A CASE-REPORT OF ACUTE PANCREATITIS ASSOCIATED WITH PANCREATICOBILIARY MALJUNCTION

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ABSTRACT

Background: *Pancreaticobiliary maljunction (PBM) is a rare congenital anomaly and possess potential risk of cancer.*

Aims: *To present of a case of acute pancreatitis associated with PBM and review the literature.*

Method: *Case-report study.*

Results: *A case of 16 years old female presented with acute pancreatitis and common bile duct (CBD) stones. The main features were abdominal pain, increased serum pancreatic enzymes and abnormal liver function tests. CT-Scan, MRCP and ERCP detected common bile duct dilation (10mm), long common channel (3-4cm) and suspected common bile duct stones. The patient was operated by laparoscopic surgery with gallbladder-CBD excision and Roux-en-Y hepaticojejunostomy and was discharged in a good condition.*

Conclusions: *PBM should be considered as a potential cause of recurrent pancreatitis, especially in young patients. The risk of cancer is well validated and needs appropriate management.*

Key words: *Pancreaticobiliary maljunction*

I. INTRODUCTION

Pancreaticobiliary maljunction (PBM) is a rare congenital anomaly where the pancreatic and bile ducts join outside the duodenal wall. Consequently, pancreatic juice communicates freely with bile duct and may lead to multiple complications such as: cholangitis, pancreatitis and biliary cancer[5]. PBM is frequently associated with congenital biliary cyst. If without biliary dilatation, the clinical presentations are usually non-specific and so easily missed. Hence, we present a case of acute pancreatitis associated with PBM that was incidentally detected and then operated at Trưng Vương hospital.

II. CASE PRESENTATION

A patient of 16 years old female was hospitalized for recurrent acute pancreatitis.

Laboratory examinations at hospitalization

Table 1. Laboratory examinations at hospitalization

Laboratories	Value	
WBC	16.94	K/µL
RBC	4.00	M/µL
Hb	11.9	g/dL
Bilirubin T	131.9	µmol/L
Bilirubin D	77.9	µmol/L
Bilirubin I	54	µmol/L
AST	170	U/L
ALT	249	U/L
Amylase	464	U/L
Lipase	709.08	U/L

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Imaging at hospitalization

US: Intra / Extra hepatic biliary dilatation

CT scan: Intra / Extra hepatic biliary dilatation, suspected CBD stones 5mm and abnormal pancreatic duct

Pre-operative Imaging to confirm PBM

- MRCP: Intra / Extra hepatic biliary dilatation, CBD stones and PBM (Fig. 1)

- ERCP: Normal papilla of Vater, CBD stones, abnormal junction between pancreatic duct and CBD situated 4cm proximal to papilla of Vater.

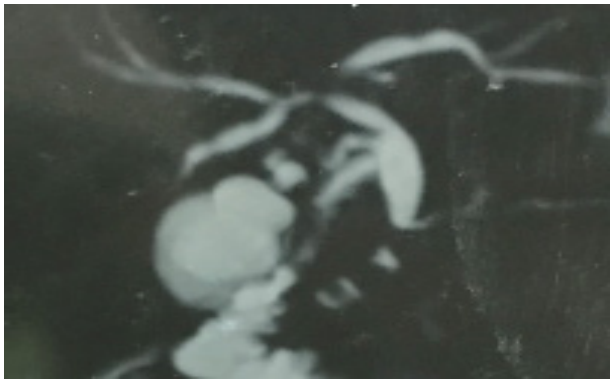

Fig. 1- MRCP

Laparoscopic surgery was indicated for biliary pancreatitis with suspected PBM.

The intra-operative modalities utilized to confirm PBM

- Dosage of pancreatic enzymes in CBD bile:

Amylase / Bile: 103751 U/L

Lipase / Bile: 545564 U/L

- Intra-operative cholangiography via CBD: PBM, long common channel with 4cm length similar to MRCP (Fig. 2)

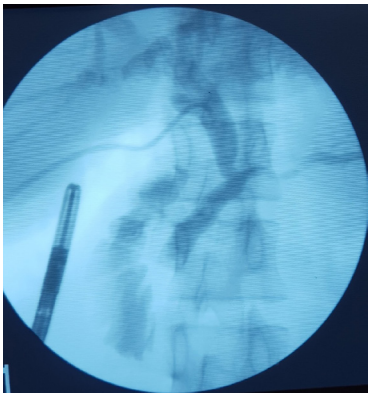

Fig. 2- Intra-operative cholangiography

- Choledochotomy and cholangioscopy: Some stones of 3-4mm in CBD, pancreatic duct and common channel. The junction of pancreatic duct and CBD was identified exactly similar to previous diagnostic images (Fig. 3). The stones were smooth, light gray-yellow color, and friable, like protein plug. The sphincter of Oddi was patent with softly contraction, the rest of biliary tree was smooth without stenosis and stone.

