Forme Fruste Choledochal Cyst

Ashish Wakhlu Y.K. Sarin Rajiv Agarwal Brijesh Lahoti A.K. Sharma

We report a case of 'forme fruste choledochal cyst' due to a congenital web of the lower end of the common bile duct (CBD) and highlight the unusual features of this entity.

Case Report

A 3-year-old girl was admitted to the Pediatric Surgical Service with intermittent episodes of jaundice and spiking fever with chills for 1 year. There was a history of passing acholic stools but the family and past medical history were unremarkable. Physical examination showed a pale, jaundiced and malnourished child with firm non tender hepatomegaly 5 cm below the costal margin. She had a hemoglobin of 7 g/dl, the liver function tests showed a total bilirubin of 8 mg/dl (direct 6.6 mg/dl) and the transaminase levels were minimally elevated. Ultrasonography of the abdomen showed a dilated CBD (1.5

From the Department of Pediatric Surgery, S.M.S. Medical College and Allied, Sir Padampat Institute of Mother and Child Health, Jaipur, Rajasthan.

Reprint requests: Dr. Ashish Wakhlu, B 1-99, Sector 'G', Aliganj, Lucknow 226 020

Received for publication: February 16,1994; Accepted: January 1,1995 cm) with dilatation of the intrahepatic bile ducts. The liver parenchyma was normal. Endoscopic retrograde cholangiopancreatography showed dilatation of the CBD with a partially obstructing web in its lower end. There was associated dilatation of the intrahepatic ducts as well (Fig. 1). There was an anomalous pancreatobiliary union in the form of a long common channel (length 16 mm, diameter 4 mm). The pancreatic duct was otherwise normal.

At operation the dilated CBD was resected at the junction of the right and left hepatic ducts proximally and at the pancreatobiliary junction distally. The distal part of the duct was oversewn, the web was included in the resected portion. A roux en Y hepaticojejunostomy was performed into a 30 cm long defunctionalized loop of jejunum brought up through a retrocolic window. The post operative recovery was uneventful and the patient has remained well during a follow up of 18 months. Histopathological examination of the resected CBD showed dense connective tissue in the wall along with some smooth muscle. There was an intense inflammatory infiltration and the epithelial lining was deficient except for some flattened patches. These features were identical to those of a choledochal cyst.

Discussion

Choledochal cysts are associated with several pancreatobiliary variants(1). The term forme fruste choledochal cyst was proposed by Lily *et al.*(2) to describe one of these variants in which the cystic dilatation of the common duct is minimal or absent but there

INDIAN PEDIATRICS VOLUME 32-JULY 1995

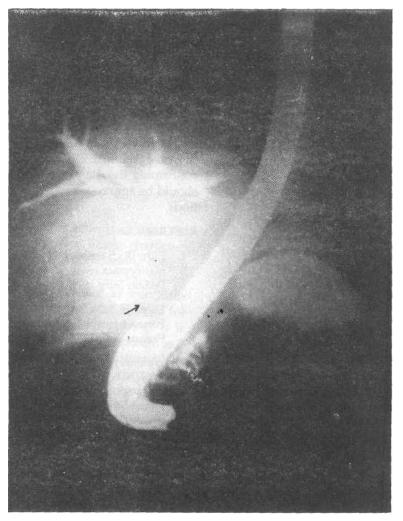


Fig. 1. Endoscopic retrograde cholangiopan creatogram showing minimal dilatation of the common bile duct with a partially obstructing congenital web in the lower end.

is a long pancreatobiliary union, partial obstruction of the lower common bile duct and histopathological changes identical with choledochal cyst in the wall. Additionally, the patient presents with

recurrent cholangitis and some have evidence of gross or microscopic cholecystitis, and abnormalities of the intrahepatic biliary channels. The authors described four patients and

categorically stated that the only satisfactory treatment was complete excision of the diseased ducts and restoration of continuity with intestinal anastomosis to healthy ducts.

The present patient satisfies most of the above criteria regarding presentation, histopathology and intrahepatic ductal morphology but the obstruction of the common bile duct in this case was caused by a congenital web in the lower end. There was an anomalous pancreatobiliary union in the form of a long common channel as described by Weidmeyer et al. (3). On the basis of the above it is reasonable to label this patient as a type of forme fruste choledochal cyst caused by an unusual type of CBD obstruction. ERCP made the preoperative diagnosis of this entity possible and we emphasize that this investigation should form an integral part of the management of such patients.

Treatment by primary complete excision was successful in providing relief from symptoms. Anastomosis of intestine to healthy biliary ducts was not possible as the intrahepatic biliary chan-

nels were also involved. Short term follow-up has shown complete recovery but longer observation is necessary.

Forme fruste choledochal cyst is uncommon but not rare(1). As there is minimal dilatation of the common bile duct, a high index of suspicion is necessary to diagnose it in patients presenting with recurrent cholangitis. The presence of an anomalous pancreatobiliary union and demonstrable lower CBD obstruction are helpful. ERCP is valuable in confirming the diagnosis and treatment should be aggressive to avoid complications.

REFERENCES

- Lilly JR. Surgery of coexisting biliary malformations in choledochal cyst. J Pediatr Surg 1979,14: 643-645.
- 2. Lilly JR, Stellin GP, Kramer FM. Forme fru'lte choledochal cyst. J Pediatr Surg 1985, 20: 449-451.
- 3. Weidmeyer DA, Stewar ET, Dodds UJ, *et al.* Choledochal cyst, findings on cholangiopancreatography with emphasis on ectasia of the common channel. AJR 1989,153: 969-972.