Case Report

Todani IA Choledochal cyst- Presentation of the disease with a case report

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Abstract

Choledochal cyst is a rare disease of the biliary tract. There are five main types of choledochal cysts with a few recognized sub-types. The etiology of choledochal cysts still is unclear. The incidence of biliary tract cancer in patients with choledochal cysts increases with age. In the past, choledochal cysts were often treated using drainage procedures; however, the optimal treatment used today is likely to involve the complete excision of the extrahepatic duct, cholecystectomy, and Roux-en-Y hepaticojejunostomy. In 1995 Farello et al. first reported laparoscopic choledochal cyst excision. We report a case of Todani type IA choledochal cyst with choledocholithiasis. Delay in the diagnosis increases the frequency of associated biliary pathology, malignant alternation and suboptimal surgical therapy. Often, intraoperative finding of choledochal cyst is the first contact with this rear entity, so awareness of possible presence of this uncommon disease is very important for surgeon.

Key words : common bile duct, choledochal cyst, cholangiography

Introduction

Choledochal cyst is a rare entity, a congenital dilatation at any portion of the biliary tree that appears more often in the main part of the common bile duct. Statistics shows one case per 100,000 to 150,000 live births. 75% are diagnosed in childhood and 20% of case in the adult. It is more common in women than men (3-4:1)[1]. Choledochal cysts were described for the first time by Vater in 1723. There are several theories that attempt to explain the etiology of choledochal cyst. It is believed that its origin may be related to abnormal coledoco-pancreatic-ductal union allowing chronic reflux of pancreatic enzymes into the bile duct, which re-consultation in weakening and dilation of the road, and the subsequent formation a quiste[2,3]. The widely accepted classification system for choledochal cysts, devised by Todani and collaborators, is based on the cholangiographic morphology, location and number of intrahepatic and extrahepatic biliary tree cysts (Fig-1). Type-I is the most common (80–90%). The treatment depends on the type of the cyst classified according to the Todani's classification, and has the excision of the cyst as a general principle. There are many options for the biliary tract reconstruction, and some studies have been conducted to compare them. The laparoscopic surgery is a feasible approach (such as the robotic surgery), with the advantage of being a minimally invasive procedure.

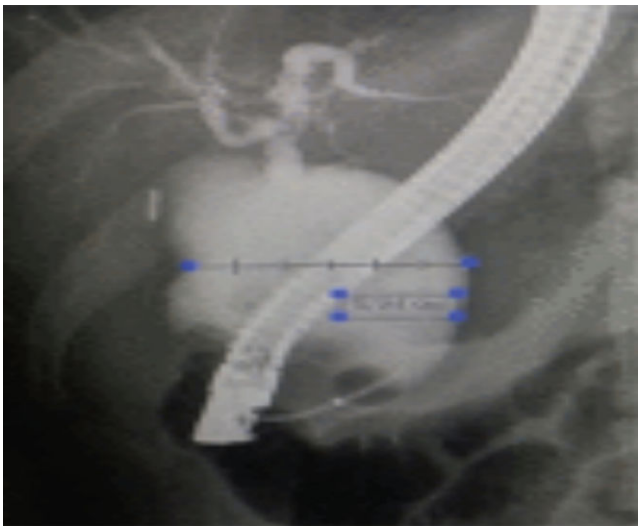

Fig-1 : Endoscopic retrograde cholangio pancreatography that shows the choledochal cyst of 5.94 cm

Case report

A 16-year-old female patient, who was a student without any associated disease, presented with epigastric pain since she was 6 years old and with post-alimentary vomits after fatty food ingestion in the last year. There was no history of jaundice. Investigation with abdominal

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ultrasound and Magnetic resonance Cholangio-pancreatography (MRCP) revealed a severe cystic dilation of 5 cm in diameter in the choledochal duct with choledocholithiasis classified as Type IA of Todani's [Figure-2]. On admission, the serum bilirubin level was 0.2 mg/dl, alkaline phosphatase was 239 U/l, and the activities of gamma glutamyltranspeptidase, aspartate transaminase and alanine transaminase were 40 U/l, 56 U/l, and 68 U/l, respectively. Surgical treatment was indicated and open cholecystectomy plus excision of the biliary cyst with Rouxen-Y hepaticojejunostomy reconstruction was performed. The surgical specimen is shown in Figure 4 and the histopathologic analysis did not reveal any signs of malignancy. The postoperative curse was uneventful.

