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Case Report

Remnant intrapancreatic choledochal cyst with symptomatic cystolithiasis

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ABSTRACT

A young female who had undergone choledochal cyst excision as a child presented with retained stone in the remnant intrapancreatic choledochal cyst which caused acute pancreatitis. This case is being reported to emphasise the importance of excising the choledochal cyst through the entire dilated part and to report the possibility of patients having symptoms due to protein plugs or retained stone in the intrapancreatic remnant.

Keywords: Remnant choledochal cyst, Intrapancreatic choledochal cyst, Protein plugs, Cystolithiasis

INTRODUCTION

Choledochal cyst is abnormal dilatation of the biliary system most commonly involving the extrahepatic biliary tree. The lower extent of the dilatation can sometimes extend up to the intrapancreatic portion making complete excision difficult.[1] The lower limit of excision is decided by the level of tapering of bile duct diameter but not defined by standard recommendations. Occasionally patients have a large remnant intrapancreatic cyst which can lead to pancreatitis secondary to protein plugs.[2] Our patient also had a remnant intrapancreatic choledochal cyst with a retained stone.

CASE REPORT

Our patient was a 23 year old female who had undergone extrahepatic bile duct excision with roux en y hepaticojejunostomy for type 4A choledochal cyst as a 9 year old child in 2003 at another center. She was well for the next 14 years but then presented with recurrent episodes of abdominal pain, which had increased in frequency and severity over the last 3 months before presentation. She did not have any episodes of fever or jaundice during these episodes of pain. She was not icteric or pale and there was no palpable mass or organomegaly on clinical examination. Her liver function test was normal. But she had an episode of mild acute pancreatitis with only elevated lipase level (1988 IU/l) and no local complications secondary to the pancreatitis. Serum CA 19-9 level was normal. magnetic resonance cholangiopancreatography scan showed an impacted calculus in the remnant intrapancreatic choledochal cyst. The intrahepatic bile duct was grossly dilated because she had a type 4a choledochal cyst to begin with, but the hepatocojejunostomy was normal. Since it was a distal calculus, endoscopic retrograde cholangiopancreatography (ERCP) with stone extraction was attempted twice but the ampulla could not be visualized. On exploration, the blind end of the roux limb was adherent to the remnant cyst and the stone was impacted in the intrapancreatic remnant choledochal cyst. The adherent blind end was taken down from remnant cyst and stone was extracted. The jejunal end was reinforced in two

layers and remnant cyst was closed with 2 O vicryl in single layer interrupted sutures. She had an uneventful recovery in the postoperative period and is symptom free for 24 months now.

Figure 1: Remnant cyst restricted to intrapancreatic portion.

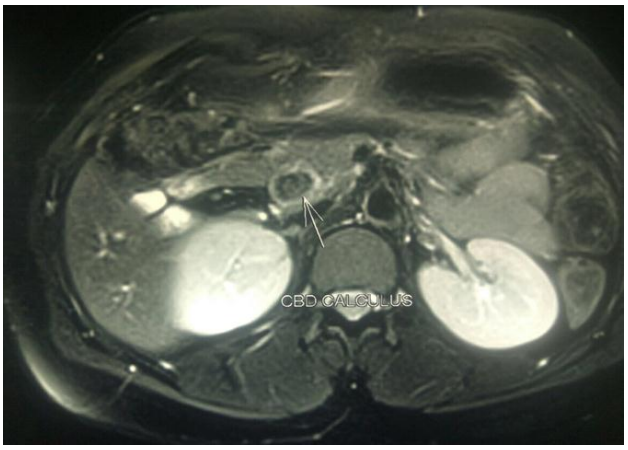

Figure 2: Remnant cyst with stone.

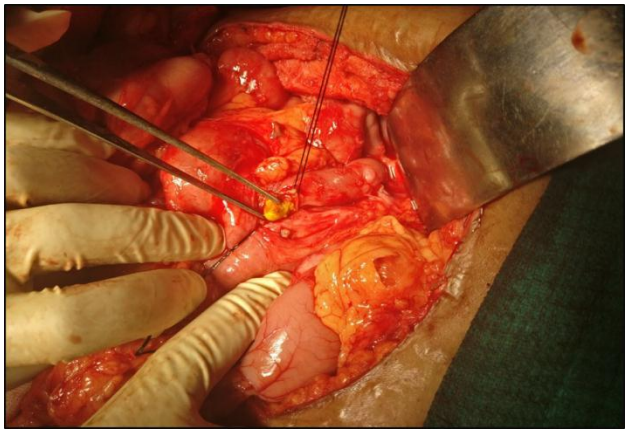

Figure 3: Stone retrieved from the remnant cyst.

DISCUSSION

Choledochal cysts are classified based on the extent of dilatation of intra and extrahepatic biliary tree. However the lower intrapancreatic part of the distal bile duct can also be grossly dilated which is not addressed in the current classification system. In such circumstances, complete excision is not feasible without Whipple procedure as there would be a very high risk for inadvertent pancreatic duct injury and pancreatic fistula. Injury to the pancreatic duct during operation may cause pancreatitis, stone formation, and leakage of pancreatic juice. Incomplete resection on the other hand can lead to stone formation, recurrent pancreatitis, and cholangiocarcinoma in remnant intrapancreatic CCs (IPCCs). Protein stones or plugs developed in remnant IPCCs in 5% and 30% patients.[1,2] Studies have reported successful outcomes for several patients with protein stones or plugs by simply using ERCP.[3]

