Pediatrics and Neonatology xxx (xxxx) xxx

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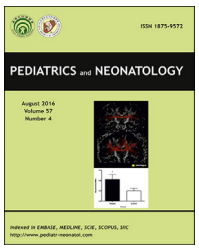

Images

Inspissated bile syndrome: A rare cause of neonatal cholestasis

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Received Aug 3, 2019; received in revised form Sep 18, 2019; accepted Mar 5, 2020
Available online ■ ■ ■

A male neonate with a birthweight of 3220 g was transferred to our hospital on the eighth day of life because of progressive neonatal hyperbilirubinemia. His serum total bilirubin and direct bilirubin levels were 22.3 and 0.8 mg/dL, respectively. After 72 h of adequate hydration and phototherapy, these levels had increased to 26.0 and 5.8 mg/dL, respectively. Other liver function tests were normal except for gamma-glutamyl transpeptidase (234 IU/L) and alkaline phosphatase (230 IU/L). An obstruction of the common bile duct (CBD) due to biliary sludge was demonstrated on abdominal ultrasonography (Fig. 1A and B) at 12 days of age and in follow-up magnetic resonance cholangiopancreatography 3 days after ultrasound examination (Fig. 2). Intraoperative cholangiography via the punctured gall bladder was performed at 16 days of age because of no dissolution of the bile sludge and no improvements in bilirubin levels after oral ursodeoxycholic acid (UDCA) therapy. Inspissated black sludge was noted in the drainage catheter after saline lavage of the bile duct (Supplementary Fig. S1). Ultrasonography on the fourth day post operation revealed significant clearance

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https://doi.org/10.1016/j.pedneo.2020.03.002

Please cite this article as: Chen S-M et al., Inspissated bile syndrome: A rare cause of neonatal cholestasis, Pediatrics and Neonatology, https://doi.org/10.1016/j.pedneo.2020.03.002

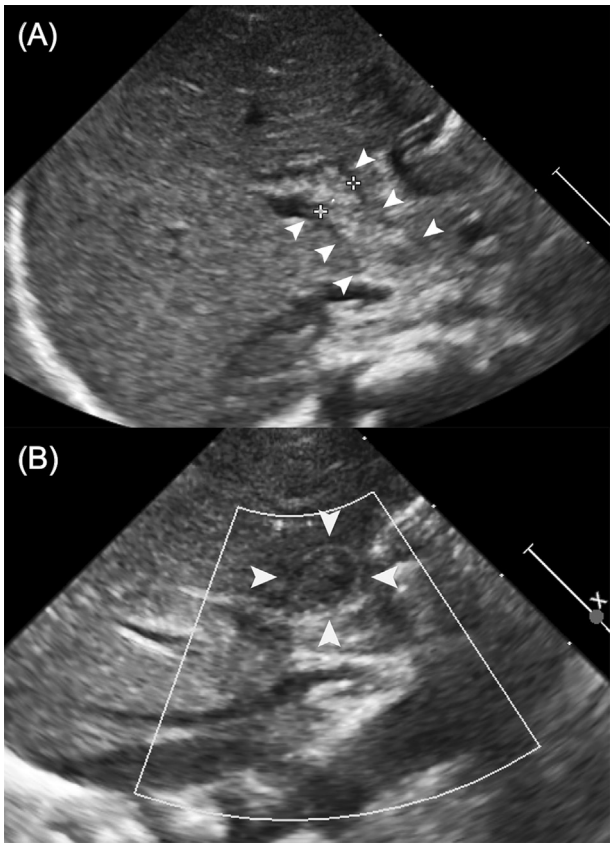

Fig. 1 Abdominal ultrasonography showed mild intrahepatic biliary dilatation and a dilated common bile duct (CBD) (6 mm) filled with a slurry-like substance (A; indicated by arrow). A transverse ultrasound scan of the CBD showed that it was dilated and filled with slurry (B).

and decreased the diameter of the CBD (Supplementary Figure S2). The patient's serum bilirubin levels returned to normal 40 days after surgery, and other biochemical test results normalized at 3 months of age.

Inspissated bile syndrome (IBS) is defined as extrahepatic obstruction of the bile duct by bile sludge without congenital biliary malformations, bile acid synthesis defects, or hepatocellular causes of jaundice. The estimated incidence of IBS is 1 in 175,000 live births, accounting for 8% of all types of surgical jaundice during infancy.[1] Several risk factors have been associated with the formation of biliary sludge in infancy, including prematurity, parenteral nutrition, dehydration, hemolytic diseases, cystic fibrosis, congenital heart diseases, cefotaxime, ceftriaxone, and sepsis.[1] In some cases, biliary sludge has been reported to resolve spontaneously with or without UDCA treatment.[2] Surgical intervention is indicated when the bile duct is dilated to >3 mm with persistent jaundice.[2,3] Berger et al. described their clinical experience of successfully treating a 6-week-old infant with IBS via laparoscope-aided cholecystostomy with biliary duct lavage.[3]

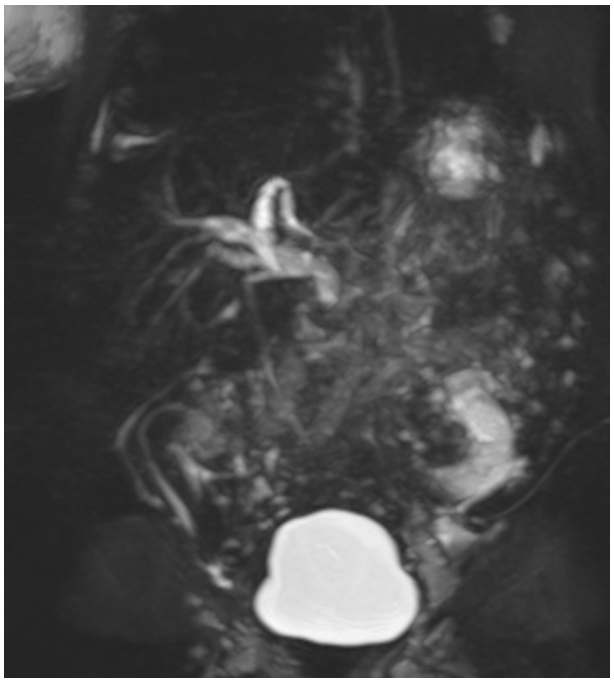

Fig. 2 Magnetic resonance cholangiopancreatography showed a small gallbladder, diffuse moderate dilatation of intra- and extrahepatic biliary ducts, and abrupt tapering of the CBD.

Declaration of Competing Interest

The authors declare no conflicts of interest regarding this manuscript.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.pedneo.2020.03.002.

Please cite this article as: Chen S-M et al., Inspissated bile syndrome: A rare cause of neonatal cholestasis, Pediatrics and Neonatology, https://doi.org/10.1016/j.pedneo.2020.03.002