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CASE REPORT

Anomaly of The Biliopancreatic Junction with Associated Hepatic Duct Anomaly-A Rare Case Report

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Abstract

Pancreaticobiliary maljunction (PBM) or anomaly is defined as an anatomical maljunction of the biliary duct and pancreatic duct outside of the duodenal wall beyond the influence of the sphincter of Oddi. Children with this anomaly can present with choledochal cyst with recurrent attacks of acute pancreatitis, and their condition can be worse when not treated appropriately. Pancreaticobiliary maljunction or anomaly is considered to be a major risk factor for biliary tract cancer. In patients with this anomaly, free reflux of pancreatic juice into biliary tract may cause biliary tract damage. This can result in biliary malignancy. Therefore, in this condition, total excision of the extrahepatic bile duct with hepaticojejunostomy is recommended. Early diagnosis of this condition and early surgical treatment is recommended to prevent further complications. Proper follow-up is necessary to detect biliary tract malignancy in the early post-operative period, especially in patients demonstrating post-operative complications. In this article, we presented a case report involving a young male with an abnormal pancreaticobiliary junction (PBM) with choledochal cyst associated with hepatic duct anomaly, biliary stones, and pancreatic stones causing recurrent attacks of acute pancreatitis.

Key Words: *Pancreaticobiliary maljunction; Acute pancreatitis; Hepaticojejunostomy; Choledochal cyst; Hepatic duct anomaly*

Introduction

Anomalous Pancreaticobiliary junction (PBM) is a rare condition. Normally, the major pancreatic duct and the common bile duct open separately with a valve or after joining as a common channel into the second part of duodenum. The length of this common channel usually between 1-12 mm, with a mean length of about 4-5 mm. In adults, a common channel longer than 6 mm is considered abnormal while in infants, it is considered abnormal if length is longer than 4 mm [1]. In PBM, the connection or joining of the main pancreatic duct and common bile duct is located outside of the duodenal wall [2]. This junction of the common bile duct (CBD) and the pancreatic duct (PD) is critical to maintain sphincteric control of biliary and pancreatic juice drainage. If this junction is outside of the duodenal wall, it may cause regurgitation of bile and pancreatic juice resulting in lysis of CBD wall. This can result in the formation of the dilated biliary tract known as choledochal cyst [3] that could also be attributed to congenital weakness of a part of wall of duct [4]. According to the pattern of joining of the biliary duct and main pancreatic duct outside of the duodenal wall, PBM is classified into three categories.

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These are V type, B-P type, and P-B type:

V type: Pancreatic duct and common bile duct both join into the duodenal wall separately without a common channel.

B-P type: The common bile duct drains into the pancreatic duct and then forming a common channel before draining into the duodenal wall.

P-B type: Pancreatic duct drains into the biliary duct and forms a common channel before draining into the duodenal wall.

Different studies have been described that the P-B type is equal to an acute angle, and the B-P type is equal to a right angle [5,6].

In this article, we presented a case report involving a young male with an abnormal pancreaticobiliary junction (PBM) with choledochal cyst associated with hepatic duct anomaly, biliary stones, and pancreatic stones causing recurrent attacks of acute pancreatitis.

Case Report

A 17-year-old male was admitted to the hospital with complaints of pain epigastrium and vomiting for three days. The pain was radiating to his back and relieved upon sitting. History of the same complaint five months back that was relieved by conservative management. No history of jaundice or fever was recorded. No past surgical history was found. On examination, vitals were stable. Abdominal tenderness was present in the epigastrium. No palpable lump was present. Chest and other systemic examinations were normal. On blood examination, total bilirubin was slightly raised (2.5 mg/dl). Amylase (2900 U/L) and lipase (1900 U/L) were significantly raised. Other blood investigations were normal.

A Magnetic resonance cholangiopancreatography (MRCP) was completed that showed the right posterior hepatic duct was draining into the left hepatic duct (LHD)-congenital type IIIA anomaly (Figure 1). The cystic duct was draining into the right hepatic duct in its medial wall with stone in the cystic duct (Figure 2). The right anterior hepatic duct continued as the right hepatic duct. A sigmoid course of the ventral and dorsal pancreatic duct with multiple stones within it was observed (Figure 3). An anomalous pancreaticobiliary junction (PBM) was discovered outside of the duodenal wall with a common channel of 2 cm - Komi classification type 2.

