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Choledochocele Presenting as Recurrent Pancreatitis: A Case Report

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Abstract **Case Report**

Introduction*:** Choledococele is a rare congenital abnormality involving the intramural segment of the common bile duct. We report the case of a patient presenting with acute pancreatitis who was diagnosed with choledochocele. ***Case report: A 17-year-old boy, with a history of hospitalization 2 years ago for acute pancreatitis stage E, admitted for acute epigastralgia radiating to the back with notion of vomiting. Abdominal examination revealed epigastric tenderness without other abnormalities. Laboratory data reported leukocystosis (WBC: 16000/ul); elevated CRP at 111 mg/l; elevated Lipase at 412 IU/L. Abdominal CT scan shows stage D pancreatitis with an intra duodenal cystic formation in front of the ampulla of Vater suggesting a choledochocele. Abdominal MRI demonstrated a cystic dilatation of the lower bile duct classified as Todani type III. Echo endoscopy was performed indicating an ampullary cystic formation measuring 22,8x15,3mm which continues with the CBD evoking a choledocal cyst with presence of stones in intra-cystic. As a result of these findings, the patient was diagnosed with acute pancreatitis secondary to intraduodenal choledochocele. He had surgery with resection of the choledochocele. ***Conclusion:*** Choledococele is extremely rare. The diagnosis is facilitated by advances in imaging. Management and therapeutic choice are not yet consensual regarding radical surgical treatment or conservative endoscopic treatment.

Keywords: Choledochocele, Bile duct cyst, pancreatitis.

INTRODUCTION

Choledochal cyst is one of the most common congenital anomaly of the common bile duct (CBD). Various types have been described. The most known classification is by Todani *et al*., in 1977, with five types of dilatation [1].

Type III or choledochocele is extremely rare and diagnosis is complicated by absence of specific clinical signs. The complications are angiocholitis or pancreatitis [2].

We report the case of a patient presenting with acute pancreatitis who was diagnosed with choledochocele.

CASE REPORT

A 17-year-old boy, with a history of hospitalization 2 years ago for acute pancreatitis stage E, admitted for acute epigastralgia radiating to the back with notion of vomiting. He had no fever or jaundice. He had no family history of pancratic or liver problems.

Abdominal examination revealed epigastric tenderness without other abnormalities. Laboratory data reported leukocystosis (WBC: 16000/ul); elevated CRP at 111 mg/l; elevated Lipase at 412 IU/L; normal AST, ALT and alkaline phosphatase; and slightly elevated total bilirubin at 34.

Abdominal CT scan shows stage D pancreatitis with an intra duodenal cystic formation in front of the ampulla of Vater suggesting a choledochocele.

The patient was rehydrated with intravenous fluids and the pain controlled with analgesic.

Further investigation of the etiology of the pancreatitis included abdominal MRI which demonstrated a cystic dilatation of the lower bile duct classified as Todani type III (Figure 1).

To confirm the diagnosis, an echo endoscopy was performed indicating an ampullary cystic formation measuring 22,8x15,3mm which continues with the CBD evoking a choledocal cyst with presence of stones in intra-cystic (Figure 2 & 3).

As a result of these findings, the patient was diagnosed with acute pancreatitis secondary to intraduodenal choledochocele. He had surgery with resection of the choledochocele.

