The THAI Journal of SURGERY 2011;32:80-83.
Official Publication of the Royal College of Surgeons of Thailand

Case Report

Malignant Choledochal Cyst : A Case Report and Reviews

Pichai Boonyongsunchai, MD[1]
Chutcharn Kongphanich[2]
Yuthapong Srikoch[3]

[1]Department of Surgery, [2]Department of Radiology, [3]Department of Pathology, Phramongkutklao Hospital and College of Medicine, Bangkok, Thailand

Abstract **Choledochal cysts (CDC) are uncommon biliary lesions with well documented congenital dilatations of the intra and/or extrahepatic biliary trees that carry significant morbidity if not recognized and treated early. This case report described the long history of abdominal dyspepsia in a 44-year-old woman who had a cholangiocarcinoma in a CDC diagnosed with magnetic resonance cholangiopancreatography (MRCP) and subsequently underwent hepatic hilar resection with Whipple operation. It illustrated the need for early diagnosis and complete excision of a CDC to prevent their complications such as cholangitis and malignant transformation.**

Key words: **Cholangiocarcinoma, cholangitis, choledochal cysts**

INTRODUCTION

A choledochal cyst (CDC) was firstly described by the German anatomist Abraham in 1725[1]. Focal dilatation of biliary tree consistent with the modern concept of CDC was described by Douglas in 1852, the disease remained difficult to define and categorize until the classification of Alons-Lej et al and Todani et al. The incidence in western countries varies between 1 in 100,000-150,000 individuals. The rate of incidence is higher in Asia and more frequently occurs in woman (1 male: 4 female). The diagnosis of CDC is often made in childhood and 25% are initially seen in adults. The most common symptoms of CDC are abdominal pain, jaundice and abdominal mass. Abdominal pain is the most common presenting symptom in adult cases and most had nonspecific clinical symptoms.[3,6] The importance of the condition lies in their complication, of which carcinoma is the most serious condition . Carcinoma that occurred in CDC had been reported about 2.4-14% and varied considerably with age.[2]

Correspondence address : *Pichai Boonyongsunchai, MD, Department of Surgery, Phramongkutklao Hospital and College of Medicine, Bangkok, Thailand; Telehone: +66 2354 7711; Fax: +66 2354 7707; E-mail: pichai3798@gmail.com*

Case presentation

A 44-year-old woman was referred to Phramongkutklao Hospital because of abdominal discomfort and jaundice for one month. She had a history of recurrent dyspepsia for 5-10 years and received medication from a local hospital. She was diagnosed with obstructive jaundice which may be caused by pancreatic cancer. On physical examination, she had normal vital signs ,deep icteric skin and sclera, palpable distended Gallbladder at right subcostal area with mild tenderness. The laboratory tests revealed 22.39 mg% total bilirubin and 20.35 mg% direct bilirubin. Abdominal ultrasound and magnetic resonance cholangiopancreatography (MRCP) showed a large heterogeneous mass in dilated common bile duct size about $7 \times 5 \times 11$ cm. with extension of dilatation to intrahepatic duct which suggestive of malignant CDC type IVa (Figure 1).

A preoperative diagnosis of CDC with carcinoma was made and the operation was performed. The whole dilated common bile duct was contained with tumor extended from distal common bile duct to common hepatic bifurcation and left main hepatic bile duct (Figure 2) and frozen section suggested adenocarcinoma. Cholecystectomy, hepatic hilar resection including standard pancreaticoduodenectomy was performed. This patient recovered uneventfully in 10 days and was discharged 12 days after surgery.

Histopathology showed dilated common bile duct measuring $8.5 \times 7.0 \times 6.0$ cm. contained poorly differentiated adenocarcinoma with signet ring cell feature and one lymph node had malignancy. This confirmed cholangiocarcinoma in the CDC.

