PREOPERATIVE DIAGNOSIS OF ASYMPTOMATIC CHOLEDOCHAL CYST BY ROSE BENGAL LIVER SCAN

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The number of choledochal cysts reported in the literature has increased in recent years. The classic triad of abdominal pain, jaundice, and an abdominal mass leads to suspicion of the diagnosis in less than 20% of cases. The purpose of this report is to emphasize that rose bengal liver scanning is the most definitive and atraumatic method of diagnosing choledochal cyst in jaundiced or nonjaundiced patients.

Choledochal cyst is a congenital cystic dilatation of any segment of the extrahepatic biliary ducts, and most commonly the common bile duct is involved. The disease occurs predominantly in children and young adults (*1*). The disorder is more common in females than males (*1,2*). Orientals are affected more frequently than any other race (*3*). The clinical triad of abdominal pain, jaundice, and abdominal mass is the most helpful diagnostic clue; however, the classic triad is present in only about 20% of the cases (*4*). A roentgenographic triad of soft-tissue mass, duodenal impression, and nonvisualized gallbladder is also a quite helpful finding. In the absence of jaundice an intravenous cholangiogram may be specific for the diagnosis of choledochal cyst (*2,4,5*). In 1970 Williams, et al first reported a choledochal cyst diagnosed by hepatoscintigraphy (*6*). The purpose of this paper is to re-emphasize the diagnostic value of rose bengal liver scanning in this disorder.

CASE REPORT

JN, a 12-year-old boy, was admitted to the pediatric service of the Thomas Jefferson University Hospital on March 9, 1973 for evaluation of a mass in the right upper abdomen. The mass was found on routine examination by his local physician. There was no history of vomiting, diarrhea, abdominal pain, or jaundice. The patient had been in excellent health until 3 years before when he was diagnosed elsewhere as having infectious hepatitis and subsequent to the initial episodes, he experienced recrudescent hepatitis 3 weeks and 1½ years later. The patient had physiologic jaundice after birth, and there was no history of blood incompatibilities.

Physical examination revealed a well-developed, thin white male in no acute distress. He was afebrile, with pulse of 105, respiratory rate of 22, and blood pressure of 115/80. The sclerae were not icteric. The heart and lungs were essentially normal. Abdominal examination revealed a large mass in the right upper quadrant which was firm and smooth to palpation. Vertical measurement of liver was 18 cm, and the spleen was not palpable. Bowel sounds were normal.

The hemoglobin was 14.2 gm% and the hematocrit was 39.6%. The white cell count was 9,900 with a normal differential. Urinalysis, blood urea nitrogen, and electrolytes were within normal limits. Total bilirubin was 0.7 mg% with 0.3 mg% of direct bilirubin. Serum cholesterol (240 mg%; normal = 120–230) and SGOT (72 mg%; normal = 7–40) were slightly elevated and alkaline phosphatase LDH were normal.

A scout radiograph of the abdomen demonstrated

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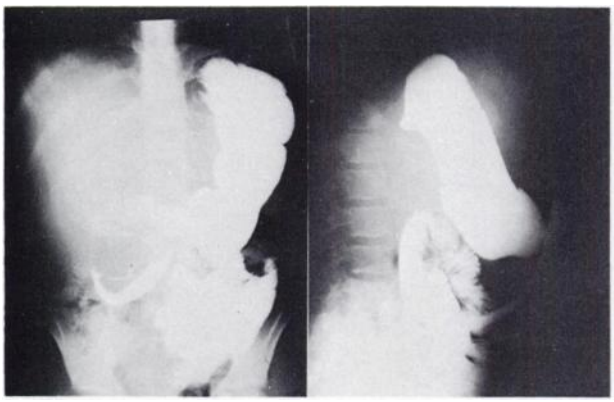

FIG. 1. AP and lateral views of upper GI series demonstrate displacement of stomach to left and anteriorly. Duodenal bulb is displaced antero-inferiorly. Second and third portions of duodenum are stretched by large extrinsic mass and notice filling of main pancreatic duct with barium.

