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Case Report

Malignant Transformation: Dreadful Complication of A Choledochal Cyst

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Abstract

Choledochal cysts are a rare congenital anomaly, the malignant degeneration is the most dreadful complication of this pathology with a very unfavorable prognosis, and we report an observation illustrating this complication and its prognostic in a young woman taken in charge in our unit.

Keywords: Choledochal cyst; malignant transformation.

INTRODUCTION

Considered a true pre-cancerous condition, choledochal cyst is a rare congenital anomaly resulting essentially from a bilio pancreatic junction anomaly, and requiring adequate surgical management.

Considered a true pre-cancerous condition, cystic dilatation of the choledochus and a rare congenital anomaly resulting essentially from a biliopancreatic junction anomaly, and requiring adequate surgical management.

OBSERVATION

We report the case of a 31 year old patient admitted for management of cholestatic jaundice associated with epigastric pain, the abdominal ultrasound had revealed a cystic formation of the main biliary tract, which was completed by MRI (Figure 1) which showed a bile duct dilated to 5 cm thick and tortuous down to the bottom with visibility of the Wirsung achieving a grade I according to the Todani classification.

The patient Benefited surgical treatment with first resection of the bile duct cyst (Figures 2, 3, 4, 5) and sent for extemporaneous examination, the result was in favour of a malignant transformation and the procedure was completed by a cephalic duodenopancreatectomy. The postoperative follow-up was simple and the anatomopathological examination was in favour of a biliary adenocarcinoma. 5 months after surgery the patient was admitted with diffuse abdominal pain with vomiting and alteration of the general condition the abdominal CT scan was in favour of generalized peritoneal carcinosis, patient died 6 months postoperatively.

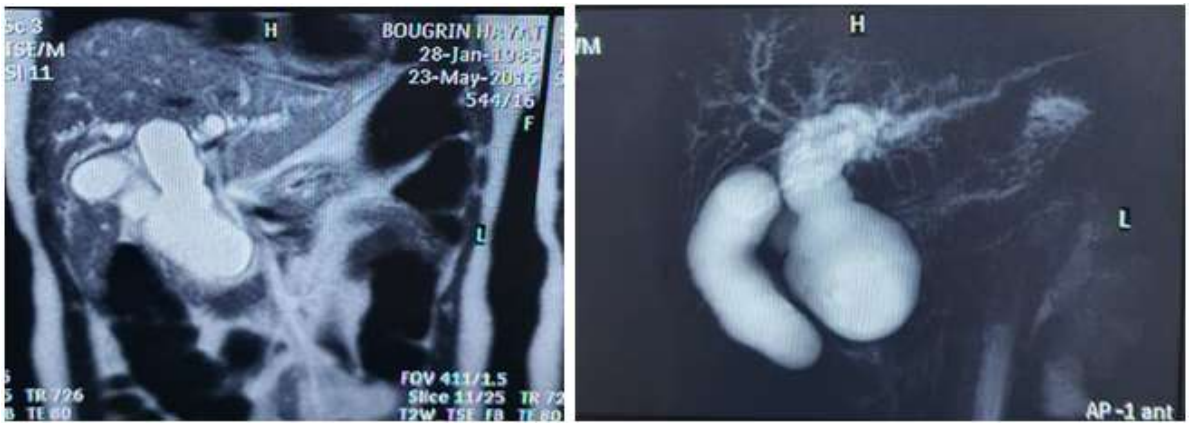
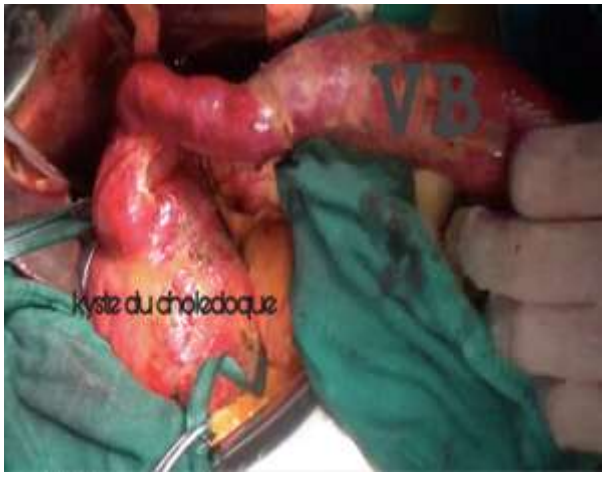
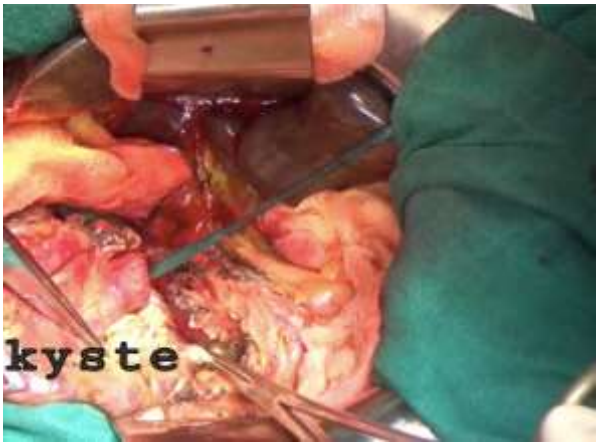