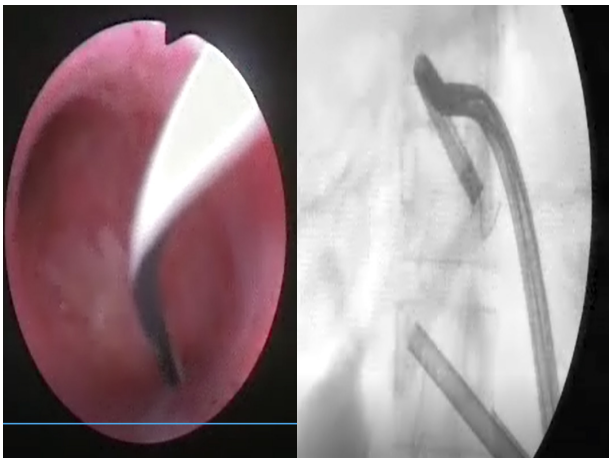

Fig. 3- Identification of pancreaticobiliary junction with cholangioscopy

Bile duct and pancreatic duct were then irrigated for clearance of stones. Cholecystectomy and extrahepatic duct resection were performed with the proximal end at hepatic duct and the distal end at 5mm proximal to pancreaticobiliary junction. The biliary was reconstructed by Roux-Y bilioenteric anastomosis.

Postoperative recovery was normal, the patient was discharged uneventfully. Pathology result was chronic inflammation of gallbladder and bile duct.

III. DISCUSSION

PBM with the pancreaticobiliary junction located outside the duodenal wall, and the sphincter of Oddi cannot exert any influence or control on the junction (Fig. 4). The free flow and reflux between pancreatic juice and bile may result in many consequences: pancreatitis, cholangitis, cancer of the gallbladder and bile duct.

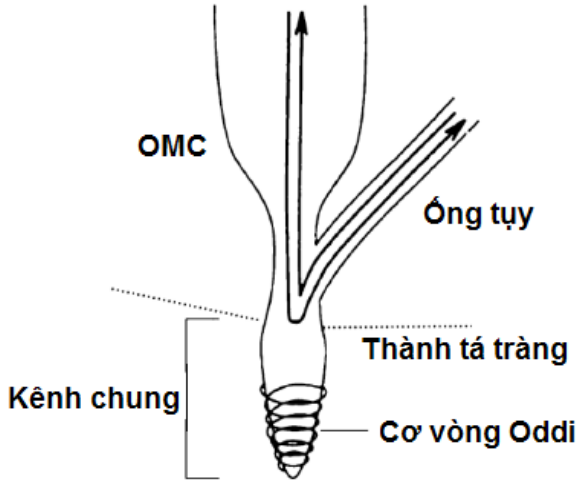

Fig. 4- PBM
(*Source: Kamisawa, 2012*) [5]

The diagnostic criteria of PBM in the literature are: pancreaticobiliary junction lies outside the duodenal wall demonstrated by imaging or anatomically by operation or autopsy. The signs in imaging consist of: common channel longer than 1.5cm, the junction is located outside the duodenal wall, and the sphincter fails to exert any effect on the junction [5], [6].

The high pancreatic enzymes levels in the bile duct and/or the gallbladder (usually > 10000U/L) serve as an auxiliary diagnosis [5], [6].

We utilized the diagnostic modalities proposed by many authors to confirm PBM: CT scan, MRCP, ERCP, Cholangiography, and pancreatic enzymes in the bile (except EUS) [5]. Particularly, because of dilated CBD in this case, we performed also cholangioscopy to identify the PBM and to examine completely the biliary tree. All the results satisfied the diagnostic criteria of PBM.

Normally, PBM is classified as PBM with and without biliary dilatation. The PBM with biliary dilatation is common and usually termed "congenital biliary cyst" (especially type Ia, Ic and type IVa), but in minor form the bile duct may be slight dilated, unlike the "cystic" form [3], [5].

The diagnosis of PBM with biliary cyst is practically not difficult due to its typical presentation in both children and adults, and eventually easily found on US or CT scan. On the contrary, the diagnosis of PBM without biliary dilatation or with slight dilatation is very easily missed due to multiple atypical clinical situations: pancreatitis, cholangitis, gallstones, in which the usual imaging techniques (US, CT scan) are incapable to find out PBM.

In presented case, initially PBM was not noticed but the incidental finding of abnormal pancreatic duct on CT scan forced us to realize additional techniques to verify PBM.

PBM may be classified based on the pattern of pancreaticobiliary junction (Komi's classification). Recently, a new pediatric PBM classification was proposed by Japanese authors, including 4 types (Fig. 5). According to this, our case is type B.

