

Case Report

Metastatic cholangiocarcinoma following choledochal cyst excision: an unusual cause of abdominal pain in a 35-year-old female

Justin Ong, William Campbell, Mark A Taylor

Accepted 29 November 2011

INTRODUCTION

Choledochal cysts are anomalous arrangements of the pancreaticobiliary tract and a well-known aetiology for cholangiocarcinoma. The risk of cholangiocarcinoma in patients with an unresected choledochal cyst remains at 20-30%¹. Of the five types of choledochal cyst, by far the most common is the extrahepatic fusiform (Type I). Management involves complete excision of the extrahepatic bile duct and hepaticojejunostomy. This reduces the risk of cholangiocarcinoma in this group of patients to as low as 0.7%². Because of this, it is rare to encounter a patient with cholangiocarcinoma following previous choledochal cyst excision particularly when the incidence of choledochal cyst is approximately 1:100,000 in western countries³.

BACKGROUND

This 35-year-old lady was referred to the hepatobiliary clinic with symptoms of abdominal bloating, right upper quadrant abdominal pain and reflux for a number of years. At the age of 8 she had an excision of a Type I choledochal cyst with roux-en-y hepaticojejunostomy reconstruction. The symptom profile was in keeping with gastric outlet obstruction; OGD at the time revealed gastritis with excess bilious secretions seen in the stomach. Due to symptom progression and weight loss she was admitted for investigation.

Liver function tests were unremarkable but computed tomogram scan (CT) of the abdomen and magnetic resonance cholangiopancreatogram (MRCP) revealed multiple dilated intra-hepatic bile ducts with pneumobilia, raising the possibility of anastomotic stenosis. In addition however, there were dilated loops of jejunum distal to the biliary anastomosis and a large retroperitoneal mass (Figure 1). Due to progressive jaundice, biliary drainage was established via PTC in addition to core tissue biopsy of the retroperitoneal mass. Unfortunately this revealed adenocarcinoma from the pancreaticobiliary tract. As her obstructive GI symptoms progressed and to give her a chance at curative intervention, the opportunity for an exploratory laparotomy was taken where a chronically obstructed roux limb secondary to an internal hernia through the mesentery defect was found. Exploration also revealed liver metastasis, lymphadenopathy

in the mesentery and a fixed nodal mass encasing the coeliac axis. The hernia was reduced, mesenteric defect closed and the roux limb refashioned but unfortunately the tumour was unresectable due to advanced disease. Histopathology from a mesentery lymph node revealed metastatic carcinoma likely secondary to cholangiocarcinoma. The patient was referred for palliative chemotherapy and survived for 8 weeks following surgery.



Fig 1. MRCP image demonstrating grossly dilated jejunal loop (arrow) anastomosed with the common bile duct. Also seen are dilated intrahepatic ducts (star)

DISCUSSION

Choledochal cysts are single or multiple dilatations of the intra or extrahepatic biliary tree. They fall into five anatomical subtypes and are typically detected in early childhood. Standard treatment involves complete resection of the extrahepatic bile ducts with subsequent hepaticojejunostomy.

Regional Hepatobiliary Surgical Unit, Mater Hospital, Crumlin Road, Belfast BT14 7AB

Correspondence to: Justin Ong

ong_justin@yahoo.com

This prevents cholangitis, pancreatitis, liver abscesses and ultimately cholangiocarcinoma. If not excised, the risk of cholangiocarcinoma in the retained cyst is as high as 20-30% in early adulthood (by the second decade of life)¹. Within the literature, cholangiocarcinoma following previously resected choledochal cyst is a very rare occurrence. Rates vary from 0.7% - 6%². In one study of 56 patients with previous history of surgical excision, 3 patients were noted to develop cholangiocarcinoma within a range of 2 years to 19 years post surgery⁴.

