UNCOMPLICATED ADULT CHOLEDOCHAL CYST: MANAGEMENT CHALLENGES IN A DEVELOPING ECONOMY

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ABSTRACT
Choledochal cyst is a rare congenital dilatation of the extrahepatic and/or intrahepatic billiary tract. Most patients present within the first few years of life and more than 60% of all cases are diagnosed in the first decade. Presentation in adulthood is rare and usually associated with complications of the cyst. The diagnosis of uncomplicated cases in adults requires a high index of suspicion. We present a 37-year-old female with uncomplicated adult choledochal cyst who was diagnosed intra-operatively thereby highlighting the diagnostic and management challenges in a developing economy.

Keywords: *choledochal cyst, management challenges, developing economy.*

INTRODUCTION

Choledochal cyst is a rare congenital dilatation of the extrahepatic and/or intrahepatic billiary tract. Most patients present within the first few years of life and more than 60% of all cases are diagnosed in the first decade.(1,2) Presentation in adulthood is rare and usually associated with complications of the cyst. The incidence ranges from one in 13,000 to one in two million people(3) and varies widely across countries. It is much more common in Asians and is four times more common in females than males.(4) Many theories have been proposed to explain the aetiology of choledochal cyst, however the most widely accepted theory is the common channel theory proposed by Babbit. He suggested that the cystic dilatation of the billiary tract results from an anomalous pancreatobilliary duct junction (APBJ) . Here the pancreatic duct joins the common bile duct more than 1cm proximal to the ampulla, resulting in a long common channel and free reflux of pancreatic secretion into the billiary tract leading to inflammation, ectasia and finally dilatation.(5,6) The presence of APBJ has been demonstrated by a number of investigators in many patients with choledochal cyst.(7,8)

The diagnosis can be established with ultrasound or CT scanning but may be overlooked if there is no high index of suspicion. Cholangiography (endoscopic, transhepatic or magnetic resonance) is a more specific diagnostic modality and may be required to determine the type of choledochal cyst, delineate the billiary anatomy accurately and plan extent of operative treatment. These imaging modalities are however not available in most hospitals in developing countries like ours.

We report a case of choledochal cyst in a 37-year-old woman diagnosed intraoperatively and treated by cyst excision and hepaticojejunostomy.

CASE PRESENTATION

A 37-year-old female presented to the accident and emergency unit of the Jos University Teaching Hospital with a 6-year history of recurrent episodes of right upper quadrant pain. Present episode started a week prior to presentation and was associated with an abdominal mass. There was no history of jaundice. She had earlier presented a year ago at the general outpatient department of the same hospital with dyspepsia for which she was given a series of investigations to do but defaulted.

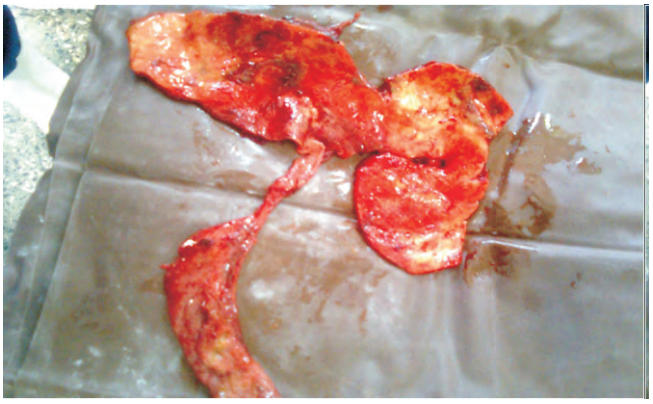
On examination, she was neither pale, febrile nor jaundiced but was in painful distress. Abdominal examination revealed a tender ill defined intra-abdominal mass occupying the right hypochondrium and epigastrium. All other systems were essentially normal.

