Turk J Gastroenterol 2006; 17 (1): 70-73

An adult choledochocele case presented with gastric outlet obstruction: A rare presentation

Mide çıkış obstrüksiyonuna neden olan erişkin bir koledokosel olgusu: Nadir bir başvuru şekli

Mehmet Fatih CAN, Nihat KAYMAKÇIOĞLU, Gökhan YAĞCI, Semih GÖRGÜLÜ, Turgut TUFAN

Department of General Surgery, Gülhane Military Medical Academy, Ankara

Bile duct cyst is a biliary tract disease that is less common among adults compared to children, and it is accepted to have a congenital entity. The classical findings constitute a triad including abdominal pain, jaundice and abdominal mass. However, infective findings also occur in case of developed cholangitis, pancreatitis and cholecystitis. Duodenal obstruction leads to a rarely seen case in patients accompanied by nausea and vomiting, and it may easily be confused with other obstruction causes. We present a case report of an adult male patient with complaints of abdominal pain, and intermittent nausea and vomiting. Due to failure of attempted endoscopic approach for treatment, we performed transduodenal cyst excision during surgical procedure on the patient diagnosed to have type III biliary tract cyst. The complaints of the patient decreased significantly after the surgical therapy compared to the previous period and no complication was observed.

Key words: Bile duct cyst, choledochocele, gastric outlet obstruction

Safra yolu kisti erişkinde çocukluk dönemine göre daha az görülen ve doğumsal olduğu kabul edilen safra yolları hastalığıdır. Karın ağrısı, sarılık ve karında ele gelen kitle klasik klinik triadı oluşturur. Bununla birlikte kolanjit, pankreatit ve kolesistit gelişmesi durumunda enfeksiyona ait bulgular tabloya eşlik eder. Duodenal obstrüksiyon hastalıkta çok nadir gözlenen, bulantı ve kusmanın eşlik ettiği bir tabloya neden olur ve diğer obstrüksiyon nedenleri ile rahatlıkla karışabilir. Bu yazıda, karın ağrısı ile birlikte zaman zaman ortaya çıkan bulantı kusma şikayetleri bulunan erişkin bir erkek hasta sunulmuştur. Tip 3 safra yolu kisti (koledokosel) tanısı konulan hastaya öncelikli olarak uygulanan endoskopik girişim ile sonuca ulaşılamadığından hasta opere edilmiş ve transduodenal kist eksizyonu gerçekleştirilmiş, ameliyat sonrası hastanın şikayetleri gerilemiş ve komplikasyon gözlenmemiştir.

Anahtar kelimeler: Safra yolu kisti, koledokosel, mide çıkış obstrüksiyonu

INTRODUCTION

Bile duct cyst is a biliary tract disease which was first defined by Vater in 1723 (1). It is specifically prevalent in East Asian countries and is more common among females compared to males. It commonly arises as a congenital disease of the childhood period, when symptoms generally present (2). However, in recent years, there has been an increase in the literature regarding cases in whom symptoms were first observed as adolescents or adults. Although the classical triad composed of abdominal pain, jaundice and abdominal mass is more common among children compared to adults, in infants in particular, jaundice alone presents more frequently than the classical state (3). It is an exceptionally rare case that a symptom occurs due to obstruction of intestinal passage by a bile duct cyst. Moreover, most cases with accompanying duodenal obstruction can be attributed to a further pathology (i.e. annular pancreas, duodenal atresia). In this paper, we report an adult male choledochocele case who presented with symptoms other than the classical presentation due to the obstruction of duodenal transit by a type III cyst.

CASE REPORT

A 21-year-old male patient presented to the surgery department with abdominal pain, and intermittent nausea and vomiting complaints which

Address for correspondence: Mehmet Fatih CAN
Department of General Surgery, Gülhane Military Medical Academy, 06018 Etlik, Ankara, Turkey
Phone: +90 312 304 51 10 • Fax: +90 312 304 51 13
E-mail: mfcan@gata.edu.tr

Manuscript received: 24.05.2005 **Accepted:** 28.12.2005

had appeared periodically for about six months. These attacks generally occurred after meals and repeated a few times once they occurred. Furthermore, he had persistent blunt pain in the right hypochondrium that became more severe during these nausea and vomiting episodes. No positive finding except sensitivity with palpation on the right hypochondrium was observed in the physical examination. Reasonable hematological and biochemical results were received in the laboratory examination. Ultrasound (US), stomach-duodenum radiogram (Figure 1), computerized tomography (CT) and magnetic resonance cholangiopancreatography (MRCP) were performed subsequently and all displayed similar findings in regard to an intraduodenal cystic lesion in the second portion of the duodenum. Furthermore, MRCP showed that the cyst had the same intensity as the biliary tract and might have originated on the common bile duct (Figure 2). Endoscopic retrograde cholangiopancreatography (ERCP) failed due to bad location of ampulla of Vater. Based on these findings, exploration of the patient was decided and he was taken into operation. It was possible to enter the second part of the duodenum with anterior duodenotomy after the Kocher maneuver. A soft and pedicular mass, roughly 3x3x5 cm in size, was observed in the medial wall of the duodenum. The lesion, which included the ampulla of Vater, had a smooth surface covered with duodenal mucosa (Figure 3a). When a silastic tube was placed in the ampulla, bile was observed (Figure 3b). After total excision of the lesion, together with the duodenum mucosa covering it, the

