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Case Report

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Congenital Absence of the Gall Bladder Associated with Forme-Fruste Choledochal Cyst: A Rare Case Report

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Abstract

An 11-year old girl presented with episodes of upper abdominal pain, fever, and non-bilious vomiting. Abdominal ultrasound (US) revealed a dilated common bile duct with non-visualization of the gallbladder. A magnetic resonance cholangiopancreatogram (MRCP) confirmed absence of the gallbladder and cystic duct, a 'forme fruste' choledocal cyst (FFCC) and no evidence of pancreatobiliary malunion (PBMU). Surgical excision of the dilated extrahepatic bile duct (EHBD) and a Roux-en-Y hepaticodocho-jejunostomy was successful. The discussion centers on the extremely rare association of congenital absence of the gallbladder (CAGB) with FFCC.

Keywords: Agenesis Gallbladder, Forme Fruste Choledochal Cyst, Pancreaticobiliary Malunion, Magnetic Resonance Cholangiopancreatogram.

Introduction

Congenital absence of the gallbladder (CAGB) is a rare finding with a reported incidence of 0.01 to 0.075 % [1, 2]. According to Liu et al CAGB is more common in females (f:m, 3: 1) [3]. More than 50 % patients with CAGB are symptomatic, usually with symptoms suggestive of cholecystitis, and require surgical intervention [3, 4].

'Forme fruste' choledochal cyst (FFCC) is a choledochal cyst that has minimal or no dilatation of the extrahepatic bile duct (EHBD) and is associated with pancreatobiliary malunion (PBMU), the maximum diameter of the common bile duct being less than 10 mm [5]. Most patients present with symptoms suggestive of classic choledochal cyst [6, 7]. This report describes a rare association of FFCC in an 11-year old girl with ageneis of the gall bladder and cystic duct. The patient was successfully managed by excision of the dilated common duct and a Roux-en-Y hepaticodocho-jejunostomy.

Case Report

An 11-year old girl presented with a history of recurrent episodes of mild to moderate upper abdominal pain for the past 4 months. The episodes of abdominal pain were accompanied by low-grade fever (< 101 degrees F) and non-bilious vomiting. Clinical examination was normal. Routine hematological and biochemical investigations, including liver function tests and 2-D echo were normal.

An abdominal ultrasound (US) showed fusiform dilatation of the common bile duct (CBD) with a maximum diameter of 9.2 mm. The gall bladder was not identified. A magnetic resonance cholangiopancreatogram (MRCP) showed a Type 1c choledochal cyst (CC) with maximum diameter 10 mm suggestive of 'forme-fruste' choledochal cyst. The gall bladder and cystic duct were absent and the intrahepatic bile ducts were normal (Figure 1). There was no evidence of pancreato-biliary malunion (PBMU).

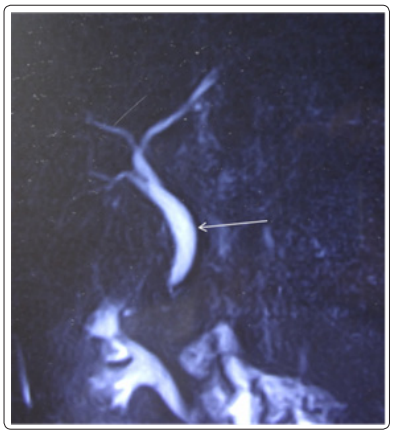

Figure1: Magnetic resonance cholangiopancreatogram (MRCP) showing 'forme fruste' choledochal cyst (arrow) with absence of the gallbladder and cystic duct.

At laparotomy, a Type 1 CC was found with absence of the gall bladder and cystic duct (Figure 2). The site of the gall bladder fossa was marked by a white line of fascia. The CC was dissected and excised after suturing its distal end with 4-0 Vicryl (polydiaxone). A Roux-en-Y hepaticodocho-jejunostomy was performed. The postoperative period was uneventful. Histological examination of the resected cyst showed features of a choledochal cyst with inflammatory changes in the mucosa and submucosa.

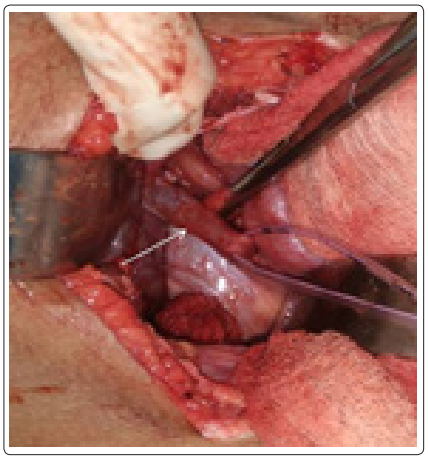

Figure 2: Photograph taken during surgery showing the dilated common duct and absence of the gallbladder and cystic duct (arrow).

Discussion

According to Liu et al, CAGB is caused by agenesis of the tail or caudal branch of the liver diverticulum during the 4th week of the embryonic period [3]. Bennion et al classified this condition into 3 subtypes, based on the clinical features: Type 1, associated with multiple congenital anomalies (12.8 %); Type 2, asymptomatic (31.6 %); and Type 3, symptomatic (55.6 %). Tang et al suggested a modified classification in which Type 1a CAGB included cases associated with potentially fatal malformations while Type 1b consisted of cases associated with non-lethal anomalies such as malrotation, right liver agenesis, cryptorchidism, choledochal cyst, choledochoectasia [2, 8]. The term Type 2 CAGB should be reserved for asymptomatic cases [8]. The vast majority of cases (55.6 %) are of Type 3 CAGB which are often mistaken for cholecystitis in adults as they present with biliary symptoms such as right upper-quadrant pain, nausea and vomiting, lack of appetite, jaundice, and fever [8].

In cases of FFCC, the associated PBMU is believed to be responsible for presentation with symptoms similar to those of choledochal cysts [6, 7]. Shimotakahara et al reported 17 cases of FFCC out of 281 cases of choledochal cyst managed at their center [5]. Surgery with excision of the EHBD and a Roux-en-Y hepaticodocho-jejunostomy was the treatment of choice. Thomas et al reported 6 children, all with symptoms of classic choledochal cyst, who had FFCC with further imaging revealing a long pancreatobiliary channel [7]. There was a high success rate of surgery provided a pancreatobiliary disconnection was incorporated in the operative management [7]. The imaging of choice in pediatric cases remains a MRCP scan which shows the diameter of the EHBD and provides evidence of PBMU [6, 7].

The association of these two anomalies is rare and there are no similar reports in the literature from India. However, Tang et al reviewed 77 cases of CAGB, including 75 cases from a Chinese registry and 2 of their own and found that 3 had lower common bile duct narrowing, 3 had choledochoectasia, 1 had a lower common bile duct valve, and 1 had situs inversus viscerum [8]. It can be presumed that choledochoectasia refers to a condition similar to FFCC. In our patient, MRCP was conclusively diagnostic of both FFCC and CAGB, although PBMU was not detected. Considering the association of Type 1b CAGB with biliary ductal disorders, it can be recommended that an MRCP should be performed with careful evaluation of common duct diameter in patients diagnosed with CAGB on ultrasonography [8]. Treatment with excision of the EHBD and hepaticodocho-jejunostomy, as in our case, is straightforward with good results [7].

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