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CASE REPORT / ПРИКАЗ БОЛЕСНИКА

Large choledochal cyst initially interpreted as Mirizzi syndrome – case report and literature review

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SUMMARY

Introduction Choledochal cysts are congenital anomalies manifested as focal or diffuse cystic dilatation of the bile ducts. They are mostly diagnosed in childhood. The magnetic resonance and surgical management are the gold standard diagnostic and treatment modality.

Case outline We present a male patient who was presumed to have Mirizzi syndrome. This presumption was discarded by additional imaging procedures and by later surgical treatment. It was confirmed that it was a large choledochal cyst filled with stones. Considering the diagnosis and good patient's general condition, we opted for surgical treatment.

Conclusion There are several surgical techniques that can be used in the treatment of choledochal cysts, wherein each is intended as a complete resection of the cyst with histological confirmation. Operative techniques do not affect the outcome of the treatment, but the time and extent of surgical resection, as well as any metaplasia of the epithelium, do.

Keywords: choledochal cyst; Todani; common hepatic duct; icterus; Mirizzi syndrome

INTRODUCTION

Choledochal cysts are congenital anomalies that manifest as focal or diffuse cystic dilation of the biliary tree. It is a rare clinical entity with an incidence of 1/150.000 births approximately [1]. There is a significant female predominance with a female to male ratio of 4:1 [2]. Five types of cysts have been described, of which a choledochal cyst type I has around 68% of percentage coverage out of all subtypes [1, 2].

The majority of patients are usually diagnosed in childhood, in the first decade of life. Only 20% of cases are diagnosed in adults [3]. Symptomatology is different in children and in adults. In children, a classical triad of symptoms in the form of abdominal pain, a palpable mass in the upper right quadrant of the abdomen and obstructive jaundice frequently occur. In adults, the ailments are related to biliary and pancreatic symptoms, accompanied by abdominal pain. The ruptures of the cysts are rare and such an occurrence is reported only in neonates [4].

In a differential diagnosis, biliary lithiasis, sclerosing cholangitis, and pancreatic pseudocyst may be taken into account. Biliary atresia is often associated with choledochal cysts and therefore should be excluded at neonatal obstructive jaundice [2, 4].

Choledochal cyst Type I treatment involves complete surgical resection with reconstruction with an isolated small bowel segment by the method of Roux-en-Y. Sometimes, the scope of surgical resection may be much more radical. When the cyst is spreading into the head of the pancreas a cephalic duodenopancreatectomy is preferred surgical option [5].

The objective of this paper is to present the rare disease, the surgical technique, and the literature review.

CASE REPORT

In our paper, we present a male patient, 53 years of age, who was admitted to our hospital because of pain in the upper abdomen, subicterical and afebrile. The initial abdominal ultrasound was performed. Distended gallbladder, with a thickened wall and with multiple stones in the lumen were seen. Choledochus was with concrements inside the lumen, and in close relationship with the gallbladder, Mirizzi syndrome differential diagnosis. A magnetic resonance imaging (MRI) and magnetic resonance imaging cholangiopancreatography (MRCP) were performed as a part of additional diagnostics, in which a large fusiform ductus choledochus cyst was seen. A large choledochal cyst (CC), 7 cm in longitudinal diameter and 4 cm in transverse diameter, was localized at about 2 cm below the primary biliary confluence, without communication with the lumen of the gallbladder, in close contact with the portal vein and with proper hepatic artery. The cyst was filled with numerous stones of different sizes (Figure 1). The laboratory work-up showed elevated WBC 18 × 10^9/L, CRP 65 mg/L, total bilirubin 49 mg/L, and alkaline phosphatase 197 IU/L.

