Whipple Procedure in the Treatment of Choledochal Cyst

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Abstract

Introduction: Choledochal cysts are malformations of the intra- and extrahepatic biliary tree, congenital or acquired, occurring at low frequency in adulthood. Case report: We report the case of an adult patient (36 years) diagnosed with choledochal cyst during investigation for persistent abdominal pain, with choledocholithiasis associated with the clinical case. The reported case has some divergent characteristics of the reported cases of choledochal cyst, including that the patient did not present the classic triad of symptoms (abdominal pain, abdominal mass and jaundice) that accompanies the choledochal cyst and the surgery performed to treat the lesion was of great complexity (Whipple procedure).

Conclusion: The case shows the importance of always considering a diagnostic hypothesis of choledochal cyst in patients with persistent pain in right hypochondrium concomitant with laboratory changes typical of cholestasis. In addition, it adds to the current literature evidence indicating that total resection of the choledochal cyst should be emphasized and partial excision abandoned.

Keywords: Choledochal cyst, Obstructive jaundice, Biliary disease, Whipple procedure

Introduction

Choledochal cysts are malformations of the intra- and extrahepatic biliary tree, congenital or acquired, of little known pathogenesis [1]. Different theories or mechanisms have been proposed to explain the development of these cysts. Congenital cysts have been attributed to dilatation of the common bile duct, secondary to choledochal stenosis. Those acquired in 70% of the cases are associated with anomalies in the pancreaticobiliary junction [1,2]. There are five subtypes of choledochal cysts (I to V), [2] of which subtype I represents 67.9% of all cases. It is estimated that the incidence of choledochal cysts varies from 1: 100,000 to 1: 150,000, being more frequent in populations of Asian countries [3]. Women are more affected by the disease, with an estimated occurrence of 3: 1 to 4: 1 [2, 4, 5]. Choledochal cysts may be asymptomatic or symptomatic, leading to biliary cirrhosis. They can cause chronic or intermittent abdominal pain, which is more common among children and adolescents. Complications such as recurrent jaundice, cholangitis, gallstones, pancreatitis, common bile duct perforation, and malignant transformation of the cyst may occur [3]. Both diagnosis and classification of choledochal cysts subtypes can be made by abdominal ultrasonography, computed tomography, magnetic resonance cholangiopancreatography (MRCP), endoscopic retrograde cholangiopancreatography (ERCP), and endoscopic ultrasonography.

With increasing understanding of the clinical evolution and etiology of choledochal cyst, its treatment changed from “unobstructed biliary drainage” to “complete excision of the cyst” in order to eliminate risks of carcinogenesis by “performing of biliopancreatic derivations” [6]. - The choledochal cyst should be excised as much as possible during surgery, particularly its intrapancreatic portion [7].

Case Report

E.P.B., 36 years old, female, natural and from Rio de Janeiro [RJ], was attended at the emergency room of the Carlos Chagas State Hospital and referred to the General Surgery Service with complaints of epigastric abdominal pain and colic type hypochondrium for 15 days. The patient presented with coluria, jaundice, nausea, episodes of vomiting and weight loss. He denied fever and other constitutional symptoms. She reported medical history without known comorbidities, prior surgeries, or regular use of medications. On physical examination, the patient presented a regular general condition, with humid...
mucous membranes, but with icteric, blood pressure of 110/70 mmHg, heart rate of 81 bpm and axillary temperature of 36 °C. Inspection of head, neck, thorax and abdomen did not present alterations. No respiratory or cardiovascular changes were observed either. Abdominal examination revealed presence of air-fluid noises and flat, slightly depressed abdomen, with voluntary pain relief on the right palpation of the right hypochondrium, without hepatomegaly or splenomegaly.

