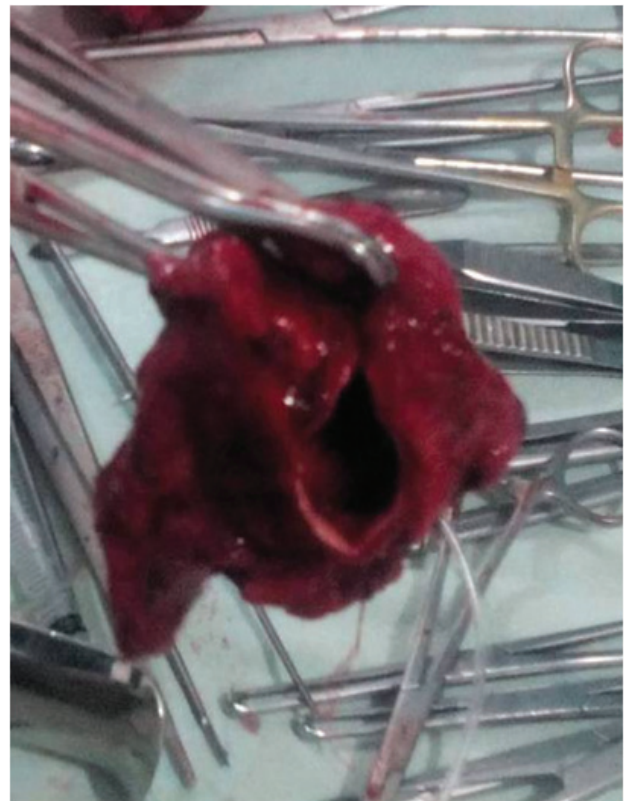
The patient was followed up for six months and no obvious complications like upper abdominal pain, jaundice, peritonitis were detected.

Discussion:

Choledochal cyst is a very rare congenital anomaly and has been established to occur in 1 in 13 thousand to 1-2 million live births.¹² Mostly occurs in Asia and have a male female ratio ranging from 3:1 to 8:1.^{13,14} Clinical presentation of adult choledochal cyst differs from that of neonates or children.¹⁵ Presentation in adults is nonspecific and often leads to delay in diagnosis. The classic triad of abdominal pain, right upper quadrant mass, and jaundice are more common in the pediatric population than adult (85 vs.25%, respectively).¹¹ Most adult like our patient usually present with abdominal pain, with or without



MRCP shows Suggestive of choledochal cyst, both CHD and CBD were dilated (8.7x5.1 cm) and Mild right Pleural thickening



jaundice, or symptoms suggesting pancreatic, or biliary origin which prompts hepatobiliary imaging before the diagnosis is made.

USG is a very important auxiliary method for the diagnosis of choledochal cyst type I. Diagnosis and management of choledochal cyst are important because patients may develop cholangiocarcinoma, gallbladder carcinoma etc.^{16,17}

Radical resection and Roux-en-Y anastomosis is the standard method for the management of congenital choledochal cyst. For patients with type I cysts, complete resection of cyst was performed together with reconstruction with hepaticojejunostomy. Unfortunately, the management efficiency for the other three types of choledochal cysts still not well defined.

In this case, we present the experiences of management of choledochal cyst coexisting with

gallbladder carcinoma in adult. In this study, we present one case of patients received resection and Roux-en-Y anastomosis. No recurrence or postoperative complications were reported in the 2-6 months follow up.

Gall bladder cancer for most patients is a lethal disease with a grim prognosis. The median survival is less than six months and a five-year survival figure of 5 per cent reported in an early case.

Conclusion

We reported an adult Type I choledochal cyst associated with gallbladder carcinoma in a woman who was successfully managed in our hospital. We suggest high index of suspicion on the part of surgeon managing such an adult patient presenting with upper abdominal pain, vomiting and anaemia. Early diagnosis and aggressive surgical resection may provide the only hope of prolonged/long survival.

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