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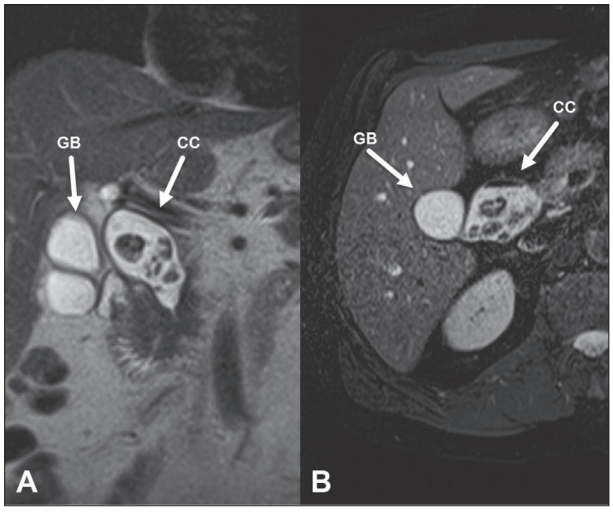

Figure 1. Abdominal magnetic resonance imaging – gallbladder (GB) and Type I choledochal cyst (CC) in sagittal (A) and axial (B) plane

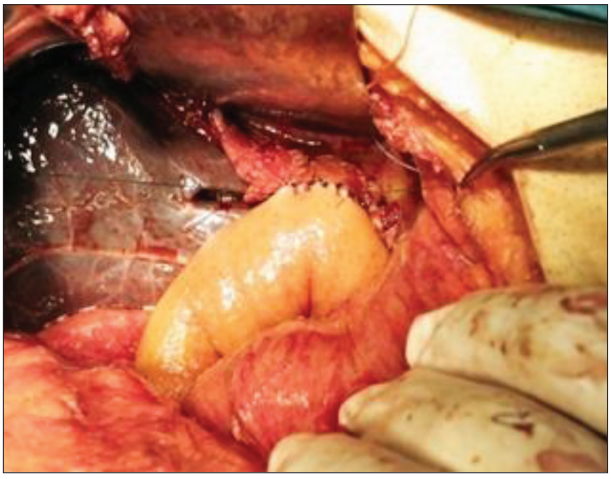

Figure 2. Intraoperative photograph showing hepaticojejunostomy performed using interrupted suture at about 1 cm below the primary biliary confluence

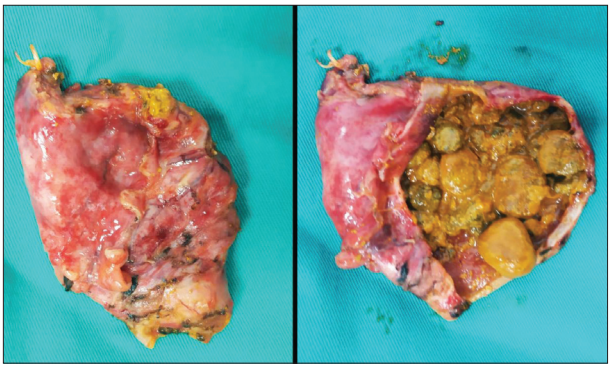

Figure 3. Macroscopic image of the specimen, before and after opening, showing multiple stones in its lumen

In his medical history, the patient provided information that gallbladder calculus was verified by abdominal ultrasound more than 10 years previously. In addition to the examinations that were performed several times during this period, no additional diagnostic procedures were performed. The patient stated that he repeatedly felt difficulties in the form of biliary colic and sometimes spontaneously resolved mild jaundice, which were treated in a conservative manner.

