Type IVa choledochal cyst: Results following haemihepatectomy and mucosectomy of intrahepatic cyst

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Abstract Choledochal cyst is a rare condition characterised by congenital dilatation of the biliary tree. Commonly seen in the oriental countries, patients usually present with a varying combination of abdominal pain, jaundice, lump or cholangitis. Untreated patients or incomplete removal of the cysts usually leads to portal hypertension and cholangiocarcinoma. Almost one-third of the cases have intrahepatic cyst, making complete cyst excision often impossible. We are reporting a 9-year-old girl with type IVa choledochal cyst presented to us with recurrent pain abdomen and intermittent jaundice for 1 year. Excision of extrahepatic cyst, left hepatectomy, mucosectomy of the residual cyst wall of right lobe of the liver and a wide bilio-enteric anastomosis was done. Patients followed up with an magnetic resonance cholangiopancreatography (MRCP) 2 months later showed shrinkage of the residual cyst and good bile drainage. Mucosectomy of intrahepatic cyst may prevent recurrent cholangitis, calculus formation and cholangiocarcinoma in the long run.

Keywords Type IVa choledochal cyst · Hepatectomy · Mucosectomy

Introduction

Choledochal cysts are rare congenital anomalies of the biliary tract. Although very rare in the western countries, this condition is seen frequently in the oriental countries [1]. Todani et al. classified choledochal cyst into five different types [2]. Type I being the commonest comprising almost two-thirds of the cases followed by type IV cysts. Treatment of the intrahepatic cysts is a challenging problem and are associated with complications even after wide bilio-enteric anastomosis. Hepatic lobectomy can be done if cysts are restricted to one side of the liver. The residual intrahepatic cyst can be cause of recurrent cholangitis, calculus formation and cholangiocarcinoma. Excision of the mucosa of the residual intrahepatic cyst may eliminate these risk in later life.

Case report

S. D., a 9-year-old girl presented in paediatric surgery Out-patient Dept. with recurrent colicky, right upper abdominal pain with intermittent jaundice for 1 year. There was no associated fever or vomiting. Her growth and development were normal. There was no pallor or icterus. Deep tenderness was present in the right upper quadrant without any palpable lump. Liver and spleen were not palpable and examination of other systems were also found to be normal.

Liver function tests revealed normal serum bilirubin and hepatic enzyme levels except raised gamma glutamyl transpeptidase (GGTP) (191 U/l). Ultrasonography of the hepatobiliary system showed cystic dilatation of both extra and intrahepatic biliary tree and a suggestion of distal common bile duct (CBD) stricture was put forward. An MRCP revealed huge cystic dilatation of extra and intrahepatic

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bile ducts with predominantly left lobar involvement. The pancreatobiliary junction was found to be normal. Patient was explored through a right thoracoabdominal incision. The extrahepatic cyst was excised (Fig.1). Left hepatic lobectomy was done. Cyst in the right hepatic lobe was opened widely and mucosa of the cyst was excised in piecemeal. A Roux-en-Y hepaticojejunal anastomosis was performed. The girl recovered uneventfully and was discharged on the 11th postoperative day.

A follow up MRCP scan 2 months after surgery revealed significant shrinkage of the residual cyst size with good biliary drainage. Patient has been well till last follow up at 2 years after surgery. The histology of the resected cyst showed chronic inflammatory changes.

Discussion

Choledochal cysts are focal or diffuse dilatation of the biliary tree that are believed to be congenital in origin. Although rare in western countries, choledochal cyst are frequently seen in Asian countries, especially in Japan.

Alonso-Lej et al. first classified choledochal cyst into three major types: cystic, diverticular and choledochocoele [1]. Todani et al. modified the classification system, which currently includes five types based on cholangiographic morphology and number of intrahepatic and extrahepatic bile duct cysts [2]. With regard to the shape of extrahepatic and intrahepatic ducts, type IVa is further classified into three subtypes: cystic-cystic, cystic-fusiform and fusiform-fusiform [2].

Type IVa cysts are observed in 28.5% of the cases reported in a survey by the Japanese Society of Paediatric Surgeons [1]. In our institute, during the last 10 years 14

cases of choledochal cyst were seen, only one of them had type IV cyst (7.1%), the rest were cystic in 8 (57.2%), fusiform in 4 (28.6%) and choledochocoele in 1 (7.1%).