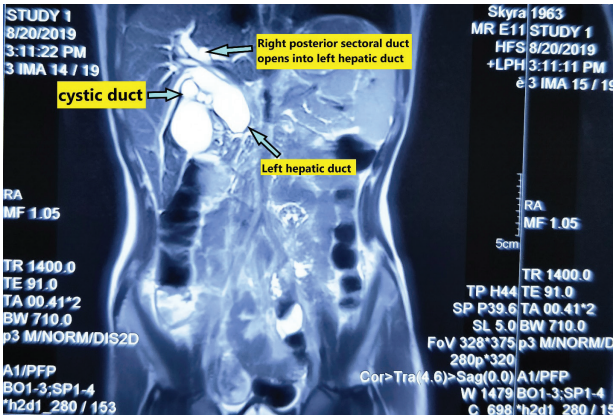

Figure 1) *MRI photograph showing hepatic duct anomaly type IIIa showing the right posterior sectoral duct opening into the left hepatic duct.*

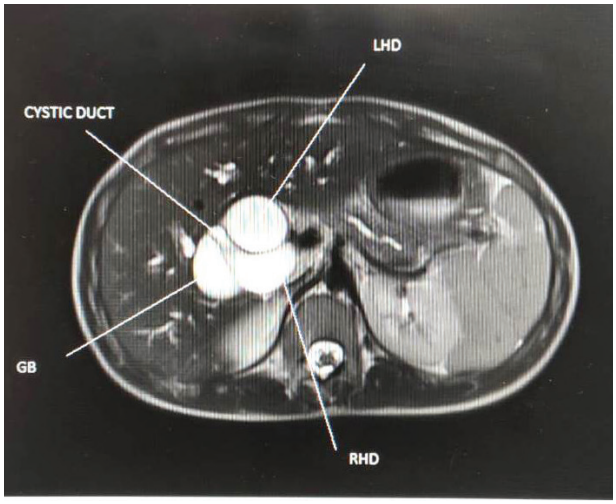

Figure 2) *MRI photograph showing the cystic duct draining into the right hepatic duct in its medial wall.*

LHD=Left Hepatic Duct; GB=Gall Bladder; RHD=Right Hepatic Duct.

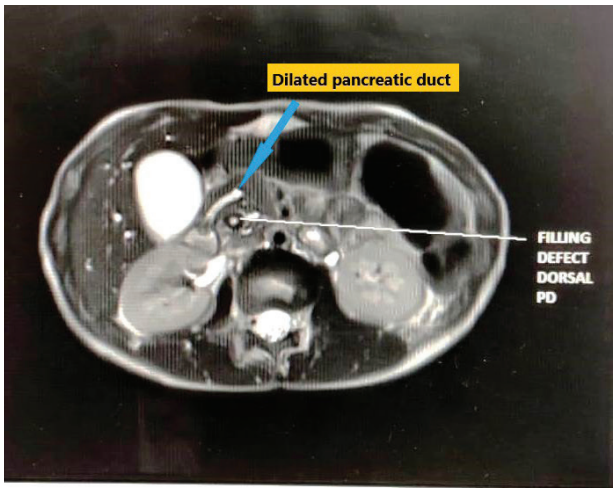

Figure 3) *MRI photograph showing dilated sigmoid course of main pancreatic duct.*

PD=Pancreatic Duct.

As per these findings, the diagnosis of recurrent acute pancreatitis was made, and the patient was managed conservatively. Later, the patient was discharged with the plan of excision of choledochal cyst with hepaticojejunostomy with lateral pancreatojejunostomy.

Discussion

PBM or maljunction is a well-known but rare condition, and often less known cause of acute pancreatitis, especially in young patients. It may also a predisposing cause of biliary tract or gall bladder cancer. In one study, anomalous ductal union was found in 16.7% of the patients with gallbladder cancer in comparison with an incidence of 2.8% with various other hepatobiliary and pancreatic conditions, who did not have gallbladder carcinoma [7]. The frequency of PBM varies from 1.5-3.2% [7]. Diagnosis of this condition can only be made by a high index of suspicion to prevent further complications. Komi et al. described a new classification for it after studying 51 patients of PBM. According to them, 35.5% were type I, 21.6% were Type II, and 43.1% were type III [8] (Figure 4).

