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Choledochal Cyst with Cirrhosis and Portal Hypertension in A Child: A Rare Presentation

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Abstract

Choledochal Cysts (CDC) are rare congenital cystic dilation of the biliary tract. Usually, the CDC presents in the first decade of life, and its prognosis is excellent. There is no debate regarding managing a typical CDC with the usual presentation, while complicated CDC still has challenges. The persistent obstruction to bile flow can lead to changes in the liver; these changes are usually transient and improve after surgery. In a few cases, there are progressive changes in the liver, especially in type IVA cysts in which the intrahepatic portion is not dealt with or involves the liver so extensively that it is not feasible to excise it completely. We here discuss the management of a five-year-old female child with type IVA CDC presented with features of liver failure and portal hypertension, along with a review of the relevant literature.

Introduction

Choledochal Cysts (CDC) are rare congenital anomalies of the bile ducts, usually present with female preponderance in the first decade of life. The classical triad includes abdominal pain, jaundice, and lump. It is more frequently seen in children than adults, and 85% of children have at least two features of the triad at diagnosis, compared with only 25% of adults [1].

These are classified into five types according to Todani's classification, among which Type I is the most common, followed by Type IV. CDC Type I with back pressure effect may result in dilatation of the intrahepatic ducts and thus can mimic a type IVA cyst [2].

Type IVA choledochal cysts pose a challenge as they involve extrahepatic and intrahepatic biliary tree. The surgical intervention for these types of CDCs requires an individual-based approach. Surgical treatment of extrahepatic cysts is possible by excision of the cyst with bilio-enteric anastomosis. The need and type of surgery for the intrahepatic cyst are decided based on anatomy and the patient's clinical condition. The implications of residual intrahepatic cysts must be considered during follow-up [3]. Here, we present a case of type IV-A choledochal cyst that presented with signs of liver failure, which was managed by a multidisciplinary team.

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Case Presentation

A five-year-old girl presented with complaints of progressive yellowish discoloration of eyes, dark yellow urine, pale stools for two months, and dull aching abdominal pain for one week. There was no history of fever, vomiting, loss of appetite, or bleeding diathesis. On examination, icterus was present, and the abdomen was distended with visible dilated veins. On palpation, the liver was firm-to-hard and palpable up to 7 cm below the right costal margin, and a fluid thrill was present, suggesting ascites. On preliminary blood investigations, hemoglobin was 7.7 g/dl, TLC was 14,900/cu mm, and total bilirubin was 8.3mg/dl with a direct bilirubin of 4.7 mg/dl. Liver enzyme levels were elevated: Alkaline phosphatase and gamma-glutamyl transferase were 453 IU/L and 261 U/L, respectively. The coagulogram was deranged with an INR of 1.65 and a PTI of 65%. On imaging, ultrasound was suggestive of an enlarged liver with mildly heterogenous echotexture with dilated central and peripheral intrahepatic biliary radical dilation, distended gall bladder, dilated Common Bile Duct (CBD) of 4.8cm, enlarged spleen, and mild ascites. MRCP (Figure 1A, B) showed fusiform dilation of CBD measuring 54 mm, with dilated central and peripheral intrahepatic biliary radicles in both lobes of the liver suggestive of type IVA choledochal cyst with hepatosplenomegaly. Upper GI endoscopy showed esophageal varices and changes of portal gastropathy and enteropathy. A possibility of choledochal cyst type IVA with secondary biliary cirrhosis and portal hypertension was made. The patient was initially managed under the pediatric gastroenterology department and received blood, fresh frozen plasma, vitamin K, and antibiotics, followed by endoscopic variceal ligation.

After optimization, the patient was transferred to the pediatric surgery department for CDC excision and bilio-enteric anastomosis. The abdomen was opened through the right subcostal incision extending towards the left of the midline. The liver was hard and nodular (Figure 1C); approximately 100 ml of high-coloured ascitic fluid was present. A 6x6 cm large choledochal cyst (Figure 1D) involving the entire CBD and common hepatic duct. Multiple portal cavernomas were noted, the dissection was difficult because of significant bleeding from collaterals. The plain could not be created between the posterior wall of the cyst and the portal vein due to portal cavernoma. A Lilly's procedure was done, and the cyst was excised, with a roux-en-Y hepatico-jejunostomy done in a standard manner; at the same time, a liver biopsy was also taken.

Hemostasis was achieved, and a subhepatic drain was placed. Postoperatively, the patient had an uneventful course -she was allowed an oral diet on post-op day four and was passing cholic stools. However, the child had high drain output initially, then gradually, drain output decreased, and the drain was removed on postoperative day 14, and the patient was discharged. After 15 days of discharge, the patient was readmitted to the pediatric gastroenterology department for spontaneous bacterial peritonitis. She was managed with IV antibiotics and underwent paracentesis for the same. A repeat upper GI endoscopy and variceal ligation were done for esophageal varices. The patient responded to treatment and was discharged subsequently. At follow-up, the patient had mild ascites but was asymptomatic otherwise, and total bilirubin had come down to 0.89 mg/dl with conjugated bilirubin of 0.72mg/dl. The liver biopsy was suggestive of marked expansion of portal tracts by fibrosis, cholestasis, and inflammation in hepatic lobules. The child was referred to the liver transplant unit for further evaluation regarding the need for transplantation. The parents provided informed consent for the publication of this case.

