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Case Report

A complex pancreaticobiliary malunion causing recurrent pancreatitis in a boy: A case report

Amen Zafar,*[1] Anwar UL Haq,[2] Dawood Shehzad,[3] Mohammad Salih,[2] Gulrez Umar,[3]

1. Shifa College of Medicine, Islamabad,

2. Department of Pediatric Surgery and Gastroenterology, Shifa International Hospital, Islamabad.

3. Rawalpindi Medical University.

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ABSTRACT

Background: Pancreaticobiliary malunion (PBM) is a distinct malformation of the pancreatic and biliary ductal system, in which the junction of the pancreatic and biliary ducts occurs above the duodenal wall. PBM has varied presentations that prompt early diagnosis and management.

Case Presentation: A 9-year-old male, a known case of chronic pancreatitis, presented with sudden intense abdominal pain and vomiting. After evaluation, the diagnosis of acute on chronic pancreatitis secondary to PBM was made and the patient underwent Roux-en y hepaticojejunostomy, along with cholecystectomy and common bile duct excision.

Conclusion: The purpose of this report is to add to the literature the unique presentation of PBM in the pediatric population in order to aid in prompt diagnosis and management. This will be the first report on PBM in the pediatric population of Pakistan.

Keywords: Congenital pancreaticobiliary maljunction, Pancreatolithiasis, Pancreaticobiliary reflux.

INTRODUCTION

Pancreaticobiliary malunion (PBM) is a disease entity of the pancreaticobiliary ductal system. It is defined as a condition where the pancreatic and biliary ducts unite at an abnormal proximal site, usually above the duodenal wall. [1] PBM is broadly categorized as — PBM occurring with congenital biliary dilatation (CBD) or without the presence of biliary dilatation. The management of PBM is contentious and depends on the “Komi” classification, which influences the selection of surgical procedures and prognosis after surgery. [2] The pathology still largely remains rare. It is common in the eastern population, however, occurrence and morbidity remain unknown.

PBM has presented with congenital choledochal cysts [4], recurrent cholangitis [5], congenital hepatic fibrosis [5], pancreatolithiasis[6], cholelithiasis[6], and pancreatitis. [7] Due to varied presentations and associated with significant complications, prompt diagnosis and treatment largely rely on imaging. [3].

CASE REPORT

A 9-year-old boy, known case of chronic pancreatitis for 5 years, presented with sudden onset of intense abdominal pain along with bouts of non-bilious vomiting. Investigations done 5 years back including an MRCP that indicated choledocholithiasis, an ERCP report revealed a distal CBD stricture with proximal dilatation and an abnormal fistula between the CBD and pancreatic ampulla, while a CT abdomen with contrast showed the pancreatic duct joining the CBD in the region of the head of the pancreas with a long common channel. An initial diagnosis of PBM was made. An MRCP at a later date confirmed the diagnosis of PBM with the evidence of a false channel between CBD and pancreatic duct that extended downwards but ruled out pancreatic divisum. For 5 years, the patient did not receive any treatment, except for intravenous analgesics for recurrent abdominal pain. After evaluation, at our facility, the diagnosis of acute-on-chronic pancreatitis secondary to PBM was made on ERCP, with the failure of efforts to cannulate the CBD (Fig. 1). After a complete

Correspondence*: Amen Zafar, 5th Year Medical Student, Shifa College of Medicine. House#603, Street 69, I-8/3, Islamabad.
Email: amenzafark@gmail.com

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evaluation, the patient was taken to surgery, where dense adhesions were broken and a Roux-en y hepaticojejunostomy, along with cholecystectomy and common bile duct excision was performed (Fig. 2).

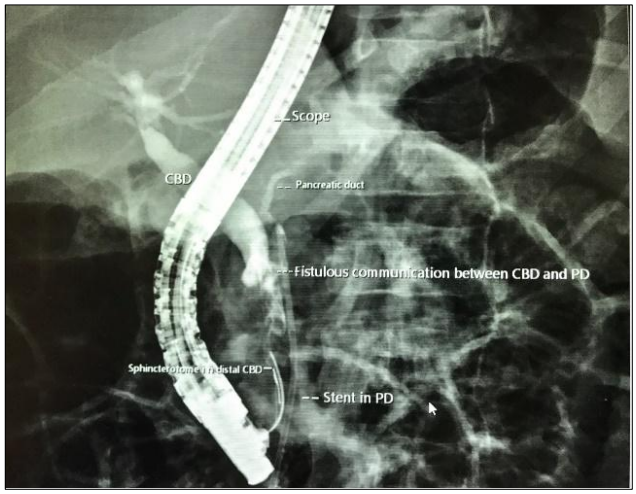

Figure 1: Preoperative Endoscopic retrograde cholangiopancreatography (ERCP) showing a fistulous communication between common bile duct and pancreatic duct and a stent being placed in pancreatic duct

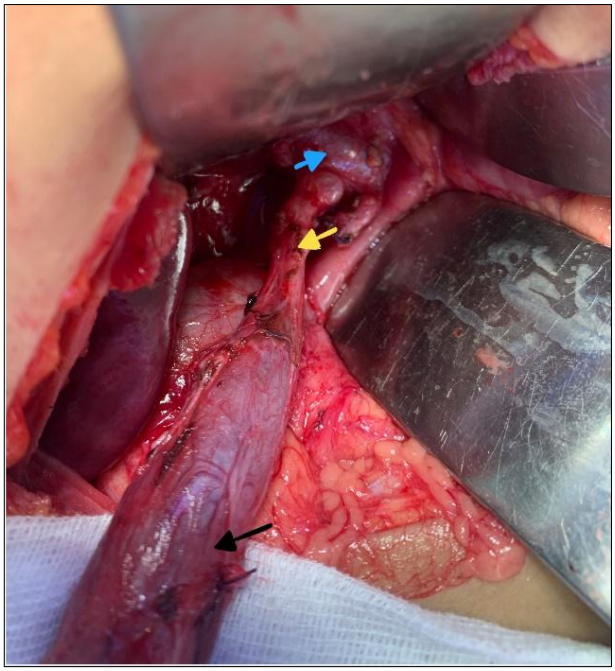

Figure 2: Shows findings during operation. Black arrow shows gallbladder. Yellow arrow specifies cystic duct. Blue arrow shows dilated common bile duct