KOMI CLASSIFICATION

TYPES		DESCRIPTION
IA	Type I A	Have single papilla & The Common Hepatic and pancreatic ducts join each other at a right angle with a non dilated common channel.
IB	B	Have single papilla & The Common Hepatic and pancreatic ducts join each other at a right angle with a dilated common channel.
IIA	Type II A	Have single papilla & The Common Hepatic and pancreatic ducts join each other at An acute angle with a non dilated common channel.
IIB	B	Have single papilla & The Common Hepatic and pancreatic ducts join each other at An acute angle with a dilated common channel.
IIIA	Type III A	Have two papilla & are equivalent to the classic pancreas divisum with biliary dilatation.
IIIB	B	Have two papilla & are characterized by the absence of the Wirsung's duct.
IIIC1	C1	Have two papilla & contain a tiny communicating duct between the main duct and the accessory ducts.
IIIC2	C2	Have two papilla with & characterized by a common channel made up of common and accessory ducts of equal caliber.
IIIC3	C3	Have two papilla with intricate network of dilated ducts that join each other by total or partial dilatation of the ductal system.

Figure 4) *Komi classification showing types of pancreatobiliary junction anomaly.*

The intrahepatic bile ducts course follows the portal veins along their anterior aspect. The right hepatic duct has its anterior segmental branch, which drains into the Couinaud's segments 5 and 8 of the liver. Also, it has a posterior segmental branch, which drains into the segment 6 and 7 of the liver. The right anterior sectoral duct (RASD) is oriented vertically, while the right posterior sectoral duct (RPSD) is oriented more horizontally. Normally, RPSD passes posterior to RASD and then joins it from its medial aspect. The left hepatic duct drains the segments 2,3, and 4 of the liver. The bile duct from the caudate lobe usually joins the origin of the left and right hepatic ducts. The right and left hepatic ducts join in the hilum to form the common hepatic duct (CHD) [9]. This typical branching pattern is seen in 50-60% of the population, with a significant number of subjects showing variation in the branching pattern. Variation in the sectoral hepatic duct is shown in below-mentioned figure [10] (Figure 5).

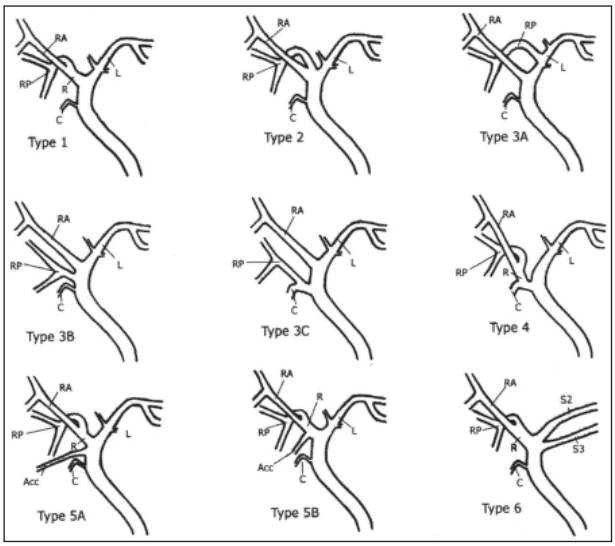

Figure 5) *Variation in the distribution of hepatic sectoral duct [10].*

MRCP is most informative and non-invasive imaging technique for the demonstration of biliary tract anatomy and anomaly of bile duct. It is the most accurate and safer modality to describe biliary anatomy. It is devoid of ionizing radiation, which allows it to be performed on patients that are allergic to iodinated contrast agents. Endoscopic retrograde cholangiopancreatography (ERCP) with cholangiogram and percutaneous transhepatic cholangiography are other investigations that can delineate the biliary tree but are invasive. Treatment varies depending upon patient signs and symptoms. Initially, conservative management should be done to relieve symptoms of acute pancreatitis. For definitive treatment, excision of choledochal cyst with hepaticojejunostomy with or without pancreatojejunostomy is required for proper drainage.

Conclusion

PBM is a rare condition that may be associated with recurrent attacks of acute pancreatitis. It should be considered as a differential diagnosis in young patients without a history of alcoholism and biliary stone disease. In our case, PBM is also associated with hepatic duct anomaly with the pancreatic and biliary stone formation, which is a rare entity. Only a few cases described in the literature to date with these findings. MRCP is the best non-invasive tool to diagnose this condition. Initial treatment is conservative to relieve symptoms of pancreatitis. ERCP and biliary stenting with or without pancreatic stenting can also be done if symptoms are not relieved by conservative management. Definitive surgery can be done once the patient is stable in the form of excision of choledochal cyst with drainage procedure as there is a high chance of malignancy later.

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