Discussion

CDC is a rare type of bile duct cyst of uncertain origin. The majority of cases reported are young women and children of Asian descent, especially in Japan and Taiwan. In North America, its incidence is estimated to be 1/150,000 and the most common

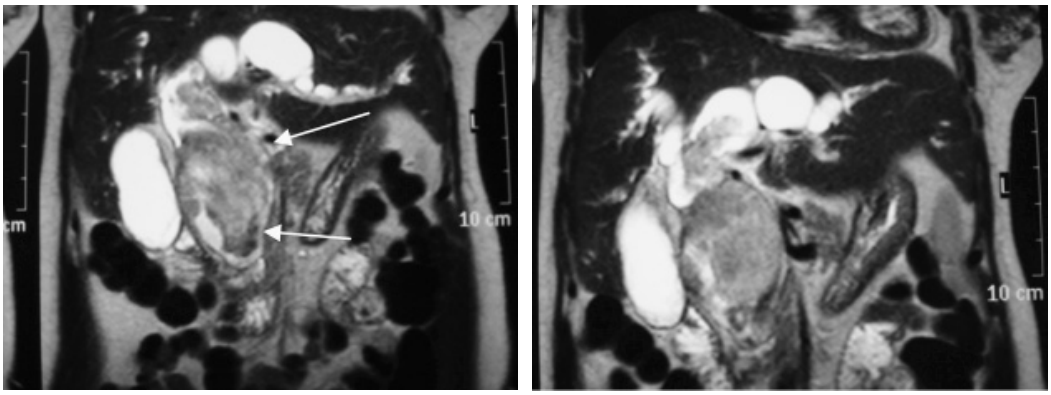

Figure 1 Heterogenous mass in choledochal cyst (Arrow)

Figure 2 Dilated Gallbladder (short arrow) : Cholangiocarcinoma in fusiform dilated CBD (long arrow)

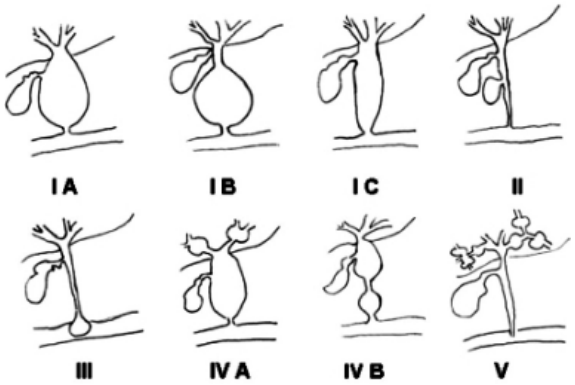

Figure 3 Modified Todani's classification cyst of choledochal cyst

symptoms of CDC are abdominal pain, jaundice and abdominal mass.[4,9] Congenital CDCs were firstly classified into three types by Alonso-Lej et al in 1959 . Todani and colleagues modified this classification in 1977 adding types IV and V and more recently Todani revised it further to reflect the presence of pancreaticobiliary maljunction.[2-5] Type IV cysts are more commonly observed in adults than in children, while type I cysts are more commonly observed in Asian patients.[7,8] In our report, this patient had CDC consistent with Type IVa which had fusiform dilation of the entire extrahepatic bile duct with extension of dilation of the intrahepatic bile ducts.

The Todani classification of bile duct cysts divides cysts of the bile duct into five groups[14,24]. (Figure3)

Type 1: Also known as a true CDC

- Account for 80-90% of all bile duct cysts and characterized by fusiform dilation of the extrahepatic bile duct. Theorized that choledochal cysts form as the result of reflux of pancreatic secretions into the bile duct via anomalous pancreaticobiliary junction.
  - Ia : dilatation of extrahepatic bile duct (entire)
  - Ib : dilatation of extrahepatic bile duct (focal segment)
  - Ic : dilatation of the common bile duct portion of extrahepatic bile duct

Type 2: Also known as a bile duct diverticulum.

- Account for 3% of all bile duct cysts and represent a true diverticulum.
- Saccular outpouchings the supra-duodenal extrahepatic bile duct or the intra-hepatic bile ducts.

