

Total Excision of The Extrahepatic Bile Duct For A Complex Pancreatico-Biliary Maljunction With Pancreas Divisum: Case Report

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1. Abstract

1.1. Introduction: Congenital pancreaticobiliary malformations are rare and their association is exceptional. It defines the type III of KOMI classification. The standard treatment is surgery. There isn't yet a standard surgical procedure.

1.2. Case presentation: We report a case of a 42-year-old-women with a complex type of pancreaticobiliary maljunction, type III of KOMI, successfully treated with a total bile duct resection with a Roux-en-Y hepatico-jejunostomy. Actually, 18 months post-operatively, she is symptom-free.

1.3. Conclusion: The coexistence of pancreaticobiliary maljunction and pancreas divisum is exceptional, it is defined as type III of KOMI classification. The major risk is the transformation in cholangiocarcinoma. MRCP is the best imaging modality of diagnosis and classification of the type of malformation. Surgery is the standard treatment of PBM and it is recommended once the diagnosis of PBM is established, Surgical treatment.

2. Introduction

Congenital malformations such as pancreaticobiliary maljunction, congenital dilatation of the bile duct, and pancreas divisum are rare. Their association is exceptional and is defined as type III in KOMI classification. Its genesis is unclear. The standard treatment is surgery but there isn't yet a standard surgical procedure.

Herein, we report a case of a 42-year-old-women with a complex

type of pancreaticobiliary maljunction, type III of KOMI, successfully treated with a total bile duct resection with a Roux-en-Y hepatico-jejunostomy.

3. Case Presentation

A 42-year-old woman was admitted for upper right quadrant abdominal pain without other symptoms for the last two years. Her past medical history includes a splenectomy for hereditary spherocytosis with cholecystectomy at the age of 13, and an emergency hysterectomy for postpartum hemorrhage at the age of 28. On admission, physical examination revealed no abnormalities especially no yellowish skin. Laboratory tests revealed normal levels of total bilirubin 16 µmol/L, direct bilirubin 4 µmol/L, alanine transaminase (ALT) 18 IU/L, Aspartate Aminotransferase (AST) 10 U/L, γ-glutamyltransferase (14 IU/L), and cancer antigen 19-9 (CA 19-9) 8.76 U/mL. Abdominal CT scan showed a congenital cystic lesion Todani type Ic. MRCP revealed a diffuse fusiform dilatation of the common bile duct compatible with congenital cystic lesion Todani type Ic, a bilio-pancreatic maljunction associated with a pancreas divisum consistent with type IIIA of komi classification (figure 1). The patient underwent resection of the principal biliary duct and hepaticojejunal anastomosis (figure 2). The pathological examination revealed a chronic inflammatory process without no malignant transformations. The immediate postoperative course was uneventful. Actually, 18 months post-operatively, she is symptom-free.

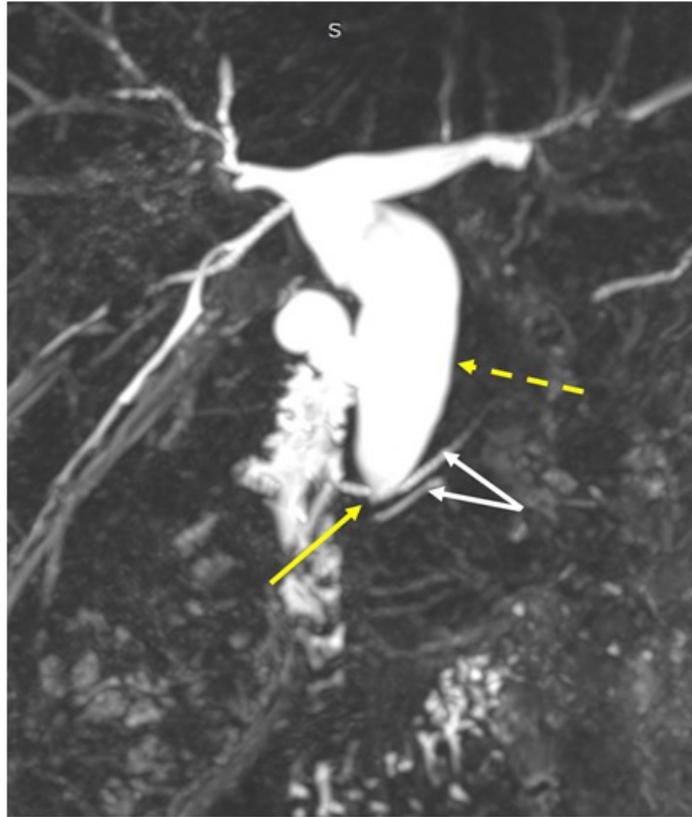


Figure 1 : MRCP showing a long common channel of pancreaticobiliary maljunction (yellow arrow), cystic dilatation of the extrahepatic bile duct (discontinuous yellow arrow), and the pancreatic ducts (white arrows)



