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Spontaneous gangrene and perforation of choledochal cyst: a rare presentation

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ABSTRACT

Choledochal cyst is a rare entity. It generally presents in children with pain, jaundice and the occasional finding of a lump in right hypochondriac region. A very rare presentation is biliary peritonitis resulting from a perforated choledochal cyst. It is usually seen in younger children. Our patient was a 14 year old female who presented with an acute abdomen. Clinically she had signs of peritonitis. Chest radiograph did not show free gas under diaphragm. The USG showed a dilated CBD of 4.1cm with a calculus at its lower end of 2 x 2.6cm, USG guided diagnostic tap revealed bile. Emergency exploration was done, a gangrenous patch of size 3 * 2cm in the anteromedial wall of large choledochal cyst was found with a perforation in it. No calculous was found. The gangrenous wall of choledochal cyst was excised and primary suturing of choledochal cyst was done with a T-tube placed through separate incision in choledochal cyst. As there is no facility for intra-operative cholangiogram in the emergency setting in our institute, a post-operative cholangiogram was done on day 7. It revealed a dilated CBD, CHD as well as right and left hepatic ducts (choledochal cyst type IA). The patient was discharged and asked to follow-up electively for excision of choledochal cyst at a later date. A rare possibility of a perforated choledochal cyst should be kept in mind in cases of biliary peritonitis, especially in younger age groups. Bile drainage would be a safer procedure in emergency condition, especially when patient presented late to hospital with hemodynamic unstability and edematous wall.

Keywords: Choledochal cyst, Perforation, T-tube cholangiogram, Gangrene

INTRODUCTION

Choledochal cvsts are uncommon congenital malformations described typically in childhood.¹ Classical triad of presentation of choledochal cyst is of pain, jaundice and abdominal mass, but are present only in 10% to 25% of cases. Diagnosis of choledochal cyst is made based on a disproportionate dilation of extra hepatic biliary duct.² They are definitively treated by excision of the cyst together with a part of bile duct and Roux-en-Y hepaticojejunostomy. The most common choledochal cyst is type I, with diffuse or segmental fusiform dilatation of the common bile duct. Perforation of a choledochal cyst is a very rare complication but can sometimes be the initial manifestation and reported to occur in 1-2% of cases in large series. Most reported cases have been managed with external drainage of the cyst followed by a second procedure to excise the cyst and reconstruct the biliary tract. 1,3,5

CASE REPORT

A 14year old female presented to us in emergency department with the complaints of pain and distension of abdomen since 2 days. Pain was severe, continuous and generalised all over abdomen. Associated complaints of nausea and two episodes of vomiting was present. The patient was febrile with tachycardia, hypotension and

signs of dehydration, pale and non-icteric. Examination revealed tenderness, guarding and rigidity all over abdomen.

A provisional diagnosis of perforation peritonitis was kept.

Investigations

- 1. Chest X ray and X-ray Abdomen standing were normal.
- USG Abdomen: Findings suggestive of gross intraperitoneal collection with tap suggestive of biliary collection. CBD dialated up to 4.1 cm with CBD calculus measuring 2 cm. No intrahepatic biliary radicles dilatation. Bowel loops were peristaltic. Solid organs were normal.
- 3. Repeat X ray chest and abdomen after RT insertion and insufflating air was normal.

Final Diagnosis

Biliary peritonitis cause with sepsis and shock.

Operative Procedure

Upper abdominal laparotomy was done. About one litre of bilious collection was drained. Intraoperative findings revealed a grossly dilated common bile duct suggestive of choledochal cyst with a gangrenous patch over anteromedial aspect of choledochal cyst proximal to insertion of cystic duct of approx. size 3*2 cm with perforation in centre of about 6*6mm. Choledochal cyst was explored and it did not reveal any intraductal calculus. Cholecystectomy was done. The gangrenous patch in choledochal cyst was excised and primary suturing of choledochal cyst was done over a 'T' tube. As there is no facility for intra-operative cholangiogram in the emergency setting in our institute, intraoperative cholangiogram could not be performed.. Specimen was sent for histopathological examination. Abdominal drain was placed in right sub hepatic space and abdomen was closed.

Post-operative cholangiogram

Done on post-operative day 7, it revealed a dilated CBD, CHD as well as right and left hepatic ducts (choledochal cyst type IA).

Histopathological examination report

Gross: Cut open structure of size 2 X 1 X 0.5 cm labelled as choledochal cyst gangrenous patch.