Histopathological analysis of the biopsied specimen showed signs of chronic inflammation with predominant lymphocytes in the gallbladder along with calcific debris found in the common bile duct lumen (Fig. 3). The postoperative investigations showed a marked decrease in serum lipase from 1249 U/L to 31U/L and serum amylase from 339U/L to 38U/L with the rest of the investigations being normal (Table 1). The patient developed no complications postoperatively and was successfully discharged on day 5 after the operation. The patient was advised to report to the outpatient department in one week. On follow up the patient was doing better, reported no pain, and was counseled on home care.

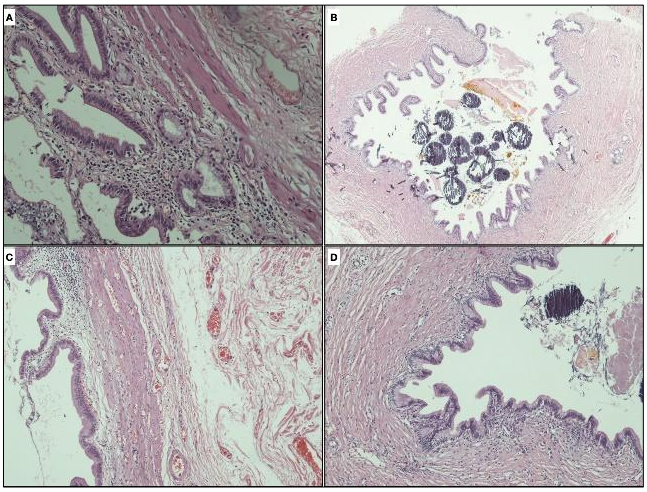

Figure 3: Histopathological findings of the resected specimen. (A) Shows gallbladder with chronic inflammation and lymphocytes. (B) Shows common bile duct with calcific debri in lumen with 10x power. (C) Shows gallbladder wall with inflammatory changes. (D) Shows common bile duct with calcific debri at 40x power.

Table 1: Investigations

Investigations	Pre-operative	Post-operative	Normal Range
Serum Lipase	1249 U/L	31 U/L	13-60 U/L
Serum Amylase	339 U/L	38 U/L	25-125 U/L
Hemoglobin	12.5 g/dL	10 g/dL	Male (13-18) g/dL
White Blood Count, Total	6,770 /UL	10980 /UL	4000-11000 /UL
Platelets	362,000 /UL	316,000 /UL	150,000-400,000 /UL
Lymphocytes	48%	14%	20-45%
Neutrophils	34%	77%	40-75 %

DISCUSSION

PBM is a congenital anomaly that is characterized by the malunion of the pancreatic duct and common bile duct outside the duodenal wall. [1] This malunion results in the defective functioning of the sphincter of Oddi. As the hydrostatic pressure inside the pancreatic duct is higher than that in the CBD, this can result in reflux of pancreatic juices into the biliary tract. Pancreatic juice contains carcinogenic substances such as phospholipase A2 and secondary bile acids which can predispose these patients to biliary tract cancers as well as chronic inflammation. [2] Similarly, the pancreas is also exposed to biliary juices and this can lead to recurrent bouts of pancreatitis [7] as in our case, along with a wide variety of other presentations. [4-6] Our patient had also developed

a high pancreaticobiliary fistula between the pancreatic duct and CBD in the region of the pancreatic head well above its ampulla which was noted during his ERCP and stenting. Such communications of the pancreas form following chronic fluid leakage and can occur on a background of acute-on-chronic pancreatitis. [8]

The therapeutic approach to this condition revolves around the prevention of mixing of pancreatic and biliary juices and their reflux into the biliary and pancreatic channels respectively. Patients that also have a congenital cyst of the bile duct need excision of the CBD to prevent cholestasis. This can be brought about by either choledochodudenostomy (end to side type) with cholecystectomy or choledochodudenostomy (Roux-en Y type) with cholecystectomy. [9] In our patient, complete excision of CBD was carried out and communication between the hepatic duct and jejunum was created in a Roux-en Y fashion along with cholecystectomy. Post-op histological examination of the removed specimens showed calcific debris in the CBD and chronic inflammatory infiltrates in the gallbladder but no metaplasia or dysplasia was found.

Biliary tract carcinoma is another significant complication of PBM with advanced cholangiocarcinoma being reported even in children as young as 11 years old. [6] For this purpose removal of the CBD and gallbladder is necessary to lower the chances of occurrence of cancer later on in life. PBM can present with or without CBD dilatation. Cholangiocarcinoma is more associated with the latter presentation. Patients without CBD dilatation are mostly asymptomatic and usually present with advanced cholangiocarcinoma.

The purpose of this report is to add to the literature the unique presentation of PBM in the pediatric population in order to aid in the prompt diagnosis, management, and follow-up of these patients. This will be the first report on PBM in the pediatric population of Pakistan.

Conflict of Interest: Nil

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