Surgery is the only treatment of choledochal cyst. Early surgical intervention protocols consisted of drainage and enterostomy [3]. Complications occurring with this approach included anastomotic strictures, calculi associated with stasis, recurrent cholangitis and most importantly, malignant disease. Currently, total excision with reconstruction of the biliary tree by means of hepaticojejunostomy is considered to be the treatment of choice. This approach reduces the risk of malignant disease by 60–70% [4]. In patients with severe pericystic inflammation and adherence to adjacent hepatic vessels, the excision of the inner layer of the cyst or mucosectomy, the technique originally described by Lilly, becomes useful [5]. In this procedure, cyst excision is accomplished using a plane of dissection between the inner and outer layers of the cyst beginning at the confluence of the cystic and the common ducts.

Treatment of intrahepatic cyst is a challenging task. It is usually impossible to remove significant portion of intrahepatic cysts but biliary drainage is improved by excision of parts of dilated right and left hepatic ducts and wide biliary enteric anastomosis. The intrahepatic dilatation tends to gradually reduce in size following sufficient biliary drainage. In addition, lateral hepatic segmentectomy may be necessary, especially in cases of strictures of the upper intrahepatic ducts and when the left hepatic lobe contains stones [6].

If the dilatation is restricted to one side of the liver, lobectomy should be done. Unroofing with drainage into a Roux loop of jejunum may be required in presence of proximal ductal obstruction. Remnant of intrahepatic cyst will cause recurrent bouts of cholangitis, biliary cirrhosis,

Fig. 1 Extrahepatic cyst with dilated common hepatic ducts

Fig. 2 Cyst wall with thickened and inflamed mucosa

Figs. 3 and 4 Preoperative and postoperative MRCP

proximal ductal ectasia and calculus formation. Incidence of postoperative cholangitis after definitive cyst excision and Roux-en-Y hepaticojejunostomy ranges from 23% to 40% in type I cyst and 31% to 44% in IVA cysts [6].

Several studies have reported malignant degeneration of the cyst wall in cases where cyst excision was not done [2, 7]. The incidence of cancer in a choledochal cyst is 20 times greater than that in the general population. Fourteen to 18% of the adult population with a choledochal cyst will develop cancer in a cyst if the cyst is not removed. Theoretically, total or intramural cyst excision should prevent malignant degeneration of the cyst wall in the long run.

Considering the high incidence of complications which can arise in the untreated or residual cyst, it becomes mandatory to do complete cyst excision, or in difficult situations, mucosectomy of the cyst. However, mucosectomy is not described for intrahepatic cyst.

In the present patient, after doing left lobectomy of the liver, we observed that the opened up cyst of the right lobe had very thickened and friable mucosa (Fig. 2), which in all probability would be a cause of recurrent cholangitis or malignant changes in the future. We also observed that the bile ducts in the rest of the right lobe were of normal calibre. So we decided to remove the abnormal mucosa, as described by Lilly et al. for extrahepatic choledochal cyst with pericystic inflammation. A wide hepaticojejunal anastomosis was performed. The patient had uneventful recovery. Follow up MRCP showed significant shrinkage of the cyst wall, a sign of good biliary drainage (Figs. 3 and 4). At two years follow up, the patient was asymptomatic with normal liver function test.

Conclusion

Treatment of type IV choledochal cyst is a difficult problem. Recommended treatment is excision of extrahepatic cyst with wide biliary enteric anastomosis. Cyst localised to a segment or lobe of the liver will require a segmentectomy or lobectomy. However, risk of postoperative complications always exist because of the residual intrahepatic cyst. Excision of the mucosa of the intrahepatic cyst may reduce the risk of the postoperative complications.

References

1. Alonso-Lej F, Rever WB Jr, Pessagno DJ (1959) Congenital choledochal cyst, with a report of 2, and an analysis of 94 cases. *Int Abstr Surg* 108:1–30
2. Todani T, Watanabe Y, Toki A, Morotomi Y (2003) Classification of congenital biliary cystic disease: special reference to type Ic and IVA cysts with primary ductal stricture. *J Hepatobil Pancreat Surg* 10:340–344
3. Lopez RR, Pinson CW, Campell JR, et al. (1991) Variation in management based on type of choledochal cyst. *Am J Surg* 161:612–615
4. Scudamore CH, Hemming AW, Teare JP, et al. (1994) Surgical management of choledochal cysts. *Am J Surg* 167:479–500
5. Lilly JR (1979) The surgical treatment of choledochal cyst. *Surg Gynecol Obstetr* 149:36
6. Lenriot JP, Gigot JF, Segol P, Fagniez PL, Fingerhut A, Adloff M (1998) Bile duct cysts in adults: a multi-institutional retrospective study. *French Associations for Surgical Research. Ann Surg* 228:159–166
7. Tsuchiya R, et al. (1978) Malignant tumours in choledochal cyst. *Ann Surg* 186:22