The patient was in good general condition, so we opted for surgical treatment. Given the size, the position of the cyst, and its proximity to surrounding structures, primarily vascular, we applied the open surgical approach. In terms of general endotracheal anesthesia, the abdomen was opened with the right subcostal laparotomy. After adhesiolysis and the inspection, the preoperative diagnosis was confirmed with an inflammatory block surrounding the hepatoduodenal ligament. Hard adhesions were obscuring normal anatomy, cholecystectomy and a careful dissection of the hepatoduodenal ligament was performed, with difficult separation of the vascular structures from the cystic structures. Resection of the common bile duct along with the large cyst was performed. The upper and lower resection was at a distance of 1 cm with respect to both ends of the cyst. Resection margins were sent to an *ex tempore* histopathologic examination. In the meantime, we performed an extensive lavage of the bile ducts, proximal part first and then the distal part from previously resected CC. Using choledochoscope, the proximal part relative to the branch of the left and the right hepatic duct, and then the distal stump of the resected hepatic ductus to the papilla of Vater were inspected. The finding was normal, with no residual stones. *Ex tempore* findings were negative for malignancy. Hepaticojejunostomy was performed using interrupted, monofilament, slowly absorbable suture (4/0) at about 1 cm below the biliary confluence (Figure 2). The abdomen was drained with two abdominal drains and the operating incision was reconstructed. The tissue of the cyst (Figure 3) and of the gall bladder was sent to histopathological examination.

Histopathological examination revealed only mild inflammatory changes in the wall of the gallbladder, with no dysplasia or other significant epithelial changes. A specimen with a cyst measuring 75 × 48 mm in diameter was extensively reviewed. It was seen that the areas of papillated and partly atypical epithelial proliferation were present in the bile duct, but coupled with poorly expressed atypia, so there were only focuses of light epithelial dysplasia (grade I–II). In the surroundings, chronic inflammation with some of the sporadic multiplicity of the diverticula of the same epithelium was present, as well as the light multiplication of such tubulo-glandular structures. None of the sections showed any invasiveness elements. Cystic part of the sections showed mostly flattened and only reactively altered, but mostly non-dysplastic epithelial changes.

The postoperative course was uneventful. Abdominal drains were removed on the third postoperative day and the patient was discharged on the seventh postoperative day. A month later, an abdominal ultrasound was performed and the finding was normal, as well as laboratory analysis. Six months after surgery, MRI and MRCP were performed showing that the anastomosis is passable and that the other findings in the abdomen were normal. The patient's condition is still monitored.

DISCUSSION

CCs were first reported by Vater and Ezler [6]. This congenital malformation is characterized by dilatation of the biliary tree without acute obstruction of the flow of bile. The cyst may be present in any part of the biliary tree. According to the volume, it may be complete or segmented, and according to the shape, it can be saccular or fusiform [7]. In our case, it was a fusiform cyst, filled with numerous concrements.

Etiology of CCs is still unclear and there is still no clear expert consensus. The most frequently mentioned hypothesis in the current literature suggests that CCs occur after partial obstruction of the bile duct, which produces an increase in pressure in the proximal part of the bile duct, which leads to the dilatation of this part [7]. CC prevalence is much higher in Asian countries such as in Japan and predominantly occurs in females [8].

The first classification of CC was announced by Alonso-Lej et al. [9] in 1959. This initial classification was completed by Todani et al. [10], wherein the choledochal cysts were classified into five types. Type I cysts represent the dilatation of the extrahepatic bile ducts. Also, they represent the most common type according to the previously mentioned Todani classification, where the incidence is 1:1,000, compared to western countries, where the incidence ranges from 1:150,000 up to 1:1,000,000 births. Type I CCs have the greatest frequency of occurrence (75–85%) compared to other types [1, 10]. Isolated cystic dilatation of the cystic duct was added to the Todani classification in 1991 as type VI [11].

CCs are presented with different symptoms, but they can often be asymptomatic. In symptomatic patients, they are commonly presented as abdominal pain, nausea, and vomiting. In these patients, biliary stones, cholangitis, liver abscess, and biliary cirrhosis are present in 60–80% of the cases [4, 12].

Ultrasound examination of the abdomen is the first diagnostic procedure, particularly in children. As an additional diagnosis, computed tomography is used. Both methods cannot always provide sufficient information. The gold standard is MRI, as well as MRCP, with an efficiency of 96–100% [1, 12]. Endoscopic retrograde cholangiopancreatography and percutaneous transhepatic cholangiography represent very reliable diagnostic procedures, but both are invasive procedures and their application is not routinely performed [12, 13].

