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Surgical Hypothesis

Mega-choledochus - Is it pathologically Similar to Megacolon?

Rajah Shunmugam, Vinodh Mutyala Kumar, Vinodh Suppiah, Ahmad Toha

Department of Pediatric Surgery, Gleneagles Hospital, Kota Kinabalu, Sabah, Malaysia.

Keywords
Aganglionosis
Bile duct pathology
Choledochal cyst
Congenital megacolon
Enteric nervous system
Etiopathogenesis

Abbreviations
CDC - Choledochal cyst
CBD - Common bile duct
CHD - Common hepatic duct

Abstract

The traditional classification and etiopathogenic theory of choledochal cyst (CDC) has recently been challenged. A 16-month-old baby girl presented with jaundice, abdominal distention and clay colored stools for 2 weeks. Investigations revealed a Type IVA CDC. At laparotomy, a hugely distended biliary apparatus with an apparent blind distal end was noted. Excision of the CDC and Roux-en-Y hepatico-jejunostomy was performed. Her recovery is marked by a transient ascites for just 2 weeks. Histopathology of the excised distal bile duct showed hypertrophied nerve bundles similar to Hirschsprungs disease. Ultrasonography after 4 mo of surgery revealed complete regression of intra-hepatic duct dilation. From these, it appears that Todani Type I and Type IVa CDC may be just variations of the same disease.

INTRODUCTION

The traditional concept of choledochal cyst (CDC) and its anatomical classification by Todani have recently been challenged.[1,2] According to modern view, CDC is part of a larger spectrum of a fibro-cystic liver disease complex that includes a group of distinct disease entities with diverse etiologies, clinical manifestations, natural courses, complications and therapeutic options.[2] This report adds yet another dimension to the pathogenesis of CDC as a disorder of enteric nervous system.

CLINICAL EVIDENCE

A 16-month-old baby girl with a birth-weight of 2.6 kg had abdominal distension, jaundice and clay colored stools for the past 2 weeks. Previously she was normal except for a short spell of physiological jaundice during the first 2 wk of life that resolved with observation. Meconium history and the stool color were normal at birth.

On examination, there was a huge cystic mass in the abdomen extending from the epigastrium to the pelvis. Hemoglobin was 6.6 g/dl and leukocyte count was $22.5x10^9$ cells/l. Serum levels of total bilirubin 80.9µmol/l (serum direct bilirubin 70.3 µmol/l), albumin 25 g/l, globulin 36 g/l, alkaline phosphatase 2466 IU/l, aspartate transaminase 402 IU/l, alanin transaminase 321 IU/l and gamma glutamyl transferase 1921 IU/l. Computed tomography and ultrasonography showed hugely dilated intra-hepatic ducts, common hepatic ducts (CHD), gall bladder, cystic duct and common bile duct (CBD). (Fig. 1) The diameter of intra-hepatic ducts was 2.1-1.6 cm and that of the extra-hepatic ducts was 6 cm.

She was resuscitated with antibiotics, packed red blood cell transfusions and albumin infusions. On laparotomy ascites and a huge cyst occupying the entire abdomen were noted. About 300 ml of thick bile was aspirated from the cyst to deliver it into the wound. The cyst was arising from the CBD. The grossly dilated gallbladder, cystic duct, CBD, and CHD were dissected. The distal end of the CBD was apparently blind ending (Fig 2). Entire cyst was resected. The bile duct was transected at the bifurcation of the CHD and a Roux-en-Y hepatico-jejunostomy was done. Histology of the lower end of the cyst wall showed hypertrophied nerve bundles without a well-demarcated muscularis propria. (Fig. 3)

She tolerated oral feeds from the day-3 of surgery. Post-operatively she developed ascites (not leak) that required spironolactone and frusemide for 2 wk. She was discharged on the 11th day of operation. Liver enzymes reverted to normalcy within 2 wk. An ultrasonography after 4 months showed near normal caliber of the intra-hepatic ducts. (Fig. 4)