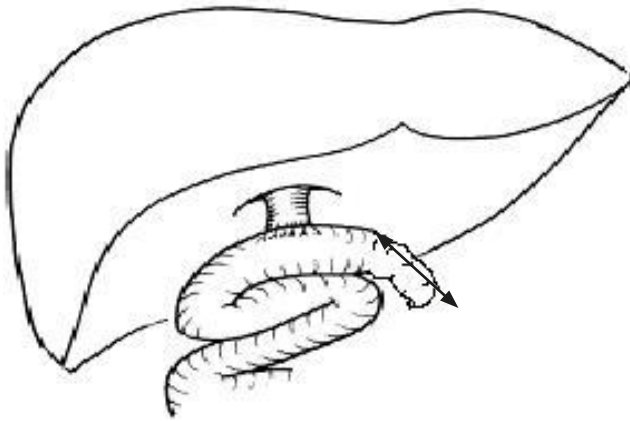


Fig 2. Arrow indicating elongation potential of blind pouch from end to side anastomosis causing potential for complications

This case is complicated however, given the evidence of chronic obstruction of the roux limb. This mechanical cause in itself may have lead to biliary stasis/reflux of enteric contents in to the bile duct thus increasing the likelihood of cholangitis and the risk of carcinogenesis. Yamataka et al. described an overall post-operative complication rate of 9% in children after excision of a choledochal cyst where stone formation/biliary stasis secondary to anastomotic stricture and bowel obstruction were the most common⁵. In a separate study by similar authors, they also stressed that the above complications post-excision in children were mostly related to elongation of the blind pouch (from end-to-side anastomosis) or redundant roux jejunal limb as they have the potential to grow or elongate in children⁶ (Figure 2). However, there were more complications seen in adult patients compared to children⁵. In the former study, majority of the patients who re-attended with complications needed a revision of their hepaticoenterostomy or exploratory laparotomy for bowel obstruction⁵.

Internal hernias of the small bowel are a recognised complication of roux-en-y bypass procedures mainly in gastric bypass surgery. However, there have been reports of small bowel volvulus secondary to internal hernia after choledocho-enteric anastomoses in transplant patients⁷. Higa et al, recommends closing all potential defects with non-absorbable sutures in a running continuous fashion⁸. Even though that study was in the setting of laparoscopic roux-en-y gastric bypass surgery, similar principals can be applied in this case.

CONCLUSION

We stress the need for close follow-up and a high index of suspicion in any patients with a previous history of a resected choledochal cyst and ongoing upper abdominal symptoms. They should be investigated aggressively with a view to ruling out the possibility of malignancy, particularly if there is evidence of obstruction or stenosis at the hepatico-enteric reconstruction. Patients with confirmed anastomotic stricture should undergo revision of either the roux-en-y hepaticoenterostomy or exploratory laparotomy if there was evidence of bowel obstruction to reduce the risk of ascending cholangitis, bile stasis or bowel obstruction which predisposes the patient to an increased risk of developing a malignancy.

The authors have no conflicts of interest

REFERENCE

1. Todani T, Toki A. [Cancer arising in choledochal cyst and management.] *Nihon Geka Gakkai Zasshi* 1996; **97** (8): 594-8. Japanese.
2. Watanabe Y, Toki A, Todani T. Bile duct cancer developed after cyst excision for choledochal cyst. *J Hepatobiliary Pancreat Surg.* 1999; **6** (3): 207-12.
3. Singham J, Yoshida EM, Scudamore CH: Choledochal cysts: part 1 of 3: classification and pathogenesis. *Can J Surg.* 2009; **52** (5): 434-40.
4. Kobayashi S, Asano T, Yamasaki M, Kenmochi T, Nakagohri T, Ochiai T: Risk of bile duct carcinogenesis after excision of extrahepatic bile ducts in pancreaticobiliary maljunction. *Surgery.* 1999; **126** (5): 939-44.
5. Yamataka A, Ohshiro K, Okada Y, Hosoda Y, Fujiwara T, Kohno S, et al. Complications after cyst excision with hepaticoenterostomy for choledochal cyst and their surgical management in children versus adults. *J Paediatr Surg.* 1997; **32** (7): 1097-102.
6. Yamataka A, Kobayashi H, Shimotakahara A, Okada Y, Yanai T, Lane GJ et al: Recommendations for preventing complications related to Roux-en-Y hepatico-jejunostomy performed during excision of choledochal cyst in children. *J Paediatr Surg.* 2003; **38** (12): 1830-2.
7. Khanna A, Newman B, Reyes J, Fung JJ, Todo S, Starzl TE. Internal hernia and volvulus of the small bowel following liver transplantation. *Transpl Int.* 1997; **10** (2): 133-6
8. Higa K, Ho T, Boone K: Internal hernias after laparoscopic Roux-en-y gastric bypass: incidence, treatment and prevention. *Obes Surg.* 2003; **13** (3): 350-4