ANJAN KUMAR DHUA
SANDEEP AGARWALA
VEERESHWAR BHATNAGAR

Corresponding Author: Dr Veereshwar Bhatnagar
Email: veereshwarb@hotmail.com

References

1. American Cancer Society (2011). Colorectal Cancer Facts and Figures 2011–2013. Atlanta: American Cancer Society.
2. Boyle P, Ferlay J. Mortality and survival in breast and colorectal cancer. Nat ClinPract Oncol. 2005;2:424–425.
3. O'Connell JB, Maggard MA, Liu JH et al. Rates of colon and rectal cancers are increasing in young adults. Am Surg. 2003;69:866–872.
4. O'Connell J B, Maggard M A, Livingston E H et al. Colorectal cancer in the young. Am J Surg. 2004;187:343–348.
5. Rao BN, Pratt CB, Fleming ID et al. Colon carcinoma in children and adolescents. A review of 30 cases. Cancer. 1985;55:1322-6.
6. Yang R, Cheung MC, Zhuge Y et al. Primary solid tumors of the colon and rectum in the pediatric patient: a review of 270 cases. J Surg Res. 2010; 161:209-216.
7. Pandey A, Gangopadhyay A, Sharma S et al. Pediatric carcinoma of rectum--Varanasi experience. Indian J Cancer. 2008;45:119-122.
8. Ries L AG, Melbert D, Krapcho M et al. SEER cancer statistics review, 1975–2005. Bethesda, MD: 2008.
9. Jackson-Thompson J, Ahmed F, German RR et al. Descriptive epidemiology of colorectal cancer in the United States, 1998-2001. Cancer. 2006;107:1103–1111.
10. Vastyan AM, Walker J, Pintér AB et al. Colorectal carcinoma in children and adolescents--a report of seven cases. Eur J Pediatr Surg. 2001;11:338-341.
11. Karnak I, Ciftci AO, Senocak ME et al. Colorectal carcinoma in children. J Pediatr Surg. 1999;34:1499-1504.
12. Poles GC, Clark DE, Mayo SW et al. Colorectal carcinoma in pediatric patients: A comparison with adult tumors, treatment and outcomes from the National Cancer Database. J Ped Surg. 2015. DOI: http://dx.doi.org/10.1016/j.jpedsurg.2015.11.005 (Article in press)
13. Sahoo SP, Gangopadhyay AN, Gupta DK et al.Carcinoma of the rectum in children. Pediatr Surg Int. 1996;11:418-420.
14. Brown RA, Rode H, Millar AJ et al. Colorectal carcinoma in children. J Pediatr Surg. 1992 ;27:919-921.
15. Hirsch BR, Zafar SY. Capecitabine in the management of colorectal cancer. Cancer Management and Research 2011;3: 79–89.

Congenital Web in the Common Hepatic Duct - An Uncommon Cause of Obstructive Jaundice in a Child

A web in the common hepatic duct (CHD) is an infrequent cause of obstructive jaundice.[1] Bile duct anomalies like hepatic duct stenosis have been reported in choledochal cysts, but bile duct webs, and especially CHD webs, are extremely rare. Herein, we report the case of a young girl with obstructive jaundice and cholangitis that was attributed to a congenital web in the CHD.

Case Report

A 3 and a half yearold girl was brought to our institute with complaints of yellow discoloration of eyes for 4 months and passage of pale colored stools for 2 months. The ultrasonography did not reveal any evidence of cholelithiasis or choledocholithiasis; however the intrahepatic biliary radicals were dilated on both sides. She was managed as cholangitis and was discharged after she improved with antibiotics. Further assessment of the cause of jaundice and cholangitis was done with an MRCP. It showed a bilobar intra-hepatic biliary radical dilatation (IHBRD) with CHD dilatation and abrupt narrowing of the extra-hepatic bile duct caliber at the site of insertion of the cystic duct, suggestive of extra-hepatic biliary obstruction with a possibility of a congenital biliary stricture.