Type 3: Also known as a choledochocele.

- Account for 5% of all bile duct cysts and represent protrusion of a focally dilated, intramural segment of the distal common bile duct into the duodenum. Choledochoceles may be successfully managed with endoscopic sphincterotomy, surgical excision, or both, in symptomatic patients.

Type 4: Multiple communicating intra- and extrahepatic duct cysts.

- second most common type of bile duct cysts (10%) and subdivided into sub-types A and B
  - type 4a : fusiform dilation of the entire extrahepatic bile duct with extension of dilation of the intrahepatic bile ducts
  - type 4b : Multiple cystic dilations involving only the extrahepatic bile duct

Type 5: Also known as Caroli disease which is a rare form of congenital biliary cystic disease manifested by cystic dilations of intrahepatic bile ducts. It is also association with benign renal tubular ectasia and other forms of renal cystic disease.

It is well accepted that a CDC is a premalignant state which occurs at the site of bile stasis, irritation and inflammation within the dilated cyst in CDC and within the Gallbladder when no cyst exists. Chronic inflammation in CDC also destroys the protective mucin-producing epithelial cells and chronic infection by gram negative bacteria such as *E. Coli* metabolized bile acid into carcinogen. The overall risk of cancer has been reported to be 10%-15% and increases with age. The risk rises from 2.3% in patients aged 20-30 years to 75% in patients aged 70-80 years.[15] The most common types of cancer in patients with CDC are adenocarcinoma 73%-84% as presented in our patient and anaplastic carcinoma 10%, undifferentiated carcinoma 5%-7%, squamous cell carcinoma 5% and other carcinoma 1.5%.The site of cancer is the extrahepatic bile duct 50%-62% of patients, gallbladder in 38%-46%, intrahepatic duct in 2.5%, the liver and pancreas in 0.7%.[16-19] Management of CDC depends on the type of cyst. The incidence of cholangiocarcinoma with non-cyst excision or non-operated CDC was 10.8%. So complete excision of CDC for types I, II and IVb with hepaticojejunostomy is the treatment of choice for CDC in adult patients. Postexcisional malignant disease, which has an incidence of 0.7-6% is thought to be due to remnant cystic tissue or subclinical malignant disease not detected before surgery. Therefore, most authors recommend intraoperative ultrasound and pathology

of frozen section to rule out malignant disease.[10-12, 20-23] Malignant disease within the biliary tree as in malignant CDC should be managed by excision of extrahepatic bile duct and adjacent liver including regional lymph nodes. In the case of distal malignant disease adjacent or within the pancreatic head requires a Whipple operation as in our case with curative intent was done by combined hepatic hilar resection and Whipple operation with regional lymph nodes clearance. Unfortunately, less than 10% of cancers are resectable at diagnosis due to metastatic disease that affects the surrounding vasculature, organ or peritoneum. A prophylactic therapeutic bile duct stent placement or gastroenterostomy to bypass the affected enteric tract and relieve obstruction may be necessary in unresectable cases. Adjuvant chemo-therapy or radiotherapy may increase survival and improve quality of life , although prognosis of cancer is very poor.

Conclusion

This case report demonstrates the diagnostic and therapeutic pitfalls to prevent cost-intensive and potentially life-threatening complications. A CDC must be considered in the differential diagnosis in all patients of any age who have an abnormal bile duct cystic dilatation and should be considered as being at risk of future malignant degeneration. Excision is not by itself protection against future development of cancer. However, liver transplantation should be considered for the patients with extensive intrahepatic cysts. All patients should have life long follow up usually with ultrasonography and monitoring of liver enzymes especially in any adult with recurrent symptoms following cyst-related surgery and must be evaluated carefully for malignancies anywhere in the biliary tract.[12-14]