Figure 2: Per operative aspect of the main bile duct dilatation

4. Discussion

Pancreaticobiliary maljunction is defined by a pancreatic and bile junction situated outside the duodenal wall. Several definition criteria have been proposed in literature. The most important one is the abnormal union between the bile and pancreatic ducts outside

the duodenal wall with a relatively long common channel [1], it varies from 8 to 15mm and is still not yet defined in literature [2]. In all cases, because of the long common channel, the sphincter Oddi is no longer able to control the pancreatico-biliary junction which allows regurgitation of pancreatic juice into the bile duct

and reciprocally. Pancreatico-biliary reflux is more frequent because hydro pressure in the pancreatic main duct is higher [3].

Pancreaticobiliary maljunction is usually associated with a common bile duct dilatation [6], which is determined by the majority of authors of more than 10mm of diameter among adult patients [7]. This association may be explained by abnormal embryology of the pancreatico-biliary system, given the fact that the common bile duct and the ventral pancreas derive from the ventral ventriculum both [1,4,8]. However, there are many cases of pancreaticobiliary maljunction without common bile duct dilatation [9].

Pancreas divisum is another congenital malformation resulting from a fusion failure between the two pancreatic ducts. The coexistence of pancreaticobiliary maljunction and pancreas divisum is very rare, and its incidence is 1,4% in pediatric cases of pancreaticobiliary maljunction [10]. This association is defined as type III of KOMI classification. To the best of our knowledge, there are only a few cases of KOMI III type reported in the literature.

The main symptoms are abdominal pain, jaundice, fever, and vomiting [1]. Those symptoms are more frequent in children and in congenital bile duct dilatation form [1]. Often they are caused by protein plugs compacted in the common channel [1]. The major risk of pancreaticobiliary maljunction is carcinogenesis due to chronic inflammation. Indeed, the mixture of pancreatic juice with bile induced ectopic activation of pancreatic enzymes, bile acid fraction modification, and mutagenic substances yield [11]. These aggressive substances may lead to chronic inflammation and DNA damage such as oncogenes (K-ras) and tumor suppressor genes (p-53) mutations [11], and carcinogenesis follows the hyperplasia, dysplasia then carcinoma sequence [3,9,11]. This carcinogenesis is directly related to bile stagnation, gallbladder carcinoma is more frequent in cases without biliary dilatation, and common bile duct carcinoma is more frequent in the dilated ones [9]. For some authors, pancreas divisum cases with dorsal pancreatic duct dominance are less frequent to have biliary carcinoma because most pancreatic juice is drained through the minor papilla into the duodenum [6,9].

Pancreatic carcinoma is less frequent than bile duct carcinomas, but its incidence is higher in pancreaticobiliary maljunction compared to the general population [11].

Radiological imaging is important for diagnosis by visualization of an abnormal long common channel with a pancreaticobiliary union outside the duodenal wall [1]. To date, Magnetic Resonance Cholangio-Pancreatography (MRCP) is the non-invasive tool of choice for the diagnosis approach [4,12]. Its detection rate for pancreaticobiliary maljunction is 82-100% in adults [1]. However, its ability is restricted by its limited spatial resolution especially in complicated form of junction and fine pancreatic duct [1]. Endoscopic Retrograde Cholangio-Pancreatography (ERCP) remains a gold standard for diagnosis [12]. It can depict pancreatic duct and

biliary duct anomalies and can be a useful therapeutic option to relieve biliary obstruction [1,12]. But, ERCP is avoided in typical cases because of the risk of post-procedural pancreatitis [9].

The standard treatment of pancreaticobiliary maljunction is surgery and is considered as soon as the diagnosis is made because of the risk of cancer [5,11]. The process of carcinogenesis may be initiated early, and hyperplasia lesions can be present from the early stages of infancy [3]. The membrane of all the biliary tract is considered a "precancerous lesion"; that's why extended or total excision of the extra hepatic biliary tract is necessary with a Roux-en-Y hepatico-jejunostomy [11]. Total resection of the retro pancreatic bile should be performed and the main bile duct should be dissected at the level just above the pancreaticobiliary junction; in order to avoid the residual mucosa [1,8]. In order to prevent per-operative main pancreatic duct injuries, some authors propose a subtotal excision, in which 5-10mm wall of the bile duct is left in intrapancreatic space, and only the mucosa will be removed [13]. In non-dilated cases, the surgical procedure still controversial, cholecystectomy without bile duct resection can be performed [11,13,14]. Some authors propose a cephalic-duodeno-pancreatectomy for KOMI-III types [15]. Our patient is a KOMI-III and underwent a total bile duct resection and cholecystectomy with an uneventful post-operative course.