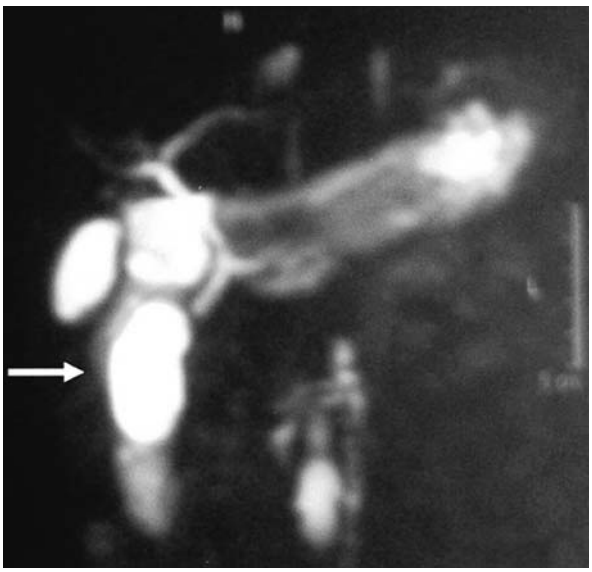

Figure 2. The MRCP appearance of the bile duct cyst

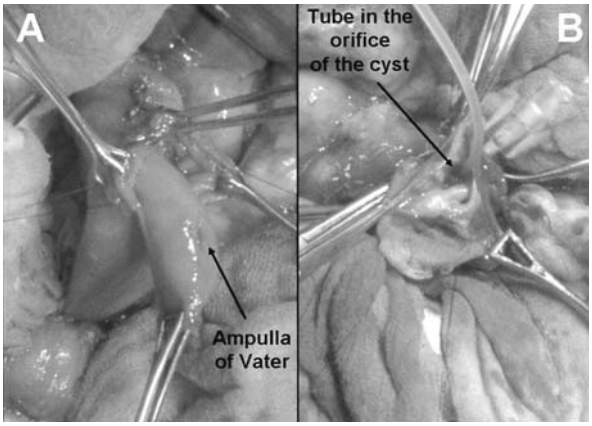

Figure 3: A) Choledochocele and ampulla of Vater after duodenotomy. **B)** Orifice of the cyst in pedicle after silastic tube placement

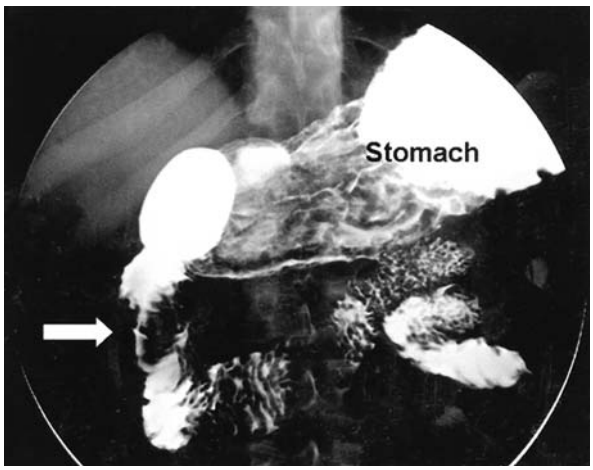

Figure 1. The appearance of the cyst as intraduodenal filling defect by stomach-duodenum radiograph

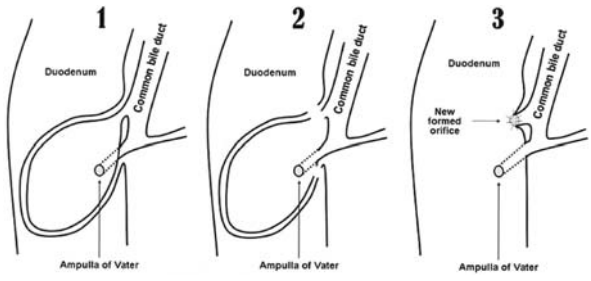

Figure 4. The schematic aspect of the operation: 1. Exploration; 2. Excision; 3. Anastomosis

intramural pedicle was anastomosed to duodenal mucosa (Figure 4). Sphincterotomy was not applied due to the decreased pressure in choledochus by new orifice on the duodenum. The duodenotomy incision was primarily closed. The patient experienced no complication in the postoperative period and was discharged from the hospital. Endoscopic follow-up in the second postoperative month revealed no problems with biliary and intestinal passage.