Laboratory tests were requested and revealed a picture compatible with cholestatic jaundice: an innocuous hemogram with no left deviation, with total bilirubin of 4.2 mg / dL at the cost of the direct fraction (3.6 mg / dL), elevation of transaminases (ALT = 180 mg / dL and AST = 90 mg / dL), platelets of 160,000 / mcl, prothrombin time of 12 seconds (activity 100% / RNI = 1.0) and elevation of alkaline phosphatase (268 mg / dL). Abdominal ultrasonography was performed, showing choledochal dilatation measuring 1.4 cm, gallbladder with gallstone and calculus in the fundus. It was requested, then, the ERCP, which showed distal choledochal stenosis with important dilatation and calculation above the stenosis (1.5 cm). Biliary tract drainage, papillotomy, and choledochal prosthesis were performed.

Six months after the care, the patient returned to the Service to remove the choledochal prosthesis and a cholecystectomy. At this time, she presented clinical symptoms of abdominal pain in the right hypochondrium, without jaundice or fever, with significant weight loss due to hyporexia and episodes of continuous vomiting. An ERCP (Figure 1) was performed, which revealed dilated hepatocoles with a rounded distal sacral formation of approximately 2 cm near the papilla and biliopancreatic anomaly. Therefore, the patient was referred for MRCP examination (Figure 2), which showed mild dilatation of the intrahepatic bile ducts and common bile duct (0.8 cm), faults in the distal choledochal filling, preserved pancreatic (Wirsung) duct and pancreas without obvious changes.

In view of the high risk for bile duct malignancies, after preoperative examinations, the patient was submitted to a procedure for complete resection of the patient’s extrahepatic biliary tract with Whipple surgery (duodenopancreatectomy with pancreatic gastrostomy, reconstruction with anastomosis gastroenterostomose lateral to billroth II, and cholecystectomy) (Figure 3). The postoperative evolution was satisfactory. The patient presented a small peri-pancreatic collection, which was resolved with antibiotic therapy, without intercurrences, and was discharged on the 30th postoperative day. The surgical specimen was submitted to histopathological analysis, which revealed acute and chronic inflammatory processes, fibrosis, elastonecrosis, vascular ectasia and foci of hemorrhage in the pancreatic parenchyma and choledochal cyst wall.

Discussion

Choledochal cyst with the classic triad of abdominal pain, abdominal mass and jaundice, as reported in the present study, is seen in only 20% of the diagnosed cases [3, 8]. In addition, it rarely affects adult individuals, predominantly in the pediatric range³. In pediatric patients, such cysts are characterized by wall fibrosis (columnar epithelium) and leukocyte infiltration [9]. In adults, inflammation and hyperplasia are observed. Most of the common bile duct cysts present some degree of pathological alteration in the liver, such as portal fibrosis, central venous distention, inflammation of the parenchyma and proliferation of the bile ducts[10]. With the exception of portal fibrosis and central venous distention, other hepatic changes cool down after appropriate surgical management [10]. Other findings common to all subtypes of choledochal cysts are: acute and chronic

![Figure 1: ERCP image showing a biliary cyst type Ib, dilated hepatocoeococoe with a rounded distal sacral formation of approximately 2 cm along the papilla and anomaly at the biliopancreatic junction.](image1)

![Figure 2: MRCP image showing dilatation of bile ducts and faults in the distal bile duct filling.](image2)

![Figure 3: Distal insulation of the lesion (choledochal cyst type I-b).](image3)
mucosal inflammation, mucosal dysplasia, and little or no mucus-producing gland.

The subtypes of choledochal cysts differ histopathologically from each other. The cysts of subtype I (and sometimes type IV) lack biliary mucosa, those of subtype II resemble biliary vesicle duplication, those of subtype III are lined by duodenal mucosa, and those of subtype V may have extensive liver fibrosis [11]. Immunohistochemical findings suggest that there is an association between the age of the patient and the risk of epithelial metaplasia and biliary intraepithelial neoplasia in the cyst walls [12, 13]. Half of the patients with choledochal cyst and age over 50 years present invasive biliary neoplasia [13, 14].

Cysts of subtypes I and IV are associated with a high risk of malignancy, while those of subtypes II, III and V have minimal neoplastic risk [15]. Evidence indicates that carcinogenesis involves several genetic events, with mutations in the K-ras gene, with increased P53 expression, observed in more than 60% of choledochal cyst-related carcinomas, followed by late inactivation of the DPC-4 gene [16].