5. Conclusion

The coexistence of pancreaticobiliary maljunction and pancreas divisum is exceptional, it is defined as type III of KOMI classification. The major risk is the transformation in cholangiocarcinoma. MRCP is the best imaging modality of diagnosis and classification of the type of malformation. Surgery is the standard treatment of PBM and it is recommended once the diagnosis of PBM is established. Surgical treatment, mostly based on a resection of the extrahepatic bile duct and Cholecystectomy.

References:

1. Kamisawa T, Kaneko K, Itoi T, Ando H. Pancreaticobiliary maljunction and congenital biliary dilatation. *Lancet Gastroenterol Hepatol.* 2017; 2: 610-18.
2. Wang C-L, Ding H-Y, Dai Y, Xie T-T, Li Y-B, Cheng L, et al. Magnetic resonance cholangiopancreatography study of pancreaticobiliary maljunction and pancreaticobiliary diseases. *World J Gastroenterol WJG.* 2014; 20.
3. Kamisawa T, Kuruma S, Chiba K, Tabata T, Koizumi S, Kikuyama M. Biliary carcinogenesis in pancreaticobiliary maljunction. *J Gastroenterol.* 2017; 52: 158-63.
4. Le Roy B, Gagniere J, Filaire L, Fontarensky M, Hordonneau C, Buc E. Pancreaticobiliary maljunction and choledochal cysts: from embryogenesis to therapeutics aspects. *Surg Radiol Anat.* 2016; 38: 1053-60.
5. Kamisawa T, Ando H, Suyama M, Shimada M, Morine Y, Shimada H, et al. Japanese clinical practice guidelines for pancreaticobiliary

- maljunction. *J Gastroenterol*. 2012; 47: 731-59.
6. Petrasek J, Huel T, Spicak J. Pancreaticobiliary malunion and incomplete pancreas divisum: an unusual cause of common bile duct obstruction. *Adv Med Sci Gruyter Open*. 2008; 53: 6-10.
 7. Tashiro S, Imaizumi T, Ohkawa H, Okada A, Katoh T, Kawaharada Y, et al. Pancreaticobiliary maljunction: retrospective and nationwide survey in Japan. *J Hepatobiliary Pancreat Surg*. 2003; 10: 345-51.
 8. Sanada Y, Sakuma Y, Sata N. Embryological etiology of pancreaticobiliary system predicted from pancreaticobiliary maljunction with incomplete pancreatic divisum: a case report. *BMC Surg*. 2018; 18: 50.
 9. Yoshimoto K, Kamisawa T, Kikuyama M, Kuruma S, Chiba K, Igarashi Y. Classification of pancreaticobiliary maljunction and its clinical features in adults. *J Hepato-Biliary-Pancreat Sci*. 2019; 26: 541-7.
 10. Terui K, Hishiki T, Saito T, Sato Y, Takenouchi A, Saito E, et al. Pancreas divisum in pancreaticobiliary maljunction in children. *Pediatr Surg Int*. 2010; 26: 419-22.
 11. Funabiki T, Matsubara T, Miyakawa S, Ishihara S. Pancreaticobiliary maljunction and carcinogenesis to biliary and pancreatic malignancy. *Langenbecks Arch Surg*. 2009; 394:159-69.
 12. Cui G-X, Huang H-T, Yang J-F, Zhang X-F. Rare variant of pancreaticobiliary maljunction associated with pancreas divisum in a child diagnosed and treated by endoscopic retrograde cholangiopancreatography: A case report. *World J Clin Cases*. 2019; 7: 1073-79.
 13. Kimura W. Congenital dilatation of the common bile duct and pancreaticobiliary maljunction-clinical implications. *Langenbecks Arch Surg*. 2009; 394: 209-13.
 14. Kobayashi S, Ohnuma N, Yoshida H, Ohtsuka Y, Terui K, Asano T, et al. Preferable operative age of choledochal dilation types to prevent patients with pancreaticobiliary maljunction from developing biliary tract carcinogenesis. *Surgery*. 2006; 139: 33-8.
 15. Al-Jiffry BO, Khurshid A, Khayat SH, Chundrigar T. Anomalous Pancreaticobiliary Junction Komi Type IIIC3, Rare Cause of Recurrent Acute Pancreatitis; Case Report. *Gastroenterol Pancreatol Liver Disord*. 2014; 1: 1-3.