Case Report

Congenital biliary web- a rare cause of obstructive jaundice in an infant: A case report

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ABSTRACT

Background: Congenital biliary web of the extra-hepatic biliary tree is becoming exceedingly rare cause of obstructive jaundice in children.

Case Presentation: We report a case of 5-month-old male baby who presented with acholic stools and persistent jaundice since birth. Magnetic resonance cholangiopancreatography (MRCP) showed contracted gall bladder and focal narrowing at mid portion of the common bile duct (CBD) with proximal dilatation of biliary channels. On exploration, a complete web was found just proximal to the confluence of cystic duct and common hepatic duct causing complete obstruction of biliary tree. A Roux-en-Y hepatico-jejunostomy was done. Postoperative recovery was uneventful.

Conclusion: We conclude that congenital biliary web is a rare entity and should be considered in the differential diagnosis of biliary atresia.

Keywords: Congenital biliary web, Obstructive jaundice, Biliary atresia, Stricture

INTRODUCTION

Biliary webs are extremely rare, with approximately less than 50 cases have been reported until now in the literature.[1] The presence of congenital variations of the extra-hepatic biliary tree is estimated to be approximately 10%. [2] The congenital causes of obstructive jaundice include choledochal cysts, biliary atresia, biliary web, CBD stricture, and extra-hepatic biliary ducts compression by vascular abnormalities.[1,3,4]

Herein, we are reporting a case of congenital complete common hepatic biliary web as a rare cause of extra-hepatic biliary obstruction in an infant. It is the youngest case of congenital complete common hepatic duct web to the best of authors’ knowledge and first one to be reported from Pakistan.

CASE REPORT

A 5-month-old male baby was referred to our gastroenterology department from a primary health care facility with history of acholic stools and persistent jaundice since birth. Past medical /surgical history, family history of such disease, hematological history and perinatal history were unremarkable. On physical examination, he was deeply jaundiced, liver was mildly enlarged (palpable 4cm below costal margin) with firm in texture and smooth surface without nodularity, and spleen was palpable. At our institute, patient was investigated for underlying cause of obstructive jaundice in the Gastroenterology department. Laboratory investigations showed conjugated hyperbilirubinemia (total serum bilirubin level: 14.4mg/dl, direct bilirubin: 10mg/dl). Alkaline phosphatase was significantly raised.
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(2864U/l) and so was Gamma-glutamyl transferase (1251U/l). Ultrasound abdomen depicted contracted gall bladder, dilated intra-hepatic biliary channels with the common bile duct (CBD) outlined. Magnetic resonance cholangiopancreatography (MRCP) was performed which showed contracted gall bladder and a focal narrowing at mid portion of CBD with proximal dilatation of intra-hepatic biliary channels (Fig.1). Provisional diagnosis of CBD stricture with the differential diagnosis of biliary atresia was made and patient was referred to Pediatric Surgery department.

After optimization, per-operative cholangiogram was performed, which showed a small but patent gallbladder, patent distal CBD as contrast outlined the duodenum and proximal jejunum. The distal CBD was clamped with bulldog clamp and the contrast material was again injected; however proximal CBD could not be outlined. Furthermore, there was a palpable thickening of CBD just distal to common hepatic duct which was slightly dilated. Aspiration of dilated common hepatic duct revealed bile in it. CBD was explored and a complete web was found just proximal to the confluence of cystic duct and common hepatic duct (CHD) causing complete obstruction of the biliary tree (Fig.2). The diaphragm at the confluence of CHD and cystic duct along with the gall bladder was excised and sent for histopathology, and “Roux-en-Y” hepatico-jejunostomy was performed. Patient was shifted to Surgical Intensive Care Unit (SICU) where postoperative stay was uneventful.

Bilirubin levels declined progressively and by 9th postoperative day, bilirubin level declined to 2.5mg/dl and stool color became normal. Patient was discharged on 15th post-operative day. The histopathology report of the submitted sample showed gall bladder and biliary tree with the signs of chronic inflammation. The patient was doing fine till 8th month of follow-up as outpatient department visits and then lost to follow-up afterward.

Figure 1: MRCP showing dilated intra-hepatic biliary ducts (Green arrow) and dilated common hepatic duct (Yellow arrow) with focal narrowing at mid portion of Common bile duct (Red arrow). The distal CBD outlined normally. GB indicating gall bladder.

Figure 2: Operative picture showing extensively mobilized and explored Common Bile Duct (CBD) and web of the common hepatic duct (arrow).

DISCUSSION

Common bile duct webs are extremely rare anomalies of the extra-hepatic biliary ducts with only a handful of cases reported in literature.[5] The embryogenic background of this anomaly seems to resemble that of the mucosal septum within intestines (type I atresia). During the development of the human embryo, the bile ducts pass through a solid stage followed by recanalization to form the lumen of the extra-hepatic biliary tree during the fifth week of gestation. Incomplete recanalization is presumed to develop congenital biliary webs.[2]

Biliary web may be partial or complete.[6] Complete biliary web usually presents in early life with jaundice, or occasionally with gallbladder perforation while partial web presents late in life with history of recurrent cholangitis and choledocholithiasis.[1,2] Only a few cases of complete biliary web in children have been reported.[6] The reported associated abnormalities include anomalous hepatic duct of the caudate lobe or anomalous junction of the pancreaticobiliary ductal system.[5]

Congenital biliary webs must be differentiated from other causes of direct hyperbilirubinemia such as
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biliary atresia, and CBD strictures. In our case, as the baby had acholic stools since birth along with persistent jaundice, our first diagnosis was delayed presenting biliary atresia. CBD stricture was documented by MRCP as our second differential diagnosis. Since in the index case, the gallbladder and distal CBD was patent as shown by passage of contrast material to the duodenum. This may be confused with type II-a of biliary atresia (Kasai classification system) where the gallbladder and distal CBD are also patent.[7] The main differentiating point was patenty of entire biliary tract with merely presence of a mucosal web as a cause of biliary obstruction. Moreover, intra-hepatic biliary dilatation is never characterized by biliary atresia.

The reported treatment for biliary web consisted of endoscopic dilatation of the (partial) web, or excision of the web with a cholecystectomy and “Roux-en-Y” hepatico-jejunostomy, excision of the diaphragm with t-tube insertion.[1,6,8,9] We performed Roux-en-Y hepaticojejunostomy as CBD was extensively dissected and could not be preserved.

In conclusion, congenital biliary web should be considered in differential diagnosis of biliary atresia especially in those cases where intra-hepatic biliary dilatation is documented on imaging. The importance of reporting this case is to highlight the significance of suspecting these rare causes of obstructive jaundice which can be easily corrected surgically.

Conflict of Interest: MBM, and NT are members of the editorial team.

Consent to Publication: The authors declared taking consent for publication from legal guardians of the patient.

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