DISCUSSION

The exact etiopathogenesis of CDC is not clearly understood. According to the most widely accepted congenital theory of Babbitt, CDC is caused by an anomalous pancreato-biliary duct junction in which the pancreatic duct joins the CBD at a point 1-2 cm proximal to the sphincter of Oddi.[3] Other theories of pathogenesis include, weakness of the CBD wall, congenital or acquired ductal obstruction and dual theory (weakness of CBD wall cum ductal obstruction).[2,3]

Rolleston[4] (1905), Weber[5] (1934) and Saltz[6] (1956) speculated that CDC may be due to defective intra-mural neurons similar to Hirschsprung disease; but they did not provide any histopathological proofs.[4-6] Kusunoki[7] proposed that oligoganglionosis in the narrow portion of distal CBD results in proximal dilation similar to achalasia cardia and Hirschsprung disease. By manometric studies, Devenport and Basu[8] suggested that the round or fusiform choledochal cysts (Todani type-1) were congenital cysts with distal obstruction due to aganglionosis and proximal dilation similar to Hirschsprung disease. Histological evidences of our case supports these views.

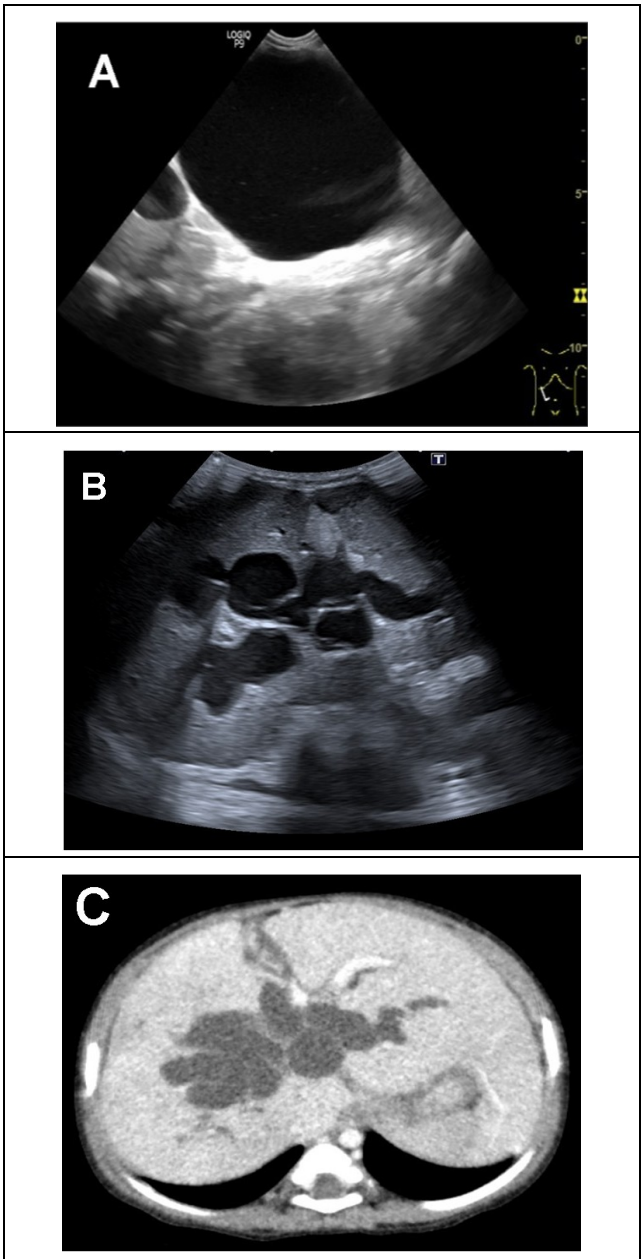

Fig 1. *Imaging studies of choledochal cyst. (A) Ultrasound showing dilated common bile duct. Ultrasound (B) and computed tomography (C) showing dilated intrahepatic biliary radicals.*