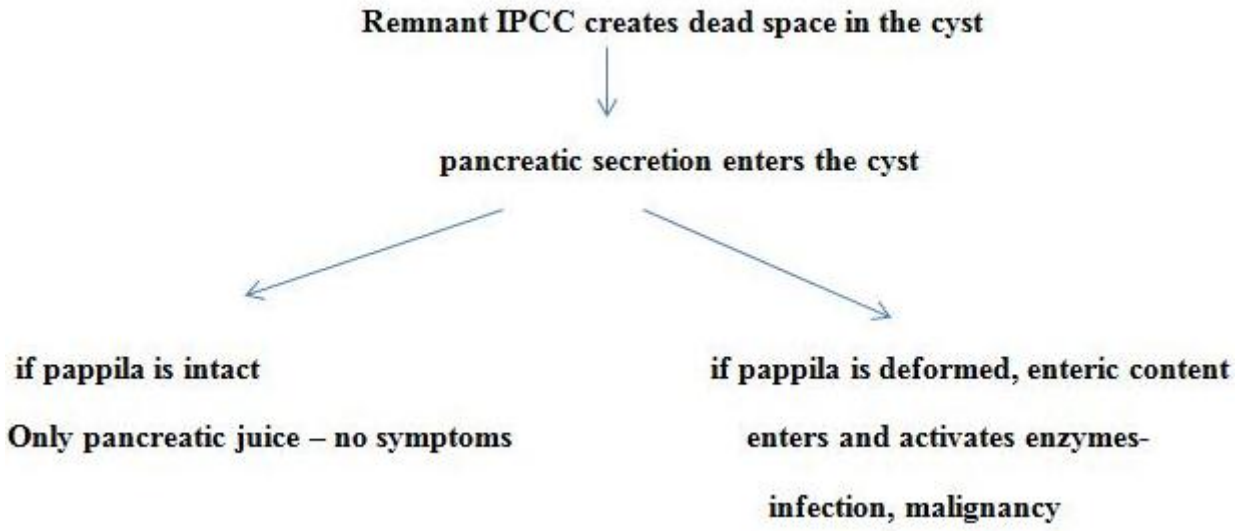

Figure 4: Mechanism of protein plug formation.

Coexistence of an anomalous union of pancreaticobiliary duct is reported in 50%–80% cases of choledochal cysts. It is important in the intrapancreatic remnant because reflux of pancreatic through the long common channel will cause protein plugs or stones in the intrapancreatic remnant choledochal cyst.

Incomplete resection of the choledochal cyst during the primary operation is the most important predisposing factor. When there is an increase in intraluminal pressure in the pancreatic duct caused by the dynamic obstruction produced by a protein plug or stone patients can have abdominal pain and can have an attack of pancreatitis. This is subsequently followed by eventual dilatation of the intrapancreatic remnant choledochal cyst. Since there is no continuity of the biliary tree with the intrapancreatic remnant, these stones are not theconventional cholesterol or pigment stones. Chemical and infrared spectrometric analyses of stones has revealed that these are protein plugs secondary to pancreatic secretions, and they are made up of albumin (more than 98%) and calcium (0.53%). These protein plugs are white and fragile. Protein plugs appear radiolucent in abdominal CT scans, and show no acoustic shadowing on EUS.[4]

The treatment options for residual intrapancreatic choledochal cyst are reoperation for complete resection, close observation, and endoscopic treatment. It is well accepted that choledochal cyst is a premalignant state, with cancer not only occurring more often in these patients but also 10–15 years earlier. The overall risk of cancer has been reported to be 10–15%, and increases with age.[5] However incidence of metachronous carcinoma after choledochal cyst excision was found to be 0.7% and risk for an individual at 15, 20, and 25 years after cyst excision was 1.6%, 3.9%, and 11.3%, respectively.[6] The risk of malignancy is also related to the type of choledochal cyst, it is higher for types I and IV than it is for types II, III, and V.[7]

When symptomatic for retained stone in the intrapancreatic remnant, ERCP and stone retrieval is the first choice of treatment for remnant choledochal cyst with symptomatic cystolithiasis. This was a rare case where stone had to be extracted operatively due to non visualisation of ampulla and failure of two attempts at endotherapy.

This patient had a remnant choledochal cyst was purely intrapancreatic and hence was not excised to avoid injury to pancreatic duct. A complete excision would have warranted a Whipple procedure but due to recent attack of pancreatitis and undilated pancreatic duct, the morbidity of Whipple procedure was very high. Hence enterotomy of the bilind loop of jejunum and opening of the intrapancreatic choledochal cyst was done to extract the stone.

This case is a rare presentation owing to stone formation in remnant intrapancreatic choledochal cyst which also caused an episode of acute pancreatitis. While complete excision of remnant choledochal cyst offers best curative option in terms of symptomatic relief and risk reduction for malignancy, complete excision is not always feasible particularly in isolated intrapancreatic remnant choledochal cyst. While there are case reports of stone extraction by ERCP for such stones, the same failed in our patient on two occasions. Hence this case is being presented for the novel approach of stone extraction via cystolithotomy without dismantling the patent hepaticojejunostom.

CONCLUSION

Remnant intrapancreatic choledochlal cyst can cause symptoms owing to stones/plugs. ERCP guided stone clearance is the preferred first choice when feasible. Risk of malignancy in remnant choledochal cyst is a realistic possibility, albeit less than an untreated choledochal cyst. When feasible surgical resection of remnant cyst should be attempted for risk reduction. In case endotherapy fails, open cystolithotomy can be done for stone extraction even if complete excision is not feasible owing to high risk and morbidty associated with Whipple procedure.

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