The majority of reported cases of malignant transformation correspond to cholangiocarcinomas. However, gallbladder carcinomas have been identified in 10% to 25% of malignancies [5]. It is believed that the anomalous junction of the pancreatic duct with the bile duct plays a role in carcinogenesis and hepatocellular damage, due to reflux of pancreatic contents into the bile duct. In addition, in patients with choledochal cyst, biliary amylase elevation is associated with an increase in the expression of the inducible nitric oxide isomerase (iNOS), suggesting that this is also a mechanism that elicits mucosal hyperplasia of the choledochal cyst and carcinogenesis [17].

In the present case report, the patient presented with pain in the upper right quadrant, jaundice, vomiting, cholestatic alterations and findings compatible with calculi biliary disease (choledocholithiasis). Initially, based on this diagnosis, she received treatment that did not result in resolution of angina and episodes of vomiting. This fact shows the importance of developing a diagnostic hypothesis of choledochal cyst in patients with persistent pain in the right hypochondrium concomitant with laboratory abnormalities of cholestasis.

Regarding the diagnosis, ultrasound of the liver and bile ducts is the initial method for evaluating patients, being able to differentiate calculus, cystic disease of the liver and cystic disease of the biliary tract. If a biliary tract dilatation with an obvious obstructive factor is detected, the next MRCP test, which has a high sensitivity (80-100%), [18] is the next one that has traditionally been performed without the risks inherent in ERCP. However, in the present report, since the patient had already undergone the procedure for biliary drainage with prosthesis, it was decided to perform an ERC before MRCP. ERC has been shown to be a viable alternative for the diagnosis of choledochal cysts and / or pancreaticobiliary junction abnormalities, since MRCP still has a limited capacity to detect minor ductal anomalies or small choledochocoele [19].

The treatment of subtypes I and IV of choledochal cysts consists of complete excision of extrabiliary biliary cyst, to the level of communication with the pancreatic duct, cholecystectomy and restoration of biloenteric continuity [3]. Attention should be paid to the preservation of the pancreatic duct and to the total removal of the intra-pancreatic portion of the cyst, as residues may lead to secondary carcinogenesis of the bile duct and morbidity. In the case of subtype V, the extent of liver resection depends on the nature of the extrahepatic component of the choledochal cyst. In some cases, the decision to excision of the extrahepatic duct is reasonable, since infrahepatic duct dilatation usually resolves spontaneously between three and six months [20]. It is important to note that the rates of bile stenosis, lithiasis or reoperation are significantly higher for patients with excision of the extrahepatic duct compared to patients submitted to extrahepatic duct excision concomitant with hepatic resection. Thus, hepatectomy is justified for subtype V cysts with a significant intrahepatic component. For cases of severe chronic pancreatitis associated with choledochal cyst and atrophic pancreatic head due to the anomalous junction of the pancreatic duct and the bile duct, a duodenopancreatectomy may be indicated [21].

The patient of the present report had a symptomatic choledochal cyst of the type 1b, according to the Todani classification, which is why a highly complex surgical procedure (Whipple) was chosen in order to avoid risks of residual neoplasms due to incomplete resection of the cyst [22]. Currently, the standard treatment for cystic disease of the bile duct is surgical resection, that is, excision of the cystic lesion, with hepatojejunostomy in Roux-en-Y, [22] when possible, or with hepatectomy if indicated [22]. In many hospitals, however, partial excision of the lesion with biliary intestinal Anastomosis continues to be indicated to treat cysts of subtype I, despite the significant risk of future complications.

Conclusion

The reported case has some divergent characteristics of the reported cases of choledochal cyst. Here, for diagnostic purposes, ERC was done before MRCP, the patient did not present the classic triad of symptoms that accompanies the choledochal cyst and the surgical procedure performed was very complex (Whipple). The reported case also shows the importance of always revealing the diagnostic hypothesis of choledochal cyst in patients with persistent pain in right hypochondrium concomitant with laboratory changes of cholestasis, since the patient had already received treatment without resolution of the condition.

References