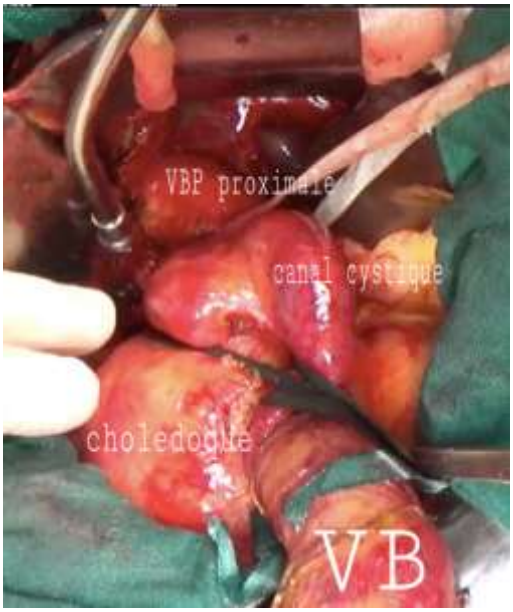
Fig-2: Total excision of the cyst with intramural dissection of the bile duct



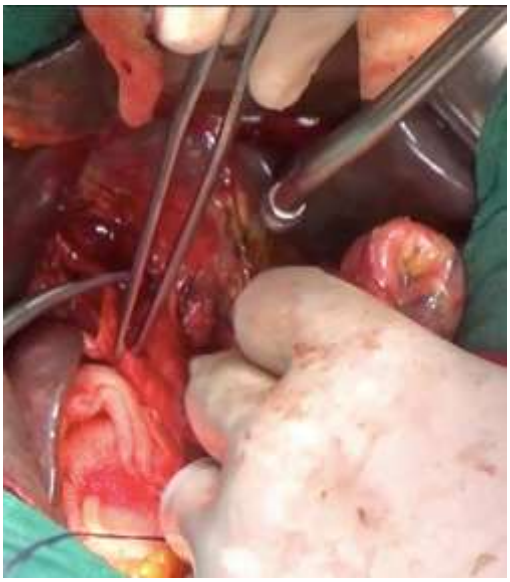
A: Cholédoque dissected and put on lake



B: Section at the low bile duct after intracancreatic dissection

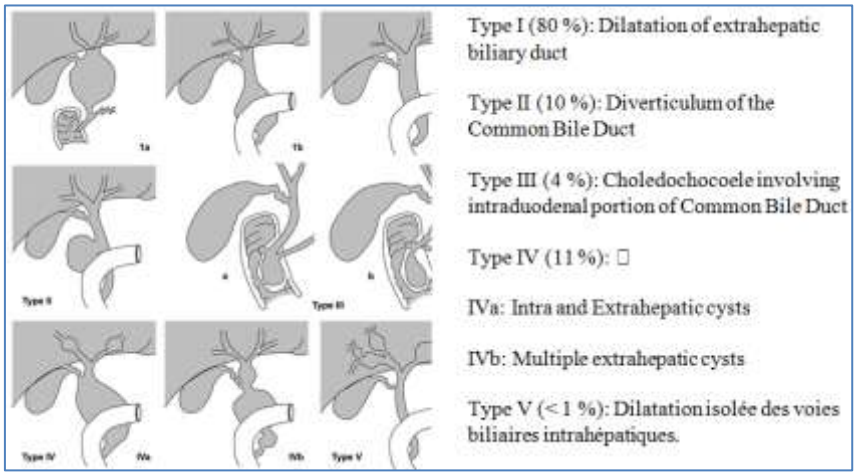


C: Dissection of proximal and distal bile duct



D: Section of the main bile duct at the level of the upper biliary convergence

Fig-3: Todani Classification



DISCUSSION

Choledocal cyst is a rare malformation of the bile ducts, described by VATER in 1723 and later [1]. It is frequently found in infants and young children in the first decade of life [2] but can occur at any age. The incidence of CC ranges from 1 in 100,000 to 1 in 150,000 in Western countries [3] to 1 in 13,000 in Japan [4], with a clear female predominance reported by most authors of 70-80% [5]. Many theories have been proposed to explain the origin of these malformations. The most commonly accepted theory is the one put forward by Babbit in 1969, which incriminated an anomaly of the biliopancreatic junction [6]. This malformation of fusion between the biliary and pancreatic ducts is characterized by three criteria a common duct more than 15 mm long; an extraduodenal junction of the two ducts at a distance from the sphincters; an angle of connection greater than 30° [7]. The triad of right hypochondrium pain with hepatic colic, jaundice and a palpable right subcostal mass is very suggestive, but only occurs in 20 to 63% of cases. Most often, the picture is reduced to abdominal pain in adults and jaundice in children. DKC can be long-lasting and may be discovered accidentally or as a result of complications [2,4,5]. The radiological step is essential for diagnosis. Hepato-biliary ultrasound shows an independent cystic mass of the gallbladder or the existence of an enormous dilatation of the bile ducts. This is more difficult in the presence of a cystic pocket full of stones. It also makes it possible to assess the state of the intrahepatic bile ducts [8]. The CT scan shows a very limited fluid tumour extended between the portal confluence and the duodenum, supplemented if necessary by a delayed cholangio-scan [9]. Direct