Abdominal ultrasound scan showed a thick walled cyst containing internal echoes extending from the right kidney to the head of the pancreas, involving the porta hepatis, with normal gall bladder and liver. Laboratory investigations including full blood count, liver function tests and urea/electrolytes were within normal limits.

She was admitted and prepared for an abdominal

exploration. The findings at surgery were those of a 10 x 16cm cyst firmly adherent to the pancreas, duodenum and portal structures containing bile, with the cystic duct opening into it and continuing proximally with the billiary system (type 1 choledochal cyst).
We undertook partial excision of the cyst leaving the adherent posterior wall behind but stripping it of mucosa and a Roux-en-Y hepatico-jejunostomy was effected. Cholecystectomy was also performed. She had an uneventful post -operative period and was discharged on the 7th post-operative day. The pathologist reported the specimen as choledochal cyst with no features of malignancy. She was followed up for 5months in the surgical outpatient department after which she was lost to follow-up.

FIGURE 1: Diagram showing bisected choledochal cyst in continuity with cystic duct and gall bladder



DISCUSSION

The significant finding of this case report was that the patient presented with choledochal cyst in her fourth decade. This is an unusual age of presentation bearing in mind that this is a congenital disease that becomes symptomatic in infants and children. Agarwal D et al in his review of 30 cases of Choledochal cyst in adults showed the fourth decade to have a low age incidence of 17%.(9)
The diagnosis was easily missed pre-operatively due to the fact that symptoms of choledochal cysts are largely nonspecific, in addition to its rarity in adulthood. Also the classic triad of jaundice, right upper quadrant pain and an abdominal mass is more the exception than the rule,(10,11,12) further compounding the problem of clinical diagnosis.
Adult presentation is characterized by complications such as pancreatitis, cholangitis and cholangiocarcinoma(13) which symptoms were largely absent in this patient. A palpable mass is rare in adults,(14,15) but this patient had a palpable mass.
Ultrasound is usually the first imaging modality due to its availability and affordability. It is sensitive in the detection of cystic structures but rather nonspecific in identifying their origin. In the index patient, the origin of the cyst could not be clearly defined and other differentials such as pancreatic pseudocyst was suggested by the radiologists. A CT was not done because it was not available at that time.
The diagnosis of type 1 choledochal cyst was made on table, according to the Todani modification of the Alonso-Lej classification.(16,17) See Table 1.

Table 1. Todani modification of the Alonso-Lej classification

Type I	Fusiform dilation of the extrahepatic bile duct
Type II	Single saccular dilation or diverticulum of the extrahepatic bile duct
Type III	Dilation of the intraduodenal portion of the bile duct (choledochocele)
Type IVa	Combined intahepatic and extrahepatic dilation of the bile duct
Type Ivb	Multiple dilation of the extrahepatic bile duct
Type V	Isolated or diffuse intrahepatic biliary dilation (Caroli's disease when associated with hepatic fibrosis)

The two basic treatments described for choledochal cysts are cyst enterostomy and cyst excision with hepaticojejunostomy. Cyst enterostomy is associated with late complications including anastomotic stricture, billiary calculi, recurrent cholangitis and malignant degeneration of the retained cyst wall(1,18,19) and is mentioned to be condemned. For this patient, cyst excision with Roux-en-Y hepaticojejunostomy is the preferred option though technically demanding.(20,21) Sometimes, circumferential dissection of the cyst wall may not be achievable as in the index patient without the risk of injury to the hepatic artery and portal vein. This may be seen when as a result of repeated inflammation the posterior wall of the cyst is densely adherent to these structures. In such situations, Lilly described the technique of entering into the cyst by opening it anteriorly and excision of the mucosa of the cyst leaving behind a portion of the posterior wall of the cyst(22) which is what was done in the index case. The stripping of the mucosa of the posterior wall of the cyst left behind is important to reduce the risk of malignant transformation occuring in the posterior wall of the cyst.(23)

CONCLUSION

This case represents the difficulties in making a diagnosis of choledochal cyst in adults, its unique presentation and the operative challenges.

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