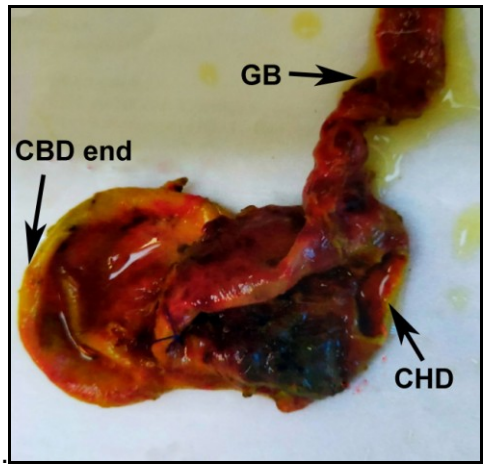

Fig 2. *Excised specimen of choledochal cyst showing dilatation of the gall bladder (GB), common hepatic duct (CHD) and the apparently blind ending dilated common bile duct (CBD)*

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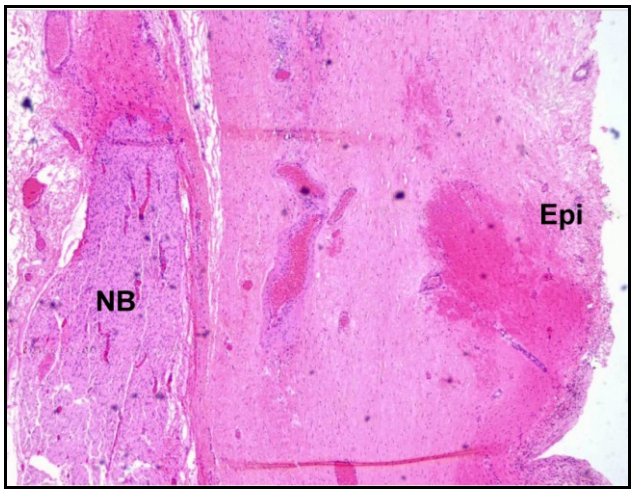

Fig 3. *Histology of the wall of choledochal cyst showing ulcerated epithelium (Epi) and hypertrophied nerve bundles (NB). Magnification 4 X, Eosin - Hematoxylin stain*

Complete regression of intra-hepatic duct dilation after hepatico-jejunostomy in our case supports the views of Visser[9] that type I and IVA cysts are variation of the same disease and the degree of intra-hepatic dilation defining one type versus the other was arbitrary. Complete occlusion of the distal end of the CDC in our case supports the

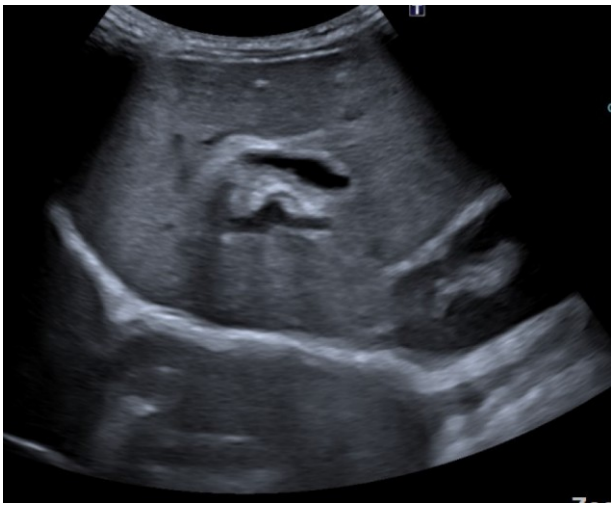

Fig 4. *Ultrasonography after 4 months of operation showing near normal caliber of the intra-hepatic ducts*

view of Diao[10] that ligation of the narrow distal stump of CDC may not be necessary.

CONCLUSION

Blind ending distal CBD may be due to segmental absence of ganglion cells with chronic infection similar to Hirschsprung disease. Complete regression of dilation of intra-hepatic ducts after surgery supports this etio-pathogenesis. More focus on Histopathology of CDC and revision of classifications need to be considered.

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Address for communication: *Dr. Rajah Shunmugam, Email: srajah1@hotmail.com*

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