opacifications of the bile ducts (endoscopic retrograde cholangio-graphy and transepatic cholangiography) allowing the jaundice to be linked to its cause were previously indispensable for preoperative diagnosis by clarifying the image of the biliopancreatic junction, currently less and less used with the advent of magnetic resonance imaging, a non-invasive examination that does not require contrast and allows reconstruction of the biliary tree, allows diagnosis and classification of lesions with high sensitivity (70% to 100%) and specification (90% to 100%) [10]. The Todani classification is the most widely used to characterize lesions (figure 6) [11].

Apart from surgical treatment, the evolution is unfavourable, marked by the occurrence of chronic cholestasis and infection followed by the development of secondary biliary cirrhosis. Acute complications such as recurrent angiocholitis, liver abscess, septicaemia, pancreatitis and more rarely DKC rupture or perforation leading to biliary peritonitis [12]. Cancerization is a rare complication but makes the prognosis bleak, the risk of DKC degeneration is 10% to 30% [13]. It occurs in young adults. In 80% of cases it is a cholangiocarcinoma. The incidence of this cancer increases with age [14] and after cysto-digestive shunts or after incomplete removal [15]. The treatment is surgical (figure 7), Total excision of the cyst with intramural dissection of the bile duct to its intra pancreatic portion followed by hepatic-jejunal anastomosis on a jejunal loop excluded in "Y" appears to be the procedure of choice [12], Laparoscopic resection gives results comparable to open resection in retrospective analyses [16,17]. The benefit of this type of surgery remains to be demonstrated because the prognosis is generally poor and survival rarely exceeds two years [18].

CONCLUSION

The risk of malignant transformation justifies surgery for cystic dilatations of the bile duct by early rese, whatever the age with intramural dissection of the common bile duct up to its intra pancreatic portion, accompanied by a hepatico-jejunal anastomosis on a Y-mounted loop.

Conflicts of Interest: No conflicts of interest.

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