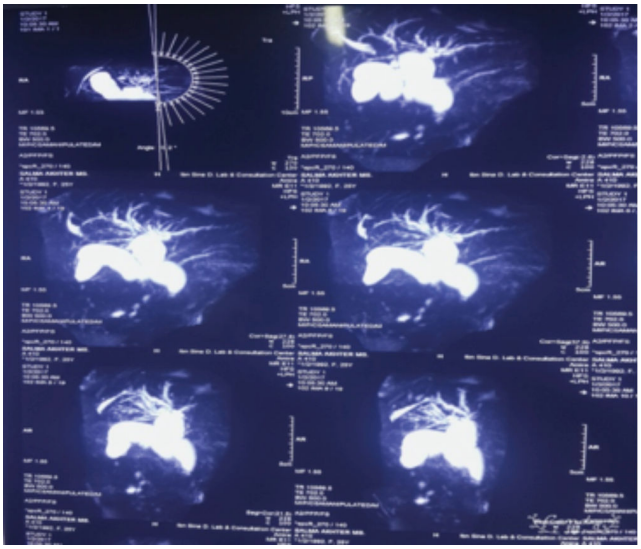

Fig-2 : MRCP showing Todani Type-1 A choledochal cyst

Discussion

Choledochal cyst is a disease almost exclusively affecting the pediatric population, but recent studies are showing increased diagnosis in adults in the West[1]. As reported in this case, Type I of Todani's classification is the most common presentation in 50%-80% of cases[3-5]. The classic triad of symptoms consisting of abdominal pain, jaundice, and a palpable abdominal mass occurs in less than 20% of the patients[6]. Mukhopadhyay et al reported abdominal pain as the most common presentation in a pediatric population[4]. It is possible to conclude that adults may have a different presentation from children and there are no specific symptoms for the choledochal cyst diagnosis. Thus, the suspicion of this pathology requires a valorization of these common symptoms and a special attention given to the imaging date and incidental findings. Association with other hepatobiliary pathologies occurs in 80% of the cases and includes benign diseases such as lithiasis in the cyst, intrahepaticlithiasis, acute cholecystitis and pancreatitis, and also malignant neoplasms, with cholangiocarcinoma being the most common. Patients with choledochal cysts have 20 times greater risk for the development of cholangiocarcinoma than the general population. Other malignant neoplasms have been reported, such as neuroendocrine tumors, but are much less common[7]. Technically, cyst excision is the main step to diminish the risk for cancer development, and must be performed when it is possible. Unfortunately, a cholangiocarcinoma may arise even after the cyst excision, as described by Nishiyamaet al[8]. The surgical strategy depends on the type of the cyst. For the Type I cyst, complete excision of the cyst is usually feasible and Roux-en-Y hepaticojejunostomy is the preferable reconstruction technique. This approach was associated with low morbidity and mortality rates and few long-term complications[9]. Reconstruction with a hepaticoduodenostomy has also been studied and found to have good outcomes. Mukhopadhyay et al., reported a review of 79 cases with hepaticoduodenostomy reconstruction and concluded that it is a quick procedure, with preservation of normal anatomy and physiology, and avoids multiple intestinal anastomoses. They suggested that this should be the preferred approach, as there were minimum complications[4]. Santore et al., compared hepaticoduodenostomy versus hepaticojejunostomy reconstructions in their series, stating that hepaticoduodenostomy required less operative time, allowed faster recovery of bowel function, and produced fewer complications requiring reoperation[10]. With this data, it is reasonable to conclude that both the reconstruction techniques can be accepted. Further studies should be conducted to better understand the advantages and disadvantages of such techniques.

The laparoscopic approach for the choledochal cyst excision is the technique of choice instead of open laparotomy[11]. It is a safe procedure and has the advantages of a minimally invasive surgery, such as lower pain, less wound complications, lower hospital stay, and faster surgical recovery. Robotic approach has also been described and seems to be as feasible as laparoscopy, although cost issues could be raised in this case[12]. It has to be emphasized that despite the advantages and feasibility of the laparoscopic approach for the treatment

of choledochal cysts, it must be reserved for experienced surgeons in complex biliary procedures and advanced laparoscopic surgery, in order to avoid lesions and complications on the biliary tract with a negative impact in the patient outcome.

Conclusion

There are some peculiarities in the clinical presentation of this disease in adults compared to the pediatric population. The aim of treatment is directed at the cyst excision, with reconstruction of biliary tract through hepaticoduodenostomy or hepaticojejunostomy. The laparoscopic approach is the preferable technique and must be performed only by experienced surgeons.

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