Spontaneous Perforation of the Extrahepatic Bile Ducts in an Infant

Spontaneous perforation of the extrahepatic bile ducts, with an accumulation of bile in the peritoneal cavity, is a rare condition. Nevertheless, it is second in frequency to biliary atresia as the cause of obstructive jaundice during the first year of life. Although approximately 60 cases of this condition have been described in the literature, it is only occasionally listed in pediatric textbooks. Nevertheless, it is important that the condition be recognized in time to perform lifesaving surgical intervention. We therefore consider it instructive to present a patient with the classic features of this condition who was cured by surgical treatment on the 29th day of life.

CASE REPORT

J.S. was a full-term, normally delivered 2,850-gm girl who was breast-fed during the first two weeks of life and then given a standard formula. She was admitted to our ward at age 24 days because of three days of feeding problems, including vomiting, irritability, and weight loss. She had discolored stools but demonstrated no jaundice. Her abdomen was not distended and there was no splenomegaly or fever. The urine was normal in color. Over the subsequent five days she did not gain weight. She was fed breast milk but had a gastric retention that increased from 25% to 60% of the administered breast milk. A roentgenologic examination of her stomach on admission disclosed no abnormalities (Figure, upper left). Laboratory values including hemoglobin level, peripheral WBC count, and electrolytes were normal. The liver enzyme levels were also normal, except for that of γ-glutamyltransferase which was moderately elevated. Serum bilirubin was 50 μmoles/liter (3 mg/100 ml). No urobilinogen, protein, galactose, glucose, or bacteria were found in the urine. Metabolic screening produced normal results.

Repeated roentgenologic examination of the infant's stomach because of further vomiting at 28 days of age revealed a deformity of the first and second parts of the duodenum (Figure, upper right, bottom left). Passage of contrast agent to the distal part of the duodenum was delayed, as compared with the previous examination. At this time the infant had mild jaundice.

Laparotomy was performed at 29 days of age. Through an upper abdominal transverse incision, a green, glittering peritoneum could be observed. Clear, bile-stained fluid was found in the peritoneal cavity. The hepatoduodenal ligament was inflamed, edematous, and bile stained. The area of papilla of Vater was found to be thickened when palpated through the walls of the duodenum. The gallbladder was not distended but its serosal lining was thickened by edema. The portahepatis area was severely inflamed. A cholecystojejunostomy was performed. A preoperative cholangiogram was not performed. A drainage tube was inserted into the area of the foramen of Winslow.

The infant's postoperative course was uneventful. Within 24 hours her stools were of normal color, and from the second postoperative day she was given increasing amounts of breast milk. Ampicillin was administered intramuscularly from the day of operation but was replaced by clindamycin after seven days, as low-grade fever and discolored stools were interpreted as ascending cholangitis. Her temperature and stools normalized during the next 24 hours. The clindamycin treatment was continued for a further three weeks.

Approximately 25 ml of bile was collected daily during the first eight postoperative days, after which no bile could be aspirated and the drain was removed 11 days after the operation.

At 12 months of age, the girl was growing well and her stools and urine were of normal color. She was being fed a standard formula. The intravenous cholangiogram was normal, as was the roentgenologic picture of her stomach and small intestine (Figure, bottom right).

DISCUSSION

Spontaneous perforation of the common bile duct with sterile bile peritonitis is a rare, subacute condition. Its diagnosis is therefore often delayed. The site of the perforation is usually in the area where the cystic and common ducts join. The inflammatory reaction forms pseudocysts in the upper abdomen, with leakage of bile into the peritoneal cavity.

Many theories have been proposed regarding the etiology of the condition, including viral infection, fat necrosis from pancreatic reflux, distal common duct obstruction, and ruptured choledochal cyst. One fairly convincing theory includes a localized embryonic mural malformation with consequent weakness of the common duct. High pressure in the common bile duct could produce either a rupture or the formation of a true choledochal cyst. Reports on spontaneous perforations mainly involve infants during their first three months of life, whereas a choledochal cyst usually arises later.

The typical clinical findings in our case included an uneventful neonatal period of three weeks followed by the onset of non-bile-stained vomiting, failure to thrive, discolored stools, subclinical to mild jaundice, and normal-colored urine. This clinical picture suggested a perforation. In addition, others have described progressive distension of the abdomen, and in some cases also bile-stained um-

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biliary and inguinal hernias and hydroceles.\textsuperscript{7,8} Usually there is no fever or significant leukocytosis. The liver enzymes may be normal but the bilirubin level is often slightly elevated. The \textsuperscript{131}I-rose-bengal fecal excretion test or \textsuperscript{131}I-cholic acid test will show accumulation of radioactivity in the ascitic fluid.\textsuperscript{8} Aspiration of bile-stained ascites has therefore been suggested as a means of confirming the diagnosis of bile peritonitis.

The treatment of this condition is unconditionally surgical but there is some disagreement regarding the method of choice. Simple drainage, as suggested by Lilly et al.,\textsuperscript{1} has been successful in many cases but proved fatal in others, probably because of distal choledochal obstruction due to inflammatory compression or by inspissated bile. At a preoperative cholangiography in these cases, a distal choledochal obstruction is often visualized. This requires bile diversion by cholecystoenterostomy which, however, has been complicated in some cases by ascending cholangitis. The use of a Roux-en-Y loop or cholecystojejunostomy combined with distal jejunojejunostomy has successfully minimized this risk.

Postoperatively performed cholangiograms have shown that the perforation heals within a few weeks

Figure. Upper left, Patient at 24 days of age with normal appearance of duodenal loop. Upper right, Patient at age 28 days with narrowing and somewhat irregular deformity of postbulbar and descending parts of duodenum (arrows). Lower left, Same examination as in

upper right with double contrast technique. Irregular deformity can be seen in region of papilla vateri as well as in postbulbar region (arrows). Lower right, Patient at age 38 days, nine days postoperatively, again showing normal appearance of duodenum.
and that free passage of bile into the duodenum is restored if drainage of the perforated area is adequate. This leads to the conclusion that common duct exploration with repair of the perforation by suturing is unnecessary and inexpedient.

The prognosis of this clinically as well as pathologically well-defined disease is good when it is recognized in time for proper surgical treatment.

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REFERENCES

NEUROPSYCHIATRIC STATUS OF INCARCERATED DELINQUENTS

The neuropsychiatric status of a sample of incarcerated white female juvenile offenders is compared with that of a sample of incarcerated white male juvenile offenders. Males and females had a similar prevalence of psychiatric and neurologic impairment. More violent males were significantly more impaired than less violent males. There was a tendency for more violent females to have greater impairment than less violent females. The study questions the prevailing literature indicating greater psychopathology in female offenders than male offenders. It explores issues of sex and race bias in the assessment of the neuropsychiatric status of offenders.

Submitted by R. J. Haggerty, MD

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