On admission, the clinical examination showed icterus and her liver was palpable 3 cm below the right costal margin. Based on the investigations, an exploration was planned. Intra-operatively **(Figure 2a)** the CHD was dilated till just short of its confluence with the cystic duct; beyond it, the CBD was narrow and collapsed. Additionally, the gut was found to be malrotated. After transecting the CHD just distal to the narrowed portion, a bulging membrane was noted that completely occluded the CHD **(Figure 2b)**. On excising the membrane, copious flow of bile was seen without any sludge or calculi from the proximal dilated bile duct system. Excision of

the CBD with a Roux-en-Y hepatico-jejunostomy and Ladd's procedure were performed. Post-operatively, the patient showed significant improvement with resolution of jaundice and fall in serum bilirubin levels within a fortnight. The histopathology report of the excised web showed hyalinised fibrocollagenous tissue focally lined by columnar epithelial cells.

Discussion

Obstructive jaundice in the pediatric age group is generally due to choledochal cysts or extrahepatic bile duct atresia. Calculi in the gall bladder or the common bile duct are uncommon as compared to the adult population. Other causes of obstruction of the extrahepatic biliary tree in adults are strictures secondary to primary sclerosing cholangitis or malignancy. Congenital stenosis of the hepatic duct is rare and is generally found in association with choledochal cyst. In addition to stenosis at the region of the confluence of both the hepatic ducts, congenital stenosis of the hepatic duct may also have anomalies of the pancreatico-biliary ductal junction or an abnormal hepatic duct of the caudate lobe.[2,3]

Congenital mucosal "webs" of the extrahepatic biliary tree areanother rare cause of obstructive jaundice. They are distinct from strictures secondary to trauma, sclerosing cholangitis, or radiation exposure. If they form a complete septum, they present early in life with progressively increasing jaundice. They may progress to secondary biliary cirrhosis if left untreated or may get complicated by spontaneous perforation of the biliary tract.[2] They often tend to have a small lumen, which results in partial occlusion only and hence they remain asymptomatic, till they get entirely occluded by inflammation because of recurrent cholangitis or stones in the proximal bile duct or gall bladder.4 In our case, the young age of the patient and the absence of any other primary pathology suggested the possibility of a congenital mucosal web that became symptomatic because of complete closure of a tiny orifice.

The extrahepatic bile duct attains a hollow tubular structure from the initial stage of the solid embryonic cord by the process of re-canalization. The webs possibly appear because of an incomplete or erratic re-canalization

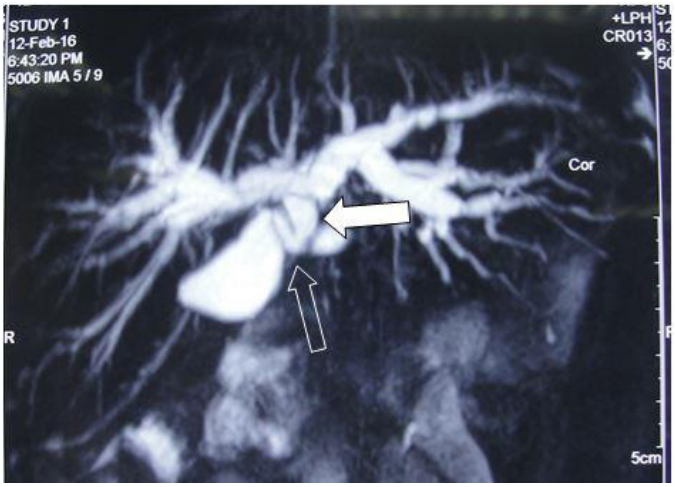

Figure 1: The MRCP showing bilobar IHBRD with dilated CHD (solid white arrow) with a sudden cut-off after cystic duct joins the CHD (arrow with white outline). The rest of the bile duct is not visualized.

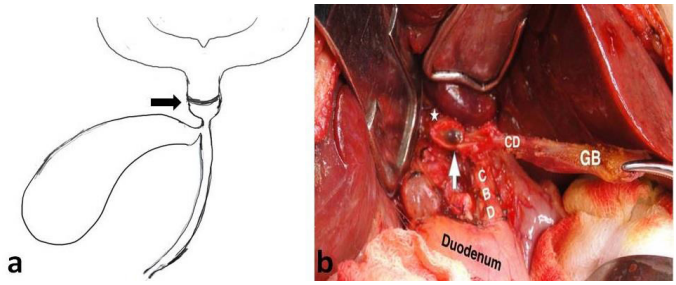

Figure 2: (a) A schematic diagram showing the position of the web in the extrahepatic bile duct. (b) Intra-operative image showing the dilated CHD (arrow) and the mucosal web (*) within it. The web was seen bulging down because of pent up bile. The gall bladder (GB) has been dissected off the gall bladder fossa (swung to the left side) and the cystic duct (CD) shown to be joining the CHD to form the collapsed CBD.

process, similar to the pathogenesis of duodenal atresia.[3] Depending on the exact site involved, the aberration may produce a web at the CBD or the CHD.