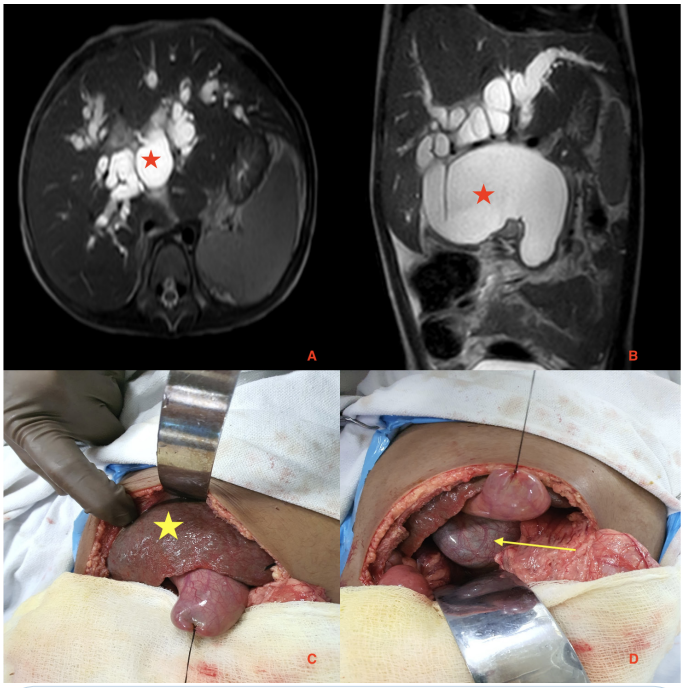

Table 1: MRI axial **(A)** and coronal **(B)** images show a large choledochal cyst (red star) with involvement of intrahepatic ducts. Intraoperative images **(C)** show a nodular liver (yellow star) and **(D)** a large choledochal cyst (yellow arrow) with multiple vascular collaterals over the cyst.

Discussion

There are various proposed theories regarding the etiopathogenesis of the CDC. The most accepted is Babbitt's theory of an Abnormal Pancreato-Biliary Duct Junction (APBDJ), which forms a common channel that allows the reflux of pancreatic juice into the bile duct and that the action of the pancreatic enzymes on the bile duct wall causes inflammation and weakening of the wall which ultimately results in dilatation and cyst formation. Although APBDJ is not present in all CDCs, the validity of this theory remains in doubt. Some have advocated that CDC occurs due to the narrowing of the distal CBD, presumably beginning in utero. The ductal plate malformation theory has been proposed for intrahepatic cysts and hepatic fibrosis [3,4].

Currently, surgical options are guided by the type of cyst and associated hepatobiliary pathology. Complete cyst excision with cholecystectomy and Roux-en-Y hepatico-jejunostomy reconstruction is the standard therapy in type I and the extrahepatic component of types IVA and IVB cysts. The extent of resection in type IVA cysts is still controversial, with concerns over the long-term complications (i.e., recurrent cholangitis, hepatolithiasis, secondary biliary cirrhosis, and malignant transformation) related to the residual intrahepatic disease following the standard approach as mentioned earlier. With such apprehension, some authors advocate segmentectomy, sectionectomy, or hemi-hepatectomy with Roux-en-Y biliary-enteric reconstruction for an intrahepatic disease localized to a resectable portion of the liver [5,6]. Conversely, in patients with diffuse involvement of the liver, biliary drainage procedures have been deemed ineffective, and a liver transplant is considered an ideal option [7].

In the index case, the ideal surgical procedure could be endoscopic stenting followed by liver transplantation, as stenting would be a temporary measure to decompress the cyst and alleviate back pressure from the liver [7]. Factors were considered

before surgical intervention in the index case: the child's clinical condition, the availability and accessibility of pediatric transplants, and the parent's financial status.

In a resource-limited country, especially in the pediatric population, an interval procedure such as the excision of the extrahepatic part with a Roux-en-Y hepaticojejunostomy can provide time for the patient till resources are arranged for definitive treatment. A successful bilio-enteric anastomosis partly alleviates the symptoms and decreases the patient's morbidity and hospital admissions/visits. We also believe that as the child's clinical/ nutritional status improved, the success of liver transplantation would also be increased.

Conclusion

Surgical intervention in the form of cyst excision and bilio-enteric drainage should be offered to selected patients with type IV-A CDC presents with signs of liver failure, even when it is not feasible to excise intrahepatic cyst. The surgery improves the patient's clinical condition and provides adequate time to make arrangements for liver transplantation.

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