DISCUSSION

The knowledge related to bile duct cysts has significantly increased since it was first defined. Current knowledge is based on Todani and colleagues' modification of the 1977 classification of the extrahepatic disease, which was primarily made by Alonso-Lej in 1959, according to cyst locations (4). Bile duct cysts have been typed to five categories according to Todani classification. The most common type among adults is type I when general prevalence is considered. Furthermore, type IV is slightly more frequent in adults than in children. On the other hand, type III bile duct cyst (also known as choledochocele) is observed in only 4% of total cases (5). It is generally accepted that the cause of the disease is congenital disorder development of the bile duct. However, there is also an opinion advocating that biliary tree mural defects are formed due to deterioration caused by reflux of the pancreatic enzymes into the extrahepatic bile duct (6). In addition, it is still controversial whether the disease may occur later in adult patients. The frequency of the classical triad of the disease is approximately 40%. While jaundice is the major complaint among children, adults usually present with nonspecific complaints related to the right hypochondrium. However, cholangitis, pancreatitis and cholecystitis are more common among these patients compared to the normal population due to their predisposition. Furthermore, symptoms and findings of these infections, such as fever, weakness and tachycardia, are also observed if these infections are present in the case. Another case resulting in increased predisposition is the development of malignancy (7). In these cases, progressive jaundice, weight loss and findings of metastasis are also observed. Symptoms due to symptomatic gallstones or cholecystitis exist in nearly half of the adults (8). Some of these cases have cholecystectomy or another surgical operation before diagnosis.

There are few data concerning whether symptoms or findings other than the above-mentioned develop among adult patients with bile duct cysts. Duodenal obstruction cases are reported in regard to children due to additional pathologies such as annular pancreas or duodenal atresia (9). Duodenal obstruction is almost impossible for these lesions except for type III cysts. Theoretically, type III cysts may lead to findings of the intestinal system (such as obstruction, bleeding or perforation) as they protrude to the duodenal lumen. However, as stated above, choledochocele constitutes only a small fraction of total cases and there is little information in the literature regarding different symptoms and findings, as most of these cysts may be asymptomatic. Our patient had neither biliary obstruction findings such as jaundice and hyperbilirubinemia nor any sign or findings in favor of infection. Thus, the cause of the nausea and vomiting complaints, which usually appeared after meals, was thought to be existence of a duodenal obstruction. The patient complained of nausea a few times and once vomited bile after a meal during the hospitalization period. However, no significant finding was encountered in the laboratory examinations of the patient during this period. There are a few case reports in the literature, mostly in childhood, regarding choledochocele, also referred to as gastric outlet obstruction, with blockage of intestinal passage in the duodenum, and this is considered exceptionally rare among the adult population (6, 10-12). The reason why the complaints of the patient only periodically arose can be explained by spasm of Oddi's sphincter or postprandial increase in the cyst content.

Imaging methods such as prior US and CT are included in the regular algorithm like in other pancreatobiliary system diseases (13). MRCP and ERCP usage has also significantly increased in recent years. The importance of MRCP in diagnosis stems from the fact that it has a similar sensitivity as ERCP as well as a significantly low risk of complication (8). Evaluation of the suspicious complaints with MRCP and ERCP in addition to US and CT, together with the increase in diagnostic ability, may help to clarify related reasons why the disease has become more common in recent years. MRCP was quite important in the differential diagnosis of duplication cyst of the duodenum in our case. Nonetheless, it has been suggested that both diagnosis and treatment of duplication cyst of the duodenum can be achieved by ERCP (14). The

stomach-duodenum radiograph for type III cyst also contributed to the diagnosis by displaying the filling defect.

The treatment modalities for bile duct cystic diseases differ according to cyst type in Tadoni classification. In recent years, cysto-jejunostomy and cysto-duodenostomy methods have not been preferred due to the higher incidence of complication and continuity of the malignancy risk (15). Total cyst excision is acknowledged as the best therapy in type III bile duct cysts. There are also publications reporting that asymptomatic cases can be observed without treatment due to the lower malignancy potential compared to other types of cysts (16). After excision, anastomosis of the biliary and pancreatic channels independently to the duodenum may be necessary due to the location of the cyst. The regression of saccular structured type III cyst after endoscopic sphincterotomy is rarely seen in contrast to other cyst types of the bile duct (particularly the fusiform ones) (17). On the other hand, it has been reported in some of the publications that resolution of the cyst can occur after sphincterotomy through ease of biliary and pancreatic drainage (18, 19). Essentially, if the method can be accomplished in these symptomatic cases, ERCP seems a preferable application due to both its diagnostic and therapeutic effectiveness in one session. In this way, patients may be followed safely due to low malignancy risk after regression of the cysts. ERCP was also attempted in our case; however, cannulation of the ampullary orifice in the posterior wall of the cyst was not successful. Thus, transduodenal total cyst excision was applied to the patient and the orifice in the pedicle of the cyst was anastomosed to the duodenum at nearly 1.5 cm proximal to the ampulla of Vater. Enabling the endoscopic approach in later follow-up of the patient is also a significant advantage of this operation technique. Endoscopic evaluation was possible in our case in the second follow-up month and bile stream through natural and new orifices was observed. Although operations of biliary-enteric anastomosis like hepaticojejunostomy and choledochojejunostomy are frequently used in the therapy of other types of bile duct cysts, they are not regularly employed methods for the therapy of choledochocele.

