

Anomalous Pancreaticobiliary Junction Komi Type IIIC3, Rare Cause of Recurrent Acute Pancreatitis; Case Report

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Abstract

Some types of Anomalous Union of the Pancreaticobiliary Duct (AUPBD) can cause recurrent acute pancreatitis and can pose a diagnostic challenge. We describe the case of a 19 year old girl with type IIIC3 malformation and recurrent acute pancreatitis. The discussion provides a review of recent literature, which supports the use of several modes of investigation and surgery as a choice of treatment.

Keywords: Anomalous Union of the Pancreaticobiliary Duct (AUPBD); Recurrent Acute pancreatitis; Komi classification; Choledochal cyst

Introduction

Anomalous Union of the Pancreaticobiliary Duct (AUPBD) is a rare condition that can pose a diagnostic challenge and some types can cause recurrent acute pancreatitis. Normally, the main pancreatic duct and the common bile duct open into the second part of the duodenum alone or after joining as a common channel. The length of the common channel ranges from 1-12mm normally, with a mean of about 4-5 mm. The anomalies are complex and are described below in the discussion.

The frequency of AUPBD in ERCP in some series varies from 1.5-3.2% [1]. Its diagnosis needs a high index of suspicion and carefully performed investigations.

We describe one such case of a type IIIC3 malformation that presented to our hospital recently.

Case Report

A 19 years old Saudi girl presented and admitted with three recent episodes of *mild* acute biliary pancreatitis. She had history of repeated episodes of abdominal pain since childhood. At the age of three, she was diagnosed to have acute pancreatitis and a pancreatic congenital anomaly was suggested, even with several visits to hospitals over several years. Ultrasound showed dilated intra- and extra-hepatic ducts with the common bile duct (CBD) measuring about 2.5cm. MRCP revealed dilated extra-hepatic duct down to the lower end of CBD. At ERCP both the major and minor papilla were separately cannulated. Injection of either

papilla demonstrated the same complex network of dilated ducts, with these ducts eventually communicating with each other and to the common bile duct (CBD) and pancreatic duct. The lower end of the CBD, at its junction with a c shaped loop, was markedly narrowed, with marked dilatation proximally, a choledochal cyst shown more proximally (see Figure 1). After reviewing the literature and comparing the pictures we found that the pattern consistent with the type IIIC3 anomaly. This, according to Komi et al. Classification, has two papilla both communicating with a complex network of dilated ducts. Absence of dilatation would make it Type IIIC2. The treatment of choice for this subtype is treated by pancreatododenectomy. She underwent pancreatododenectomy after much discussion with the family. The immediate post operative course was complicated by wound infection, which responded to appropriate treatment in the form of drainage and antibiotics. She is currently symptom free. Her last visit was in November, 2011 (30 month post surgery).

Discussion

Anomalous Union of the Pancreaticobiliary Duct (AUPBD) is a rare condition. Normally, the main pancreatic duct and the common bile duct open into the second part of the duodenum alone or after joining as a common channel. The length of the common channel ranges from 1-12mm normally, with a mean of about 4-5 mm. In infants, a common channel longer than 4mm is considered abnormal, in adults it is considered abnormal if longer than 6mm [2].

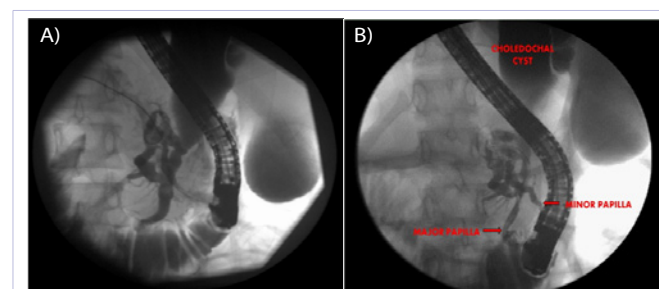


Figure 1: A) The complex dilated net-work involving the pancreatic ducts with the catheter in the major duct as seen by the arrow going into the pancreas. B) The catheter in the minor duct indicated by the arrow.

KOMI CLASSIFICATION

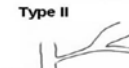
TYPES		DESCRIPTION
IA	Type I	Have single papilla & The Common Hepatic and pancreatic ducts join each other at a right angle with a non dilated common channel.
	A 	
IB	Type I	Have single papilla & The Common Hepatic and pancreatic ducts join each other at a right angle with a dilated common channel.
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

Table 1: Demonstrating a detailed description of each type.

If it appears that the pancreatic duct is joining the common bile duct it is denoted as P-B type and if the common bile duct appears to join the main pancreatic duct it is denoted as B-P type [3].

In AUPBD the connection between the common bile duct and the main pancreatic duct is located outside the duodenal wall [3].

Choledochal cyst is a rare congenital dilatation of the CBD that is known to be associated with Anomalous Union of the Pancreaticobiliary Duct (AUPBD) [4].

AUPBD is a well described though uncommon, and often unrecognized, cause of acute pancreatitis, especially in young patients. It has also been associated with gallbladder carcinoma. In one study, anomalous ductal union occurred in **16.7%** of the patients with gallbladder carcinoma in comparison with an incidence of **2.8%** among **641** consecutive patients with various hepatobiliary and pancreatic diseases studied by endoscopic retrograde cholangio-pancreatography who did not have gallbladder carcinoma. Gallbladder carcinoma occurred in **24.6%** of the **65** cases of anomalous ductal union in comparison with a 1.9% incidence of this cancer among **635** consecutive patients similarly studied and found to have normal ductal union [1].

The frequency of AUPBD varies from 1.5-3.2% [1]. Its diagnosis needs high index of suspicion and carefully performed investigations. Table 1 Komi et al. in their new classification described 51 cases of AUPBD. Of these, 35.5% were Type I, 21.6% were Type II and 43.1% were type III. Type III was sub-divided into three types (A,B and C). Type C is further divided into three sub-types (1,2, and 3) [5]. Because of its rareness there are only few reported cases in the literature, mostly from asia [6].

ERCP is the diagnostic procedure of choice [2,3]. Our patient had two papilla, communicating with each other and with dilatation of the ducts which makes it type IIIC3. Only type IB, IIB and IIIC3 are associated with recurrent pancreatitis and are treated surgically. The treatment of choice for type IIIC3 is pancreatododenectomy [4]. Out of Japan the two reported cases of type III are type IIIa of all AUPBD types [7]. We could only find one case of type IIIC3 in the English literature[8].

Komi et al. [5] didn't mention the numbers of the each subtype of type III C. Our case therefore is the second reported case of type IIIC3 in the literature.

Conclusion

In young patients, AUPBD should be considered as a cause

of recurrent acute pancreatitis [1]. The complications and long term sequelae of acute pancreatitis and risk of cancer in young patients mandates appropriate management [1,4].

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