In our case, only after completed MRI and MRCP were we able to remove the dilemma on the possible presence of Mirizzi syndrome. There was no communication of CC dilation with the lumen of the gall bladder, nor any expressed compression in the relations of biliary structures, as originally seen in the abdominal ultrasound examination. Since the only reliable and correct treatment modality of these biliary tree anomalies is complete surgical resection, surgical treatment should not be delayed, especially if you take into account that these anomalies represent the premalignant condition. Malignant alteration depends on the cyst type and that percentage is the largest for Type I cysts (about 70%), followed by Type IV cysts (about 20%), Type II cysts (about 5%), and Type III cysts (about 5%) [14].

Within frequent cholangitis and inflammatory processes of the surrounding structures and their consequences, sometimes it is difficult to achieve complete excision of the CC, especially because of the close relationship with the blood vessels, especially the portal vein. CC sometimes spreads into the parenchyma of the pancreas and it is necessary, in order to apply the adequate and radical surgical approach, to perform a cephalic duodenopancreatectomy with all the risks that this procedure can cause [5, 7].

After the excision of the cyst, the reconstruction can be done in two ways: hepatico-duodenal anastomosis or hepatico-jejunal anastomosis by the Roux-en-Y method [15]. The success of operational procedures and of the selection of anastomosis is measured by the ease of implementation, as well as by short- and long-term results of the surgical treatment. The data from the current literature suggest that the success of hepaticojejunostomy is about 92% with the complication rate of 7%, compared to hepatico-duodenostomy, with the complication rate about 42% [7, 15].

Surgical management can be carried out by using several surgical techniques. These include the classical operational approach, which we also apply, then minimally invasive or laparoscopic access, and the most modern, robotic-assisted surgical approach [16, 17].

Depending on the technical equipment and the training of the surgical team, in institutions where this type of surgery is performed, it is possible to effectively apply several surgical techniques, whose ultimate objective is the same – the complete excision of the CC with the appropriate reconstruction of the biliary ducts. After reviewing the literature and bibliographic databases (PubMed, Scopus), we came to the conclusion that the short- and long-term results of surgical treatment outcomes of patients operated on for Type I CC are similar, regardless of the applied surgical technique. It can be concluded that the applied surgical technique does not affect considerably the final result of the treatment, but the period required to diagnose the disease, the extent of surgical resection, and the presence of the bile duct epithelium metaplasia do.

Informed consent: Written informed consent was obtained from the patient for this case report publication, including the accompanying images, case history, and data.

Conflict of interest: None declared.

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Циста холедоха иницијално дијагностикована као Миризијев синдром – приказ болесника и преглед литературе

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САЖЕТАК

Увод Цисте холедоха су урођене аномалије које се манифестују као фокалне или дифузне дилатације жучних водова. Најчешће се симптоматски испоље и дијагностикују током детињства. Магнетна резонанца је златни стандард за постављање дијагнозе, а хируршко лечење је једини сигуран и ефикасан начин лечења.

Приказ болесника Болесник стар 53 године примљен је на клинику због болова у трбуху, мучнине и иктеруса. Из медицинске документације се сазнало да је 10 година уназад знао за калкулозу жучне кесе, те се после иницијалне ултразвучне дијагностике посумњало на Миризијев синдром. После спроведене допунске сликовне дијагностике утврђено је да се заправо радило о великој цисти холедохуса која је испуњена масом калкулуса. Имајући у виду добро опште стање болесника, одлучено је да се болесник лечи хируршки.

Закључак Описано је неколико оперативних техника за хируршки третман цисте холедоха. Без обзира на примењену технику, циљ је био да се одстрани комплетна циста и хистопатолошки прегледају маргине ресекције, као и сам препарат. Избор оперативне технике не утиче на исход лечења. С обзиром на познату тенденцију метапластичне промене епитела холедоха код болесника са цистом холедоха, од суштинског је значаја да се оперативном лечењу приступи на време.

Кључне речи: циста холедоха; Тодани; заједнички хепатични вод; иктерус; Миризијев синдром