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Extrahepatic biliary obstruction due to stone

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SUMMARY Jaundice developing after the neonatal period requires prompt investigation. Surgical causes of cholestatic jaundice, apart from extrahepatic biliary atresia and choledochal cyst, are rare. We describe a case of bile duct dilatation associated with an impacted calculus at the ampulla of Vater in a haemophiliac. The ultrasound finding of a dilated duct was an indication to proceed to surgery.

Full investigation of cholestatic jaundice in infancy is important to diagnose those lesions which may be correctable surgically. The differential diagnosis of extrahepatic biliary obstruction includes extrahepatic biliary atresia, bile duct perforation, stenosis and cyst, and biliary 'sludging'. We present a case of common bile duct stone which, although extremely rare and readily treatable, was difficult to diagnose.

Case report

A boy was born at term to an obligate carrier of haemophilia A. At birth his factor VIII value was 2.5%. He was breast fed and developed mild unconjugated jaundice (on day 3 his total bilirubin concentration was 158 $\mu\text{mol/l}$; conjugated bilirubin 11 $\mu\text{mol/l}$) which had cleared by day 7. At 18 weeks of age he became intermittently fretful with occasional screaming attacks. Physical examination was normal. Biochemical evidence of haematoma was excluded by finding normal haptoglobin and methaemalbumin concentrations. Total bilirubin

concentration was 50 $\mu\text{mol/l}$, aspartate transaminase 308 U/l (normal 5 to 40 U/l), and alkaline phosphatase 274 U/l (normal 35 to 130 U/l). Serological tests for toxoplasma, toxocara, rubella, cytomegalovirus, herpes simplex, syphilis, varicella zoster, and hepatitis A and B were negative. Abdominal ultrasound was normal. After one week his bilirubin concentration had fallen to 11 $\mu\text{mol/l}$, his aspartate transaminase concentration was 145 U/l, and he was much more content.

Two weeks later he again became very fretful, and developed overt jaundice with pale, foul smelling stools and pruritus. A liver edge was now palpable 4 cm below the costal margin. His bilirubin concentration at this time was 60 $\mu\text{mol/l}$ and his conjugated bilirubin was 48 $\mu\text{mol/l}$. Routine haematology and biochemical determinations were normal, as were the immunoglobulins, α_1 antitrypsin, plasma amino acids, thyroxine, sweat sodium and chloride, prothrombin time, and amylase. Urine was negative for reducing substances on three occasions. The liver function tests became increasingly abnormal, with aspartate transaminase 298 U/l, alkaline phosphatase 910 U/l, and total bilirubin concentration 138 $\mu\text{mol/l}$. His prothrombin ratio was 4. Other abnormal results included serum cholesterol 20.9 mmol/l (normal 3 to 6.5) and triglycerides 2.9 mol/l (normal 0.2 to 1.5). Real time ultrasonography on this occasion showed dilated intra- and extrahepatic bile ducts and a common bile duct of 11 mm maximum diameter. The gall bladder was distended but contained no sludge or calculi. The grossly dilated system made a diagnosis of choledochal cyst likely and he was transferred for surgery.

With adequate transfusion of factor VIII concentrate, a laparotomy was performed. The gall bladder and bile ducts were grossly dilated and an area of induration was palpated at the lower end of the common duct. Cholecystography showed a filling defect suggestive of a calculus in the ampullary region. The duodenum was opened and a stone 5×2.5 mm in diameter was found impacted at the ampulla of Vater. The calculus was extracted after sphincterotomy and a sphincteroplasty 5 mm long was fashioned to ensure free drainage of bile into the duodenum. He made an uneventful recovery after operation and received daily transfusions of factor VIII concentrate for two weeks. Within two weeks of surgery the liver function tests (bilirubin, aspartate transaminase, and alkaline phosphatase) were all within the normal range. A follow up ultrasound examination one month after surgery showed a normal common bile duct of 5 mm diameter.

Discussion

Cholelithiasis is an uncommon paediatric diagnosis and contrary to popular belief most cases are not associated with haemolytic disorders but occur in adolescent girls without any particular underlying cause. In a review of 367 cases of cholelithiasis in children only a single case occurred in an infant.¹ Calculi within the common bile duct are even less frequently described. The most common presentation is with perforation of the common bile duct and biliary ascites.^{2,3} There have been only two previously reported cases of choledocholithiasis presenting with obstructive jaundice in infancy.³

Ultrasonography has emerged as one of the most valuable non-invasive investigations for conjugated hyperbilirubinaemia. The ultrasonographic anatomy of the paediatric biliary tree has been determined⁴ and is particularly sensitive in differentiating in-

trahepatic disease from extrahepatic biliary dilatation.⁵ Distinguishing these two main groups can be very difficult and requires extensive investigation, such as liver biopsy and rose bengal faecal excretion.⁶ The finding, however, of a dilated extrahepatic biliary tree is a clear indication to proceed to surgical treatment. Liver biopsy and percutaneous transhepatic cholangiography may be performed before operation, the latter providing information on the site of obstruction and biliary anatomy, but carry the risk of bile leakage and peritonitis. In our patient these investigations were strictly contraindicated by the presence of haemophilia.

Biliary calculi are most reliably detected by ultrasound. A calculus impacted at the sphincter of Oddi is notoriously difficult to diagnose on ultrasound because the increased echogenicity of the head of the pancreas obscures the stone. Although the stone was not seen in our patient, the resultant biliary dilatation rendered further investigations unnecessary pending the results of laparotomy.

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