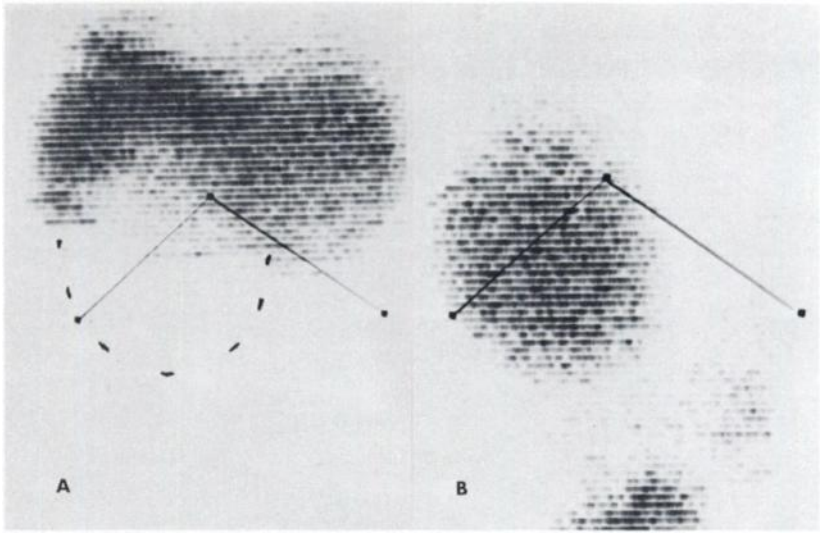

FIG. 2. (A) Anterior view of liver scan with ^{99m}Tc-sulfur colloid reveals large defect (dots) in lower portion of right lobe. (B) Anterior view of 5-hr rose bengal scan shows complete filling of large defect with isotope and some excretion of isotope into small intestine.

a large soft-tissue mass in the right upper abdomen. On an upper gastrointestinal series the stomach was displaced to the left and anteriorly. The duodenal bulb was displaced markedly anteriorly almost to the anterior abdominal wall. The second and third portions of the duodenum were markedly stretched by a large extrinsic mass. The main pancreatic duct was filled with barium (Fig. 1). An oral cholecystogram failed to visualize a gallbladder. An initial liver scan was performed with ^{99m}Tc-sulfur colloid because of clinical suspicion of retroperitoneal lymphoma, and the scan demonstrated a defect in the lower portion of the right lobe. A 2-hr scan with ^{131}I-labeled rose bengal revealed excretion of the isotope by the liver and concentration of the isotope in the area of the filling defect noted in ^{99m}Tc-sulfur colloid scan. A delayed scan at 5 hr showed good concentration of the isotope in the choledochal cyst (Fig. 2). A celiac arteriogram was scheduled, but the study was cancelled because the scan finding was thought to be pathognomonic for a choledochal cyst.

On March 15, 1973 at laparotomy, the common bile duct was converted into a large choledochal cyst containing at least 1,500 cc of bile and innumerable small pigmented calculi. The descending limb of the duodenum was circled by an extremely broad annular pancreas. A cholecystectomy, choledocho-cysto-duodenostomy, and duodeno-jejunostomy was performed. The postoperative course was uneventful, and the patient was discharged on the twelfth postoperative day.

DISCUSSION

Choledochal cyst is not a rare disease and the number of cases reported has increased in recent years (*2*). Early diagnosis and treatment of this disorder can prevent serious complications; however, choledochal cysts have in the past seldom been correctly diagnosed preoperatively (*1*). The clinical triad (abdominal pain, abdominal mass, and jaundice) and the roentgenographic triad (soft tissue mass, duodenal impression, and nonvisualized gallbladder) should lead to the correct preoperative diagnosis in a limited number of cases. Direct radiographic demonstration of the disorder by intravenous cholangiogram is possible only during a remission of the jaundice (*5*). More recently the preoperative diagnosis of choledochal cyst by rose bengal liver scan was reported (*6*), and scan findings are pathognomonic for the disorder. Since liver scanning proved to be a relatively safe procedure in children, it is the authors' opinion that a rose bengal scan should be performed before special radiographic procedures when clinical suspicion of choledochal cyst exists.

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