A mucosal web of the CHD is a rare situation, and only a handful of cases have been reported.[1] In 1994, Kato et al., described a 48-year-old lady with gallbladder carcinoma and an anomalous junction of the pancreatico-biliary channel with a 'septum' of the CHD.[2] In 1992, Furukawa reported a 66-year-old lady with jaundice who was later found to have a web in the CHD. She

also had an anomalous hepatic duct of the caudate lobe.[3] Margolis and Schein in 2001 reported a 65-year-old Chinese-American lady with the clinical features of acute cholecystitis.[1] During cholecystectomy and subsequent CBD exploration they found a mucosal web of the CHD. They also reviewed the cases of extra-hepatic bile duct webs and other similar intra-luminal causes of obstructive jaundice reported in literature till then. A search of the current literature did not reveal any pediatric patient to have been reported with an isolated mucosal web in the CHD leading to obstructive jaundice.

Diagnosis may be missed on ultrasonography, but with MRCP, indirect evidence like proximal IHBRD with an abrupt cut off of the dilated duct at an unexpected location may suggest webs. CBD exploration would be the most effective way to pick the webs, as in our case. Intra-operative cholangiography must also be used when the findings are inconclusive. During exploration, care should be taken since these thin webs can be inadvertently punctured or torn even with minimal instrumentation or probing, which may lead to a missed diagnosis.

Management of the web requires transecting the involved segment of extra-hepatic bile duct containing the web, followed by a bilio-enteric anastomosis. Simple excision and incision plus dilatation have also been described, but the long term results of these procedures are not known.[1] We removed the segment of CHD containing the web along with the distal collapsed CBD and ligated the lower end. A hepatico-jejunostomy was done to restore bilio-enteric continuity and Ladd's procedure was performed, given the associated malrotation of the midgut.

A rare cause of obstructive jaundice in the pediatric age group is reported. 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. When exploring the CBD, the surgeon must be cautious as the webs can easily be damaged and missed. Excision of the segment of bile duct and bilio-enteric anastomosis is generally curative. Prompt relief of the obstruction of bile flow also helps in avoiding morbidity.

RAVI PATCHARU[1]
ANJAN KUMAR DHUA[1]
KANIKA SHARMA[1]
ROHAN MALIK[1]
MANISHA JANA[2]
VEERESHWAR BHATNAGAR[3]

Department of [1]Pediatrics, [2]Radiodiagnosis and [3]Pediatric surgery, All India Institute of Medical sciences, New Delhi.

Corresponding Author: Prof. Veereshwar Bhatnagar
Email: veereshwarb@hotmail.com

References

1. Margolis M, Schein M. Biliary web -a rare cause of extrahepatic biliary obstruction. Dig Surg. 2001;18:317-9.
2. Kato S, Nakagawa T, Kobayashi H, Arai E, Isetani K. Septum formation of the common hepatic duct associated with an anomalous junction of the pancreaticobiliary ductal system and gallbladder cancer: report of a case. Surg Today. 1994; 24:534–7.
3. Furukawa H, Hara T, Taniguchi T. A case of septum formation of the common hepatic duct combined with an anomalous hepatic duct of the caudate lobe. GastroenterolJpn. 1992; 27:102–7.
4. Devanesan J, DeBlasi HP, Sable R, Hoe R, Kinkahabwala M: Congenital hepatic duct obstruction with perforate diaphragms. Arch Surg. 1978;113: 1452–5.

Gastric Adenocarcinoma with Yolk Sac Differentiation, Fungal Super Infection and A Large Solitary Liver Metastasis - A Histologic Conundrum

Gastric adenocarcinoma with yolk sac differentiation are rare tumours. They are usually AFP secretors and presents with high AFP levels. Also the neoplastic cells