

Choledochal Cyst in an Adult – A Case Report

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Abstract

Choledochal cyst, an anomalous dilatation of the biliary tract, is a congenital disease. However, a very few cases of this condition have been diagnosed in adults. We report a case of a 58-year-old man with symptoms related to biliary tract pathology. On investigations, he was found to have a type-II choledochal cyst.

Keywords: *choledochal cyst, biliary tract*

Introduction

Choledochal cyst is a rare congenital cystic dilatation of the extrahepatic and/or intrahepatic biliary tree. The incidence is between 1:100,000 and 1:150,000. The choledochal cyst affects females three to eight times more often than it does males. This disease is common in Japan, where most of the cases were reported.^{1,2} Almost 25% of choledochal cysts are detected in the first year of life, whereas 50% of the reported cases are diagnosed in the first decade of life.^{3,4} The detection is delayed in approximately 20% of the cases, in which the patients are diagnosed in adult life, when symptoms related to the biliary tract pathology appear.

Case Report

Mr.T, a fifty-eight-year-old heavy smoker, was admitted to the Surgical Department Immanuel Hospital in June 2009, with a history of jaundice for six months. The patient denied any history of associated fever or abdominal pain.

On the physical examination, he was icteric, characteristic of obstructive jaundice. The liver was palpable three fingers in breadth below the costal margin. However, there was no palpable mass. The cardiovascular and respiratory functions were clinically normal. The urine was dark colored, which the patient described as “cola colored.” The stools were pale and his total bilirubin serum level was 10.4 mg/dl and direct bilirubin was 7.0 mg/dl. All other laboratory findings were within normal parameters. We also found congenital disorder of his fingers which posed some irregular deformities.

Abdominal ultrasound (USG) showed a slight hindrance of intrahepatic and extrahepatic bile duct due to multiple choledocholithiasis, with suspected chronic cholecystitis. Endoscopic Retrograde Cholangiopancreatography (ERCP) result showed a big cyst in proximal common bile duct, with suspected intracystic stones (Fig. 1).

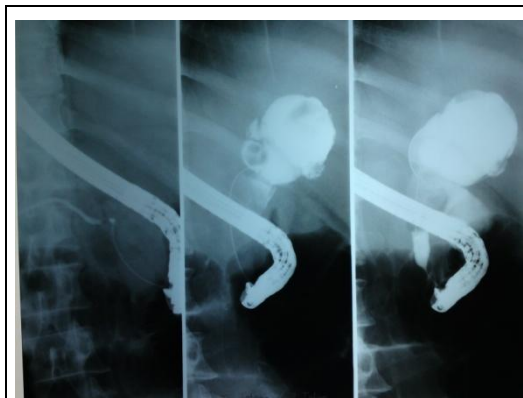


Figure 1. ERCP shows a diverticulum of the CBD, suggesting a type-II choledochal cyst.



Figure 2. Post stenting of the CBD

Discussion

Choledochal cyst is a pathological condition due to a specific weakness in a part or the whole of the wall of the common bile duct. It consists of congenital cystic dilatation of the extrahepatic biliary tree, intrahepatic biliary radicals or both.⁵

There are three classic symptoms in choledochal cyst: abdominal pain, icteric (jaundice), and a palpable mass. We only found abdominal pain and icteric in this patient, with no palpable mass. In adults, it is less likely to find those three classic symptoms of choledochal cyst than in children (25% vs. 82% respectively).⁶ In fact, this patient came with a history of jaundice, which was then examined whether it was caused by pre-hepatic, hepatic, or post-hepatic disorder so-called obstructive jaundice.

Obstructive jaundice may happen when there is an interruption of the bile flow in the biliary system. There are many causes of obstructive jaundice but the most common one is Choledocholithiasis, which mostly occurs in women.⁸

This patient was considered to have obstructive jaundice since he had a

greenish dark yellowish color, along with upper abdominal pain, pruritus, dark colored urine and pale stool⁷. His symptom did not indicate pre-hepatic or hepatic jaundice, which is usually preceded by flu-like syndrome with bright yellowish color.

The possibility of malignancy in this patient cannot yet be established since we could not make a CT scan due to financial consideration. Carcinoma of the head of the pancreas is the most likely cause of malignancy, which occurs more in men than in women, with 2:1 ratio. The possibility of malignancy in this patient is also supported by a drastic drop of body weight from 92 to 65 kilos during the last 9 months and by the fact that he was a heavy cigarette smoker, which is a risk factor.¹ Prolonged jaundice indicated a blockage that happened slowly in the common bile duct. These features most likely can be found in malignancy.

ERCP result in June 2009 led to a suspicion of a big choledochal cyst. We believe that the cyst in this patient was a type-II choledochal cyst. Alonso-Lej classified the choledochal cysts into three types, which were subsequently

modified by Todani et al. in 1977. This classification is currently the most common one used in daily clinic. A type I cyst is a fusiform dilatation of the common bile duct. An isolated diverticulum protruding from the wall is considered to be a type II choledochal cyst. A type III choledochal cyst is also known as a choledochoceles because it arises in the intraduodenal portion of the common bile duct. Multiple dilatations of the intrahepatic and extrahepatic bile ducts are considered type IVA, while type IVB involves exclusively the extrahepatic bile ducts. Type V is also known as Caroli's disease, which involves multiple intrahepatic bile duct dilatation.⁹ Usually, Magnetic Resonance Cholangiopancreatography (MRCP) approach is effective and is the first choice of imaging technique for examining these cysts.⁶ Regrettably, as previously mentioned, the patient could not afford such examination.

We also found congenital disorder of his fingers, suggesting that the cyst may as well have been a congenital condition. The "cyst" found on fluoroscopy from ERCP might be construed as the gallbladder itself, considering a strong indication of a congenital anatomical malformation.