In conclusion, type III bile duct cyst, also known as choledochocele, is a rarely seen form of an infrequent disease among adults. The patients may be asymptomatic or have nonspecific symptoms, as seen in most cases. We believe that it would be beneficial in the differential diagnosis to take into consideration the rarely seen symptoms and findings related to duodenal obstruction by the cyst together with the other supporting data.

REFERENCES

1. Chowbey PK, Katrak MP, Sharma A, et al. Complete laparoscopic management of choledochal cyst: report of two cases. J Laparoendosc Adv Surg Tech A 2002; 12: 217-21.
2. Lee KF, Lai ECH, Lai PBS. Adult choledochal cyst. Asian J Surg 2005; 28: 29-33.
3. Altman RP, Lazar ER. Neonatal biliary atresia, hypoplasia, and choledochal cyst. In: Zuidema GD, Yeo CJ, eds. Shackelford's Surgery of the Alimentary Tract. 5th ed. Philadelphia: WB Saunders, 2002; 280-9.
4. Todani T, Watanabe Y, Toki A, Morotomi Y. Classification of congenital biliary cystic disease: special reference to type IC and IVA cysts with primary ductal stricture. J Hepatobiliary Pancreat Surg 2003; 10: 340–4.
5. Nagorney DM. Bile duct cysts in adults. In: Blumgart LH, Fong Y, eds. Surgery of the Liver and Biliary Tract. 3rd ed. London: WB Saunders, 2000; 1229-44.
6. Metcalfe MS, Holden SAW, Maddern GJ. Management dilemmas with choledochal cysts. Arch Surg 2003; 138: 333-9.
7. Wu GS, Zou SQ, Luo XW, et al. Proliferative activity of bile from congenital choledochal cyst patients. World J Gastroenterol 2003; 9: 184-7.
8. Söreide K, Korner H, Havnen J, Soreide JA. Bile duct cysts in adults. Br J Surg 2004; 91: 1538-48.
9. Komuro H, Makino S, Tahara K. Choledochal cyst associated with duodenal obstruction. J Pediatr Surg 2000; 35: 1259-62.
10. Hamada Y, Tanano A, Sato M, et al. Rapid enlargement of a choledochal cyst: antenatal diagnosis and delayed primary excision. Pediatr Surg Int 1998; 13: 419-21.
11. Adamek HE, Schilling D, Weitz M, Riemann JF. Choledochocele imaged with magnetic resonance cholangiography. Am J Gastroenterol 2000; 95: 1082-3.
12. Bona S, Boiselle JC, Harb J, et al. Intra-duodenal development of low choledochal diverticula. Apropos of 2 cases. Ann Radiol (Paris) 1991; 34: 273-7.
13. Zhong L, Yao QY, Li L, Xu JR. Imaging diagnosis of pancreatobiliary diseases: a control study. World J Gastroenterol 2003; 9: 2824-7.
14. Sezgin O, Altiparmak E, Yilmaz U, et al. Endoscopic management of a duodenal duplication cyst associated with biliary obstruction in an adult. J Clin Gastroenterol 2001; 32: 353-5.
15. Shi LB, Peng SY, Meng XK, et al. Diagnosis and treatment of congenital choledochal cyst: 20 years' experience in China. World J Gastroenterol 2001; 7: 732-4.
16. Jordan PH Jr, Gross JA Jr, Rosenberg WR, Woods KL. Some considerations for management of choledochal cysts. Am J Surg 2004; 187: 790-5.
17. Ng WT, Liu K, Chan KL. Endoscopic treatment of choledochocele. Surg Endosc 1998; 12: 469.
18. Zheng LX, Jia HB, Wu DQ, et al. Experience of congenital choledochal cyst in adults: treatment, surgical procedures and clinical outcome in the second affiliated hospital of Harbin Medical University. J Korean Med Sci 2004; 19: 842-7.
19. Akkiz H, Colakoglu SO, Ergun Y, et al. Endoscopic retrograde cholangiopancreatography in the diagnosis and management of choledochal cysts. HPB Surg 1997; 10: 211-8.