REFERENCES

1. Dabbas N, Davenport M. Congenital choledochal malformation: not just a problem for children. Ann R Coll Surg Engl 2009;91:100-5.
2. Dowsett JF, Rode J, Chandiramani VA, et al. Occult carcinoma in an adult choledochal cyst. Post grad Med J 1991;67:202-5.
3. Lee HK, Park SJ, Yi BH, et al. Imaging features of adult choledochal cysts: a pictorial review. Korean J Radiol 2009; 10:71-80.
4. Wang Q-G, Zhang S-T. A rare case of bile duct cyst. World J Gastroenterol 2009;15:2550-1.
5. Visser BC, Insoo Suh BS, Way LW. Congenital choledochal cysts in adults. Arch Surg 2004;139:855-62.
6. Singhak J, Yoshida EM, Scudamore CH. Choledochal cysts. Part 1 of 3 : classification and pathogenesis. Can J Surg 2009;52:434-40.
7. Gigot J, Nagorney D, Farnell M, et al, Bile duct cysts: a changing spectrum of disease. J Hepatobiliary Pancreat Surg 1996;3;405-11.
8. O'Neill JA. Choledochal cyst. Curr Probl Surg 1992;29:361-410.
9. Singhak J, Yoshida EM, Scudamore CH. Choledochal cysts. Part 2 of 3: diagnosis. Can J Surg 2009;52:506-11.
10. Singhak J, Yoshida EM, Scudamore CH. Choledochal cysts. Part 3 of 3: management. Can J Surg 2010; 53:51-6.
11. Zheng L-X, Bo H, Wu D-Q, et al. Experience of congenital choledochal cysts in adults. J Korean Med Sci 2004;19:842-7.
12. Krissat J, Bismuth H. Choledochal cystic malignancies. Ann Oncol 1999:94-8.
13. Dhupar R, Gulack B, Geller DA, et al. The changing presentation of choledochal cyst disease: an incidental diagnosis. HBP Surg 2009:1-4.
14. Yu J, Turner MA, Fulcher AS, et al. Congenital anomalies and normal variants of the pancreaticobiliary tract and the pancreas in adults: part 1, biliary tract. Am J Roentgenol 2006;187:1536-43.
15. Feng JF, Chen WY, Chen DF, et al. Choledochal cysts with malignancy in adult: a retrospective study with an experience of twenty -two years. Pak J Med Sci 2011;27: 6-10.
16. Okada A, Hasegawa T, Oguchi Y, et al. Recent advances in pathophysiology and surgical treatment of congenital dilatation of the bile duct. J Hepatobilary Pancreat Surg 2002;9:342-5.
17. Todani T, Watanabe Y, Toki A, et al. Carcinoma related to choledochal cyst with internal drainage operations. Surg Gynecol Obstet 1987;164:61-4.
18. Todani T, Tabuchi K, Watanabe Y, et al. Carcinoma arising in the wall of congenital bile duct cysts. Cancer 1979;44: 1134-41.
19. Fieber SS, Nance FC. Choledochal cyst and neoplasm: a comprehensive review of 106 cases and presentation of two original cases. Am Surg 1997;63:982-7.
20. Kobayashi S, Asano T, Yamasaki M. Risk of Bile duct carcinogenesis after excision bile ducts in pancreatobiliary maljunction. Surgery 1999;126:934-44.
21. Franko J, Nussbaum ML, Morris JB. Choledochal cyst cholangiocarcinoma arising from adenoma: case report and review of the literature. Curr Surg 2006;63:281-4.
22. Di Sena V, de Paulo GA, Macedo EP, et al. Choledochal cyst mimicking a pancreatic pseudocyst: case report and review. Gastrointest Endosc 2003;58:620-4.
23. Atkinson HDE, Fischer CP, de Jong CHC, et al. Choledochal cysts in adults and their complications. HPB 2003;5:105-10.
24. Lenriot JP, Gigot JF, Segpl P, et al. Bile duct in adults; a multi-institutional retrospective study. Ann Surg1998;228:159-66.