

Choledochal Cysts: A Case Report of Todani Type VI

Mariama Jarti^{1*}, Hanane El Abbassi¹, Adil Ait Errami¹, Sofia Oubaha¹, Zouhour Samlani¹, Khadija Krati¹¹Department of Gastroenterology, Mohamed VI University Hospital, Marrakech, MoroccoDOI: [10.36347/sjams.2021.v09i09.019](https://doi.org/10.36347/sjams.2021.v09i09.019)

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*Corresponding author: Mariama Jarti

Abstract

Case Report

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. A new type, type VI, causing dilation of the cystic duct between the neck of the gall bladder and the common hepatic duct (CHD) has been described in medical literature which is the rarest of all these subtypes. They are commonly observed in middle aged females and are mostly symptomatic. Most of these cysts need magnetic resonance cholangiopancreatography (MRCP) for accurate diagnosis. Excision of the Choledochal cysts has shown excellent results; hence early diagnosis and management are recommended. We present an interesting case of such a rare cyst and its management in a middle aged woman.

Keywords: Choledochal cysts, Type VI, adult.

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INTRODUCTION

Choledochal cysts (CDC), are rare congenital dilations involving the extra hepatic biliary apparatus with or without dilation of the intrahepatic bile ducts. first described in 1723 by Vater [1]. It is a rare and predominantly female condition with pain as the main symptom. Its main risk is cancer. The TODANI classification establishes five groups of congenital cystic dilatations of the bile ducts. Complete surgical excision remains the treatment of choice. New type, type VI, causing dilation of the cystic duct between the neck of the gall bladder and the common hepatic duct (CHD) has been described in medical literature which is the rarest of all these subtypes. [2]. We report the observation of a patient with a cystic dilatation of the main bile duct, we recall the main characteristics of this pathology which can have a serious and fatal evolution.

CASE REPORT

A 30-year-old female presented with a history, since childhood, of recurrent dull non-radiating upper abdominal pain, usually lasting for 2-3 days, occurring every 2-3 months and relieved with pain medications. She had been taking proton pump inhibitors, prokinetic agents and enzyme supplements for the previous 4 years. She had no history of addiction or allergies to any drug or food.

On examination, the patient was afebrile with normal vital signs. She was not anemic or jaundiced. Abdominal palpation confirmed tenderness in the

epigastrium. The full blood count showed leukocytosis of 12000/mm³ and serum amylase 174U/L (normal 30–110U/L) and lipase 1297U/L (normal 23–300 U/L) were elevated. Laboratory values revealed total bilirubin of 1.2mg/dl, direct bilirubin 0.7mg/dl, alkaline phosphatase 189IU/L, AST 71U/L, ALT 73U/L. The hepatitis screens were negative. All other laboratory values were within normal limits. A diagnosis of acute pancreatitis was made.

Ultrasonography of the abdomen identified the gall bladder to be normal and the intrahepatic ducts were not dilated. A large choledochal cyst was noted high in the portahepatis. The pancreas appeared swollen and echolucent consistent with acute pancreatitis. Magnetic resonance cholangiopancreatography (MRCP) (Fig. 1) revealed a fusiform cystic dilatation of the CD measuring 4.1 cm, with a parallel course and low implantation. There was associated dilatation of the right and left hepatic ducts, and the distended gallbladder showed no evidence of cholecystitis, cholelithiasis or choledocholithiasis. There were no lymphadenopathies or other lesions observed at the pancreatic-duodenal junction. With the diagnosis of cystic malformation of the cystic duct (Todani type VI). After management of acute pancreatitis, a laparoscopic exploration was performed, finding a distended gallbladder that was normal in appearance, with a dilated the cystic duct up to its union with the bile duct. Cholecystectomy was performed and total cyst excision and Roux-en-Y hepaticojejunostomy. The postoperative

course was uneventful and patient was discharged on postoperative day 3 after surgery.

DISCUSSION

Bile duct cysts are a rare medical condition and are more frequent in children. However, the disease is becoming increasingly common in adults. They are classified by Todani *et al.* [2] into five subtypes where type I CDC is the most common, (60%) causing fusiform dilation of the common biliary duct apparatus. Type II involves saccular diverticulum of the CBD, type III involves perivaterian part of CBD, type IV involves multiple focal dilations of the bile ducts which are further subdivided into extrahepatic with intrahepatic involvement (4a) and extra hepatic involvement only (4b). Type V involves the intrahepatic bile ducts only (Caroli's disease)[2].

Choledochal cysts in adults lie dormant, present with non-specific symptoms, or are detected while undergoing imaging studies for symptoms related to its complications, such as hepatolithiasis, acute cholecystitis, acute or chronic pancreatitis, gastric outlet obstruction, portal hypertension.[3] It is important to note the risk of malignancy. The incidence of this is higher when diagnosis of cystic disease of the bile duct occurs at a more advanced age: 2% at 20 years and 43% at 60 years. Type I and Type IV cysts are pose the greatest risk of malignant degeneration. Type V poses the least risk (7%) [4].

Concerning diagnostic imaging modalities, ultrasound has a primary role in the imaging of the biliary tree, related diseases and detection of choledochal cysts, and an Magnetic resonance cholangiopancreatography is ideal to delineate the entire biliary system including the course of the cystic duct, presence or absence of abnormal pancreaticobiliary duct junction, GB thickening, presence of gall stones, intrahepatic biliary radicle, and CBD involvement. Endoscopic retrograde cholangiopancreatography (ERCP) is invasive, though providing the same information and detail regarding the biliary system as magnetic resonance cholangiopancreatography. ERCP and Tc-99m Hydroxy Imino Diacetic Acid (HIDA) scan can be used for diagnosis but are not commonly used[2].

Typical radiologic abnormalities that are specific to type VI collecting duct carcinoma include dilatation and squaring of the cystic duct, acute angulation of the common hepatic duct and cystic duct junction with a distinct plane present between the dilated cystic duct and common hepatic duct, a normal or wide (Mirrizi syndrome) opening of the cystic duct to the The common bile duct, a normal The common bile duct and associated that abnormal pancreaticobiliary duct junction[5].

The management approach of choledochal cysts depends on the cyst type and the extent of hepatobiliary pathology. As a rule, all cysts should be resected, and bile flow should be restored. The treatment of choice is total cyst excision and Roux-en-Y hepaticojejunostomy, especially for patients with choledochal cysts type I, type II and type IV. Total cyst excision might be difficult in patients with very large cysts and incomplete cyst excision with bilioenteral anastomosis is a surgical option. However, these patients have to be regularly followed up because of the risk of malignance. The therapeutical approach in patients with type III cysts depends on cyst size, and it can be treated endoscopically or surgically[6].

CONCLUSION

Choledochal cyst is a rare anomaly that is considered to be premalignant. It often poses a diagnostic dilemma. The typical presentation of this condition is non-specific. Early diagnosis and management are critical. This activity discusses the evaluation and management of choledochal cysts and highlights the role of the interprofessional team in managing patients with this condition.

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Fig-1: Magnetic resonance cholangiopancreatography revealing a todani type Choledochal Cysts

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