

Atypical Presentation of Choledochal Cyst Type IV-A in a Hispanic Girl: A case report

Victor Ortiz Justiniano*

University of Puerto Rico School of Medicine, USA

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***Corresponding author:** Victor Ortiz Justiniano, University of Puerto Rico School of Medicine, USATel: 17874000054, Email: zashavazquez@gmail.com

Abstract

A type IV-A choledochal cyst is the second most common type of this rare congenital entity that is characterized by multiple dilatations of the intrahepatic and extrahepatic biliary tract. The most common clinical findings of this entity is abdominal pain, palpable right upper quadrant mass and jaundice. However, there are other clinical presentations that differ from the classical presentation that can delay the adequate management of the choledochal cyst. The optimal treatment is total surgical excision along with a hepaticoduodenostomy or hepaticojejunostomy. We present a case of a 22 month old female with an unusual presentation of a choledochal cyst.

Introduction

Choledochal cysts are rare congenital entities characterized by single or multiple dilatations of the intra and/or extrahepatic biliary tree [1]. Its incidence in Asian population is 1:1000, but in western populations is 1:100000 to 1:150000 [2]. There is an unexplained female:male preponderance of 1.5:1 in the pediatric population and up to 4.9:1 in the adult population [1,3]. There are numerous theories of choledochal cyst etiology. The most accepted theory explaining the pathogenesis of choledochal cyst is the Babbitts theory. This theory is based in the anomalous pancreaticobiliary ductal junction system. It attributes choledochal cyst formation to the presence of an abnormal pancreaticobiliary junction outside the ampulla of Vater, resulting in a long common channel that allows the reflux of pancreatic juice in the bile duct. This reflux in turn activates pancreatic enzymes, causing inflammation and weakness of the bile duct wall and leading to cyst formation [4].

The anatomy of choledochal cyst is categorized in five types named by Todani. Choledochal cyst

- Type I** is a saccular or fusiform dilatation of a portion or entire common bile duct with normal intrahepatic duct;
- Type II** consists of an isolated diverticulum protruding from the common bile duct;
- Type III** is a dilatation of intraduodenal segment of the common bile duct (choledecele);

d. **Type IV** consists of multiple dilatations of the intrahepatic and extrahepatic biliary tract (**IV-A**) or multiple dilatations involving the extrahepatic biliary tree (**IV-B**).

e. **Type V** consists of cystic dilatation of the intrahepatic biliary ducts (Caroli's disease).

Choledochal cyst **Type I** is the most common type presenting in 50-80% of cases. Choledochal cyst **Type IV-A** is the second most common one with an incidence of 5 to 83% in different series [1,5,6]. The clinical presentation of choledochal cyst varies among patients. The most common frequent presentation is abdominal pain (93.8%) followed by jaundice (58.3%). These cysts are typically a surgical problem of infancy or childhood, however, the diagnosis is delayed until adulthood in nearly 20% of patients [7]. Choledochal cyst carry a long - term burden of morbidity and mortality, therefore the optimal treatment is total surgical excision and possibly surgical diversion [8]. We present a case report of a 22 month old Hispanic girl with an atypical presentation and a rare case of choledochal cyst type IV-A.

Case Report

Case of a 22 month old female born premature at 32 weeks complicated with low birth weight.

She was doing well until 2 months ago that she started presenting generalized pruritus. She was treated with conservative management for skin lesions without significant improvement. Patient was brought to the emergency room due to worsening generalized pruritus and multiple skin lesions.

Upon physical examination she had multiple skin lesions and a distended, nontender abdomen with a palpated mass in the right upper quadrant. Mother referred that abdominal distention became evident close to the time she started presenting pruritus. Mother denied abdominal pain, nausea, vomiting, diarrhea, or jaundice. Laboratory evaluation upon admission was significant for elevated white blood cell count of 14.8×10^3 cells/ μL , elevated erythrocyte sedimentation rate of 71 mm/h, and elevated bilirubin levels at 2.26 mg/dL.

An abdominal ultrasound was performed and revealed a large cystic mass measuring 9.1 X 6.7 X 6 cm, suggestive of a choledochal cyst. A magnetic resonance cholangiopancreatography was ordered to confirm diagnosis and study revealed a marked dilatation of the intrahepatic and common hepatic ducts with severe fusiform dilatation of the common bile duct. These findings were consistent with a Type IV-A choledochal cyst.

Patient was started on ursodiol treatment as well as intravenous antibiotics (ceftriaxone and clindamycin) for concomitant skin infections. Also, she was started on topical treatment with hydrocortisone 1% cream for suspected dermatitis. Patient was taken to the operating room for excision of choledochal cyst and anastomosis. A hepaticoduodenostomy and division of an intrahepatic membrane were performed successfully. Histological sections of the cyst surgical margins did not reveal malignancy. The postoperative course was uneventful and the patient was discharged in the postoperative day # 11, tolerating diet and stooling adequately for age, as well as significant improvement in her symptoms. During follow up examination she was doing well. Bilirubin levels had decreased to normal values and pruritus had improved.

Discussion

A choledochal cyst is a rare congenital dilatation of the biliary tract characterized by single or multiple dilatations of intra and/or extra hepatic biliary tree. The incidence of choledochal cyst was reported to be less than 1 in 13,000 to 2 million population [9]. As previously mentioned, it has a female preponderance of 1.5:1. The choledochal cyst Type IV is the second most common choledochal cyst type. Clinical choledochal cyst presentations varies and depends largely on the age of the child at presentation [1]. Literature often reports that both pediatric and adult populations presents most commonly with abdominal pain, nausea and vomits, and jaundice, in that order of occurrence. However, the classic triad of abdominal pain, jaundice and palpable mass was found only in 10.5% of the children [2]. Hau [10] also reported that the classic triad of abdominal pain, jaundice and abdominal mass were not common in newborns or infants with choledochal cyst [10]. Pediatric cases frequently exhibited cystic lesions with jaundice and abdominal pain which were more apparent than in newborn and infants [10].

Todani reported that patients with Type IV-A are commonly older children and adolescents in comparison with those without intrahepatic involvement. Most likely due to development of

symptoms appears to be delayed until the intrahepatic cysts are filled with infected bile. Other presenting features of choledochal cysts are cholangitis, pancreatitis, and biliary peritonitis from cyst rupture [11,10]. Our patient presented with pruritus and without abdominal pain, which is an extremely rare presentation. Ultrasound is the initial imaging modality of choice for children, whereas ERCP (endoscopic retrograde cholangiopancreatography) is most commonly used in adults [9,12]. Surgical treatment of choledochal cyst should be recommended to reduce the risk of serious complications such as cholangitis, pancreatitis, rupture, portal hypertension, cirrhosis and cholangiocarcinoma [9].

The primary complete cyst excision and bilioenteric anastomosis with wide stoma is the treatment of choice for choledochal cysts and should be done as early as possible to reduce the risk of malignancy. Hepaticoduodenostomy or Roux-en-Y hepaticojejunostomy are the preferably approaches. Recently, laparoscopic approaches are being used more often with good results. Our patient had a primary complete cyst excision and a hepaticoduodenostomy with concomitant division of biliary ducts performed with success.

In conclusion choledochal cyst are rare congenital entities. They can present at different ages with variable symptoms. A high index of suspicion is necessary since if left untreated choledochal cysts have an increased rate of malignant transformation and serious complications.

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