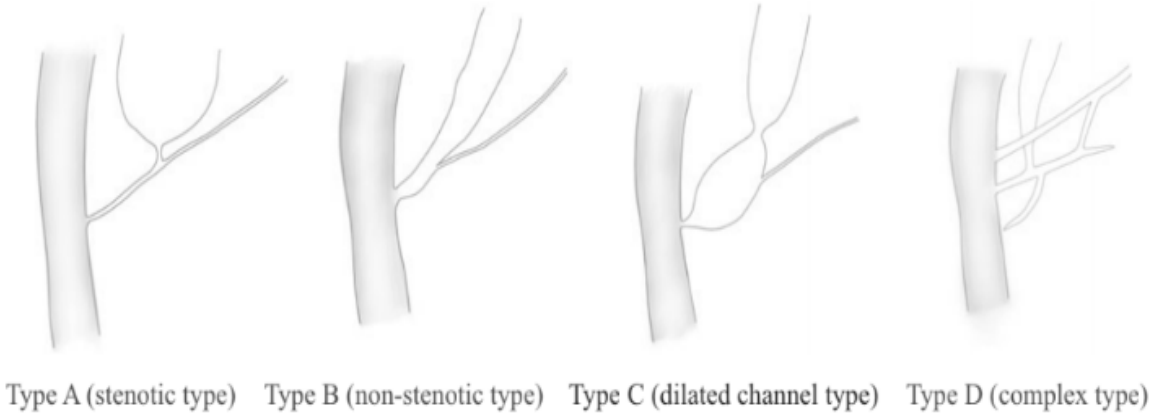

Fig. 5- Pediatric PBM classification
(Source: Urushihara, 2017) [7]

The risk of biliary and gallbladder cancer is well validated. According to Kamisawa, the prevalence of biliary tract cancers in adult PBM patients with and without biliary dilatation was 21.6 and 42.4% respectively. In PBM patients with biliary tract cancers, bile duct and gallbladder cancers were present in 32.1 and 62.3% of patients with biliary dilatation and in 7.3 and 88.1% of those without biliary dilatation respectively.

The preferred treatment of PBM with biliary dilatation is cholecystectomy and extrahepatic bile duct resection close to pancreaticobiliary junction and Roux-Y hepaticojejunostomy (also called "biliary diversion"), it's exactly our choice in this case.

On the other hand, there is considerable debate about the treatment of PBM without biliary dilatation. In many institutes, prophylactic cholecystectomy is sufficient because most biliary cancers in PBM patients without biliary dilatation are gallbladder cancers. However, some surgeons suggest that the extrahepatic bile duct should be removed together with the gallbladder in PBM patients without biliary dilatation for the prevention of bile duct cancer [1], [8].

In patients operated with the above biliary diversion procedure, the malignancy risk also remain. In longterm follow-up, the incidence of biliary cancer range 0.7%–5.4% [6]. Thus, the postoperative patients should be followed-up for life.

According to Kamisawa, PBM without biliary dilatation rarely have symptoms, so that the majority of such patients remain undiagnosed until they present with advanced-stage gallbladder cancer. So the most important problem is creating a strategy for early detection of PBM and prevention of cancer [8]. With our presented case, we found some noticeable features as: recurrent pancreatitis and young age (16 yrs), these are just the characteristics of acute pancreatitis in PBM.

Kamisawa proposed to perform MRCP for patients with gallbladder wall thickening on screening ultrasonography.

The incidence of PBM is relatively rare, approximately 1 in every 1,000 persons is affected by this disease in Japan [7], and this incidence is 4.1% in South Korea (following 10,243 ERCP cases). However, once this condition is misdiagnosed, the long-term consequences may be severe.

Therefore, we try to propose certain clinical situations in which PBM should be screened:

- Acute pancreatitis with non obvious etiology, recurrent pattern and young age
- Biliary dilatation on imaging without evident cause
- Gallbladder wall thickening on screening ultrasonography
- The cases of cholecystectomy for lithiasis or polyp, dosage of pancreatic enzymes in the gallbladder should be performed
- The cases of cholangiography, CT scan, MRCP, ERCP for any reason, PBM should be screened
- The cases of intervention on bile duct, dosage of pancreatic enzymes in bile and cholangiography should be performed
- The cases of gallbladder or biliary cancer

IV. CONCLUSIONS

PBM is a rare congenital malformation (with high incidence in Asia), susceptible to misdiagnosis (especially PBM without biliary dilatation) and may have potential risk of cancer.

The modalities and criteria for diagnosis of PBM are actually available at almost Vietnamese hospitals.

The treatment of PBM requires surgical intervention and the follow-up is for life.

Our important task is to perform the best screening strategy for early detection of PBM.

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