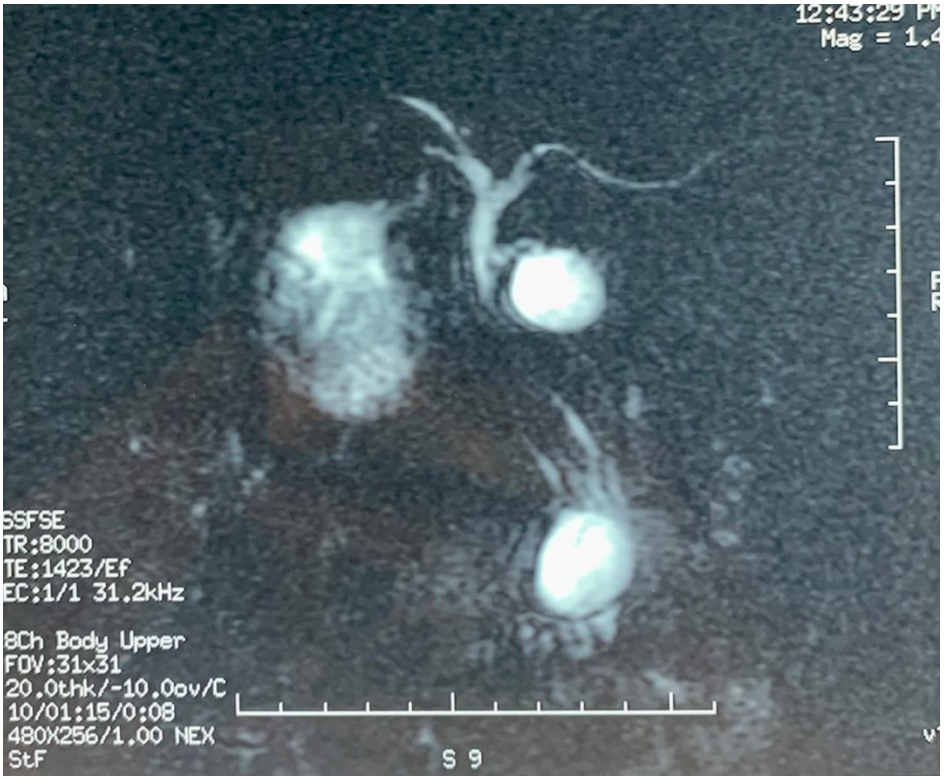

Figure 1: MRCP shows cystic dilatation of the lower bile duct

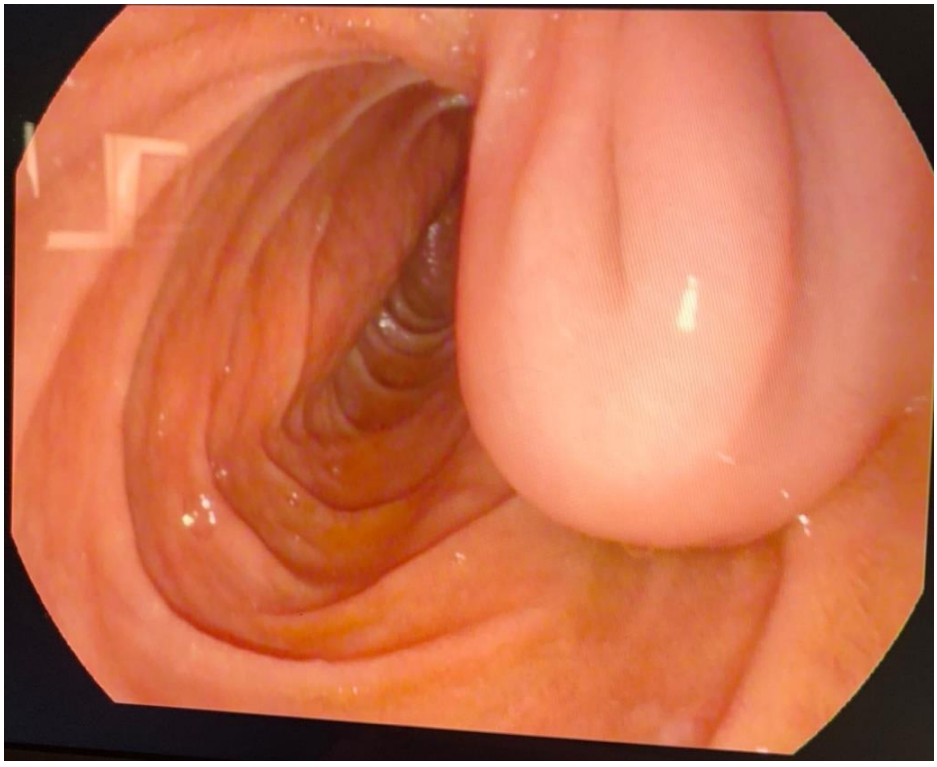

Figure 2: Intraluminal duodenal cyst

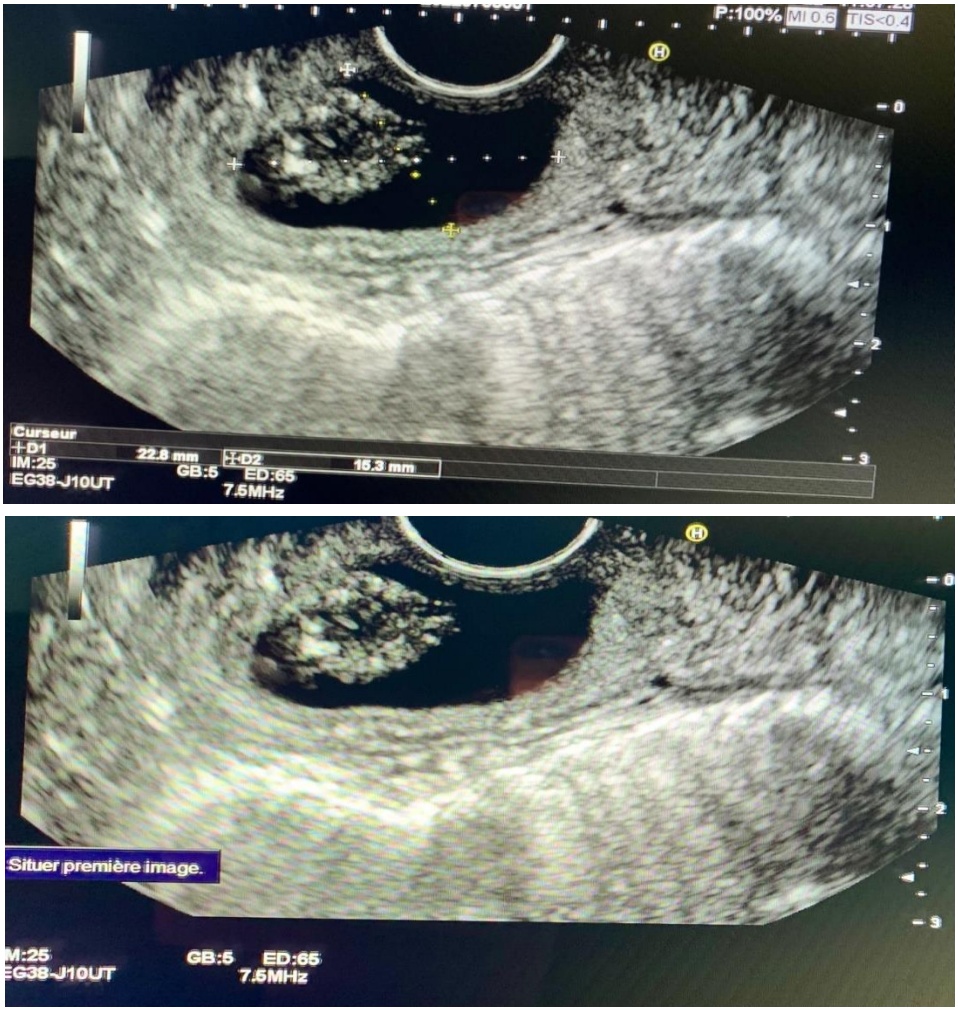

Figure 3: Echo endoscopy demonstrated an ampullary cystic formation 22,8x15,3mm → choledocal cyst with stones in intra-cystic

DISCUSSION

First described by W.C. Wheeler in 1915, choledochocele is a rare subtype of biliary cystic dilatation. It represents between 1.7 and 2% of cystic dilatations of the bile ducts [3].

Choledococele is a rare congenital abnormality involving the intramural segment of the common bile duct. It is considered the rarest of the congenital cystic dilatations of biliary tracts.

It is more prevalent in Asian countries than Western countries, with predilection for females. 67 to 80% of patients are diagnosed by the age of ten years [4-6].

Etiopathogeny of choledochocele remains unclear regarding congenital and acquired origin. The congenital theory involves a congenital malformation of the biliopancreatic junction [7]. Due to the increased frequency of this disease in adults, the acquired theory suggests a chronic inflammation of the papilla leading to dilatation of the intra luminal portion of the principal bile duct [4, 5, 8].

Todani's classification divides cystic lesions into 5 main categories. Type IA is cystic dilatation of the main bile duct, type IB is focal segmental dilatation, usually distal to the main bile duct, and type IC is fusiform dilatation of the main bile duct and the main hepatic duct. Type II is the real diverticulum of the extrahepatic duct. Type III is choledocele. Type IVA is dilatation of both intra- and extrahepatic ducts (segmental cysts) and type IVB is dilatation of multiple segments of extrahepatic ducts only. Type V is Caroli disease [1].

The clinical presentation is generally a triad of abdominal pain, jaundice and a palpable mass seen in 13 to 63% of the patients [9]. The symptoms are variable and depend on age.

CT is a major method of diagnosis of hepatobiliary disease and it is performed when the ultrasound result is not certain, the mass is suspicious and segmental obstruction is present [10].