The vast majority of patients with choledochal cyst have an anomalous junction of the common bile duct with the pancreatic duct. An anomalous pancreatic bile duct junction is characterized with the entering of pancreatic duct into the common bile duct about 1 cm or more proximal to where the common bile duct reaches the ampulla of Vater. The anomalous pancreatic bile duct junction makes pancreatic secretions and enzymes active, and this may result in inflammation and weakening of the bile duct wall. Severe damage may result in

complete denuding of the common bile duct mucosa. A cyst may also develop after trauma or from fibrosis and stenosis of the distal common bile duct or from recurrent cholangitis associated with stones in the bile duct; the early changes of this type are sometimes seen in the ERCP.⁵

Although we found multiple stones in the cyst, we believe that these stones are secondary and not the cause of choledochal cyst, since there were no previous attacks of jaundice or cholangitis. Furthermore, there was no history of previous trauma or surgery on common bile duct; thus, the most probable cause of the choledochal cyst was anomalous pancreatic bile duct junction.

USG did not reveal the cyst in the common bile duct. It was probably due to the cyst collapsing at the time of examination. It is not easy to find a cyst using USG if the cyst has collapsed.¹⁰ Nonetheless, ultrasound may help detect associated conditions and complications of choledochal cyst, such as choledocholithiasis, intrahepatic biliary dilatation, portal vein thrombosis, gall bladder or biliary neoplasms, pancreatitis and hepatic abscess.¹¹

Any type of choledochal cyst is susceptible to malignancy which is believed to be the result of chronic inflammation and metaplasia. The typical pathology is adenocarcinoma or occasional cases of small cell carcinoma. According to Atkinson et al, approximately 9 - 28 % choledochal cysts may develop to malignancy.³

All patients diagnosed as having choledochal cyst in adult life, regardless of the symptoms, should undergo complete excision of the extrahepatic duct, cholecystectomy, and Roux-en-Y hepaticojejunostomy. The outcome of this approach is expected to be

excellent.⁹ A type-V cyst (Caroli's disease) confined to one side of the liver can be treated by hepatic lobectomy. More diffuse varieties are complicated by recurrent cholangitis and stone formation, in which cases liver transplantation may be necessary.¹²

Conclusion

We have presented a type-II choledochal cyst in an adult, which is a very rare case. Choledochal cyst is mostly a congenital disease and it is rarely diagnosed in adult patients with obstructive jaundice. USG and ERCP may be used in determining the diagnosis even though MRCP is still the most excellent imaging technique. A complete excision of the extrahepatic duct, cholecystectomy, and Roux-en-Y hepaticojejunostomy is the treatment by choice, regardless of the symptoms; however, in this patient we could not perform such operation due to the patient's financial misfortune.

Bibliography

1. Oddsdottir M, Hunter JG. Gallbladder and extrahepatic biliary system. In: Brunicaudi, Andersen, Billiar, Dunn, Hunter, Pollock, ed. Schwartz's Manual of surgery. 8th edition. Part II. Chapter 31. New York: McGraw Hill, 2006; p.831-6.
2. Besner GE. Choledochal cyst: surgical perspective. 2008 [cited 2009 June 19]. Available from: <http://www.emedicine.com>.
3. Singla SL, Kadian YS, Godara R, Kajal P. Choledochal cyst in an adult - a case report. The Internet Journal of Surgery. 2009 Volume 19 Number 2 [cited 2009 June 19]. Available from: http://www.ispub.com/journal/the_internet_journal_of_surgery/volume_19_number_2_1/article/choledochal-cyst-in-an-adult-a-case-report.html.
4. Atkinson HDE, Fischer CP, de Jong CHC, Madhavan KK, Parks RW, Garden OJ. Choledochal cysts in adults and their complications. Journal of the International Hepato Pancreato Biliary Association. 5(2). 2003 [cited 2009 June 19]. Available from: <http://www.pubmedcentral.nih.gov/articlerender.fcgi?artid=2020569>.
5. Sbeh TA, Rehani H, Smadi S, Jarrah B. Adult choledochal cyst. JRMS. June 2006; 13(1):64-66. [cited 2009 June 19]. Available from: <http://www.jrms.gov.jo/Portals/1/Journal/2006/pdf%20June%202006/ADULT%20CHOLEDOCHAL%20CYST.%20A%20CASE%20REPORT.pdf>.
6. Sherlock, Sheila. Disease of the liver and biliary system. Eleventh edition. Italy, Milan: Blackwell Publishing, 2002.
7. Jaundience. [cited 2009 June 19]. Available from: <http://www.wikipedia.com>.
8. Jaundience. 2005 [cited 2009 June 19]. Available from: <http://www.surgical-tutor.org.uk/default-home.htm?system/abdomen/jaundice.htm~right>
9. Visser BC, Suh I, Way LW, Kang SM. Congenital choledochal cyst in adults. Archives of surgery. Vol 139 No.8 August 2004; 855-62. [cited 2009 June 19]. Available from: <http://archsurg.amaassn.org/cgi/content/full/139/8/855>.
10. Maheshwari M, Parekh BR, Lahoti BK. Biliary peritonitis: a rare presentation of perforated choledochal cyst. Indian Pediatrics. 2002; 39:588-592.
11. Liu CL, Fan ST, Chung M, Lo CM, et al. Choledochal cyst in adults. Archives of surgery. 2002; 137(4): 465-68.
12. Oldham KT, Colombani PM, Foglia RP, Skinner MA. Principles and Practice of Pediatric Surgery. 4th edition. Lippincott Williams & Wilkins. 2005; 1504.