Cut open gall bladder of size 8.5 X4.5X0.5 cm

Microscopy: Sections from gangrenous patch shows gangrene.

Sections from gall bladder shows evidence of chronic cholecystitis

Bile culture and sensitivity report

No growth.

Final diagnosis

Biliary peritonitis secondary to Gangrene and perforation of Choledochal cyst type IA.

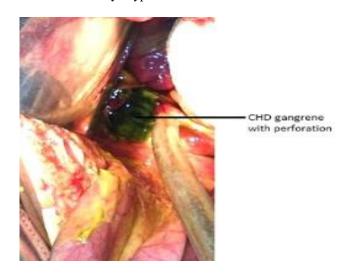


Figure 1: Choledochal cyst gangrene with perforation tube in situ.

In our case it was possible to approximate the wound edges and primary closure was achieved. CBD exploration was done through a separate incision in the CBD to rule out presence of calculi mentioned in the USG report. T-tube was clamped after output was less than 50ml on day 5 and was removed on day 12. The sub hepatic drain was removed on day 14. Patient was discharged on post-operative day 15 and was asked to follow-up for elective surgery for excision of choledochal cyst after 6 weeks.

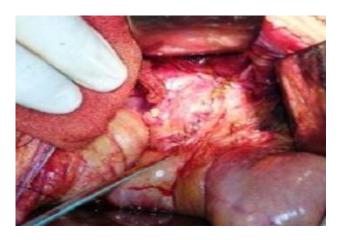


Figure 2: Sutured CBD and sutured CHD with T.

DISCUSSION

Choledochal cysts are uncommon congenital malformations described typically in childhood. The incidence in the population is 1:5000 and is three times higher in females. 2,3 Infants and children may develop pancreatitis, cholangitis, and histologic evidence of hepatocellular damage. Adults in whom subclinical ductal inflammation and biliary stasis may have been present for years may present with one or more severe complications, such as hepatic abscesses, cirrhosis, portal hypertension, recurrent pancreatitis, and cholelithiasis. Cholangiocarcinoma is the most feared complication of choledochal cysts, with a reported incidence of 9-28%.



Figure 3: T tube changingram showing dilated CBD, CHD as well as right hepatic ducts.

Spontaneous rupture of choledochal cyst had been considered as rare.⁵ Since rupture can sometimes be the initial manifestation of the disease, it should be considered in the presence of bile-like fluid at the time of emergency laparotomy, especially in a young age. The etiology remains unknown in majority of cases. The suggested explanations for spontaneous perforation include extension of those proposed in the pathogenesis of choledochal cyst in general and are a mix of congenital and acquired factors i.e. pancreticobiliary malunionpancreticobiliary reflux and epithelial irritation, distal obstruction in the common pancreaticobiliary channelanatomic or with inspissated protein plugs, mural immaturity and bile duct ischemia.

Most reported cases have been managed with external drainage of the cyst followed by a second procedure to excise the cyst and reconstruct the biliary tract as in our case, but many researchers recommend primary reconstructive surgery as the treatment of choice. ^{1,3,5,6,9} In developing countries, patients usually present late with much of septic load which do not allow for primary reconstructive surgery. In stable patient even though it has been suggested that reconstructive surgery may be tolerable, definitive surgery should be regarded as a of procedure with some risk post-operative

complications. Availability of the specialist concerned is also a limiting factor for primary reconstructive repair, especially in third world countries.

In most cases, there is only a small hole on the anterior wall of the cyst in cases of cyst rupture; but in a few cases, especially in those who present late, the whole of the anterior wall gets sloughed off after necrosis as in our case. All workers recommend the use of T tube drainage of the perforation in the choledochal cyst as was done in our case. However, in some cases due to the delayed presentation with biliary peritonitis, the region of the extra-biliary tree may be so oedematous and friable that people have resorted to cholecystectomy with excellent outcome. There is also consensus that excision of the perforated choledochal cyst should not be attempted at the initial exploration and excellent outcome is obtained with delayed excision of the choledochal cyst after an interval of one to four months.¹³

CONCLUSION

Bile drainage would be safer procedure in emergency condition, especially when patient presented late to hospital with hemodynamic instability and oedematous wall. Our method of putting T-tube and primary repair in case presenting late and having complete loss of the anterior wall hence minimizing the chances postoperative leak of bile and buying the time for definitive repair, thereby subsequently reducing the morbidity and mortality in these patients.

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institutional ethics committee

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