Choledochal cyst with Anamolous Pancreato Biliary Junction (APBJ) - Case report

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ABSTRACT

Anamalous union of Pancrato Biliary duct (APBD) has been regarded to be the etiological factor for choledochal cyst. A 27 year old lady who presented with pain abdomen and vomiting was found to have Type IV A choledochal cyst with APBD which was diagnosed by Magnetic Resonance cholangio Pancreatography (MRCP). She was successfully treated by excision of the cyst with Roux-en Y hepatico-jejunostomy. Classification of Choledochal cysts and APBJ, possible diagnostic modalities with treatment were discussed.

Keywords: Choledochal Cyst, APBJ, Pancreatic duct anomalies, Hepatico Jejunostomy

Introduction:

Choledochal cyst is a congenital dilatation of the bile duct. Anomalous union of pancreaticobiliary duct (AUPBD) has been regarded to be the etiological factor of the choledochal cyst. Approximately 20% of cysts are diagnosed in older patients. Most of the type I cysts are complicated by APBJ. The incidence of biliary tract cancer in patients with choledochal cysts was reported as 6% - 20% in the United States and approximately 15% - 20% in Japan.

Case report:

A 27 year old lady was admitted with complaints of epigastric pain since 2 months and non bilious vomitings at the onset of pain, abdominal examination was normal, her bilirubin was normal, ALP -1070U/L, MRCP (figure 1) showed type IV A choledochal cyst with common pancreaticobiliary channel outside the duodenal lumen measuring <15mm with two pancreatic ducts draining at an acute angle into common bile duct (CBD). Cyst excision with roux en y hepaticojejunostomy (figure 2) was done and patient recovered well. Patient was followed for 6 months and she is symptom free.

Figure 1A
CLASSIFICATION OF CHOLEDOCHAL CYSTS AND ANOMALOUS PANCREATICOBILIARY JUNCTION

Choledochal Cysts

Todani et al.'s classification of choledochal cysts is as Type I cysts consist of saccular or fusiform dilatation of the extrahepatic bile duct. This is the most common type and represents nearly 80% - 90% of cases. Type I cysts are further subdivided into type IA (diffuse) cysts, type IB (focal) cysts, and type IC (fusiform) cysts. Subtype IA shows saccular dilatation of the common bile duct. Subtype IB shows focal, segmental dilatation of the common bile duct. Subtype IC has fusiform dilatation of the common hepatic and common bile duct [1,2]. Type II cysts are a diverticulum of the CBD. Type III cysts are also referred to as choledochoceles, which show dilatation of the intra duodenal portion of the CBD. Type IV cysts show dilatation of either the intra- or extra hepatic bile duct, or both. Type IV cysts can be subdivided into two subtypes: IV-A involving multiple intra- and extra hepatic cysts, and IV-B, involving only multiple extra hepatic biliary dilatations. Type V cysts (Caroli's disease) present with segmental cystic dilatations of the intra-hepatic bile duct. (3,4)

APBJ (Anomalous union of Pancreatico-Biliary Junction)

APBJ refers to union of the pancreatic and bile ducts outside the duodenal wall resulting in a long common channel (usually >15 mm) (1). ABPJ is seen in up to 90% of cases of congenital choledochal cysts and is associated with increased risk of pancreatitis and cholangio carcinoma presumably secondary to biliopancreatic reflux (5).

In 1977, Kimura et al. classified APBJ into two types by analyzing the fusion pattern between the pancreatic and bile ducts. In the P-C type, the main pancreatic duct appears to join the common bile duct, while the common bile duct appears to
join the main pancreatic duct in the C-P type [6].

In 1994, the Japanese Study Group of Pancreaticobiliary Maljunction (JSPBM) classified APBJ into three sub- groups, such as type a (right angle type), type b (acute angle type), and type c (complex type) according to the type of confluence of the main pancreatic duct and the CBD. Type a and type b correspond to the P-C type and C-P type, respectively. In 2003, Tashiro et al. analyzed 1627 patients who were enrolled by the JSPBM, including 1239 patients with choledochal cysts and 388 patients with the non-dilated type of APBJ. APBJ was type a in 57.9%, type b in 32.4%, and type c in 5.6% of cases with choledochal cysts. APBJ was type b in 60.8%, type a in 29.4%, and type c in 7.2% cases with the non-dilated type of APBJ [7].

Endoscopic retrograde cholangiopancreatography (ERCP) is the gold standard for diagnosis of APBJ. MRCP is able to diagnose choledochal cysts with an accuracy of 82% - 100% [8]. MRCP is considered as the first-choice modality for diagnosing choledochal cysts and APBJ in pediatric patients because it is non-invasive and it does not require breath holding.

Currently, complete excision of a cyst with cholecystectomy and Roux-en-Y hepatico-jejunostomy reconstruction (RYHJ) is the standard therapy in Types I and IV-A cysts [9].

**Conclusion**

APBJ frequently associated with choledochal cyst may have an implication not only as an etiological factor but as an associated disorder leading to a grave clinical course. APBJ associated with choledochal cyst may be a very important factor that affects the clinical course, surgical planning and prognosis.

**References:**