Although ERCP has long been considered the gold standard for diagnosing choledochal cysts and evaluating pancreatobiliary junction abnormalities in

the past decade, numerous studies have shown that MRI cholangiopancreatography is also sensitive [11, 12].

The fact that ERCP is an invasive method, has a mortality rate of 0.2-1%, a risk of 1-7% morbidity, the need for experienced operators limit its use for diagnostic purposes. MRCP is a reliable non-invasive procedure for treatment of pancreatic and biliary system diseases [13-15]. However, ERCP provides therapeutic intervention.

The most common complications of choledocholithiasis, cholelithiasis, pancreatitis, angiocholitis, cyst rupture, and the development of cancer [12, 16]. On rare cases, patients with choledochal cysts may develop cholangiocarcinoma at a rate of 9-28%, an incidence 25-40 times higher than normal population [9, 17].

The therapeutic management is still not consensual. Several therapeutic options are possible: surgical or endoscopic treatment. For some authors, surgery remains the treatment of choice allowing a definitive and radical treatment of choledochocele and avoiding the risk of degeneration [7, 18-21].

Most authors consider endoscopic retrograde cholangiopancreatography as the reference treatment since 1974 when Dehyle *et al*., [22] described the first endoscopic biliary sphincterotomy [23-26]. The endoscopic interventions performed include an endoscopic sphincterotomy, excision of the cyst with a diathermy loop, or a combination of the two procedures [23, 27]. Indeed, the risk of degeneration exists but it is less frequent in case of type I and II cystic dilatation lesions [28, 29]. In this case, surgical treatment was a therapeutic option chosen by the patient's parents.

CONCLUSION

Choledococele is extremely rare. The diagnosis is facilitated by advances in imaging. Management and therapeutic choice are not yet consensual regarding radical surgical treatment or conservative endoscopic treatment.

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