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Spontaneous bile duct perforation in a neonate

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ABSTRACT

We present a case of spontaneous biliary perforation (SBP) in a previously healthy 3-week-old boy with worsening scleral icterus, abdominal distension, and acholic stools. Abdominal ultrasound demonstrated moderate ascites without extrahepatic ductal dilatation. The HIDA scan demonstrated accumulation of tracer within the right upper quadrant outside of the liver but not in the bowel, however there was a large amount of tracer activity seen diffusely within the abdomen that appeared to lie within the ascitic fluid and complicated the read of the scan. Intraoperative cholangiogram during an exploratory laparotomy confirmed an intact gallbladder with a mostly contained perforation at the junction of the cystic duct and the common bile duct with no contrast in the duodenum. The patient underwent an uneventful cholecystectomy with a Roux-en-Y hepaticojejunostomy and was discharged home three days later.

More than fifty cases of spontaneous biliary perforation (SBP) have been reported in infants and neonates [1,2]. Various etiologies have been suggested, including pancreaticobiliary malunion, ischemia, or an inherent weakness at the junction of the cystic and common hepatic ducts [3–5]. The majority of cases present with acute deterioration or distress at birth, but occasionally these children may have a subacute, indolent presentation. We report on a 3-week-old boy with spontaneous perforation of the bile duct managed with single-stage reconstruction.

1. Case report

A 3-week-old boy, born full term with a benign early postnatal course, presented with worsening jaundice and acholic stools. On examination, positive findings included jaundice and scleral icterus. The abdomen was soft and nondistended with a palpable umbilical hernia and a unilateral hydrocele. Serum liver enzymes were elevated (AST 116 U/L and ALT 88 U/L). Total bilirubin was elevated at 17.8 mg/dL, with a conjugated value of 11.9 mg/dL. Abdominal ultrasound demonstrated a normal liver and gallbladder with no dilation of the intra- or extrahepatic ducts, however there was mild to moderate ascites. A HIDA scan, preceded by a phenobarbital load, demonstrated immediate uptake in the liver, followed by accumulating focal activity in the upper abdomen, thought to be too posterior to represent the gallbladder (Fig. 1).

Of note, the imaging also demonstrated hazy, diffuse activity throughout the abdomen, suspected to be within the ascites, which made gut activity difficult to confirm. Given the current stability of the child, a laparotomy with intraoperative cholangiogram was performed. Upon entry into the peritoneum, 500 mL of bilious peritoneal fluid was evacuated. An intraoperative cholangiogram demonstrated contrast filling the gallbladder and traversing the hepatic ducts into the liver, however there was significant extravasation of contrast into the left side of the abdomen (Fig. 2).

This finding was confirmed with a second contrast injection after ensuring there was no leak around the cholangiogram catheter. Upon exploration, a small perforation was discovered at the confluence of the cystic and common bile ducts. Additionally, a biloma cavity was noted between the stomach and liver (Fig. 3).

A cholecystectomy and Roux-en-Y hepaticojejunostomy was performed, with an anastomotic drain. Post-operatively the neonate tolerated oral feedings with resolution of hyperbilirubinemia (total bilirubin of 3.4 mg/dL from 17.8 mg/dL on admission).

2. Discussion

A recent literature review determined the incidence of SBP to be 1.5 in 1,000,000 live births, with the median age of diagnosis at 2.4 months old [1]. There is no clear etiology for SBP however, three potential etiologies have been proposed. First, a congenital biliary wall weakness due to ischemia of the anterior common bile duct related to a posterolateral arterial supply has been postulated [3,4]. Supporting the theory of congenital wall weakness and ischemia is the observation that the

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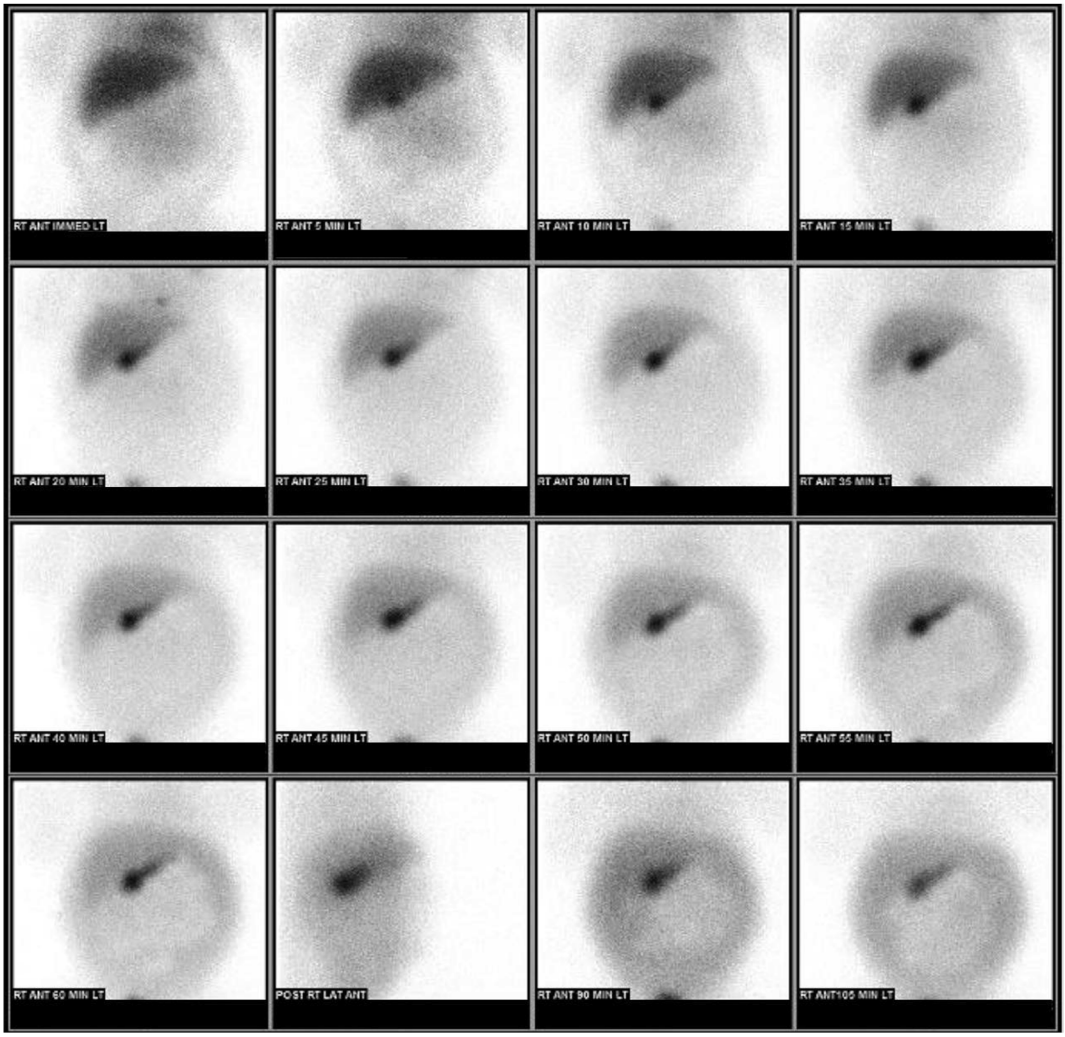

Fig. 1. HIDA series demonstrating accumulation within the gallbladder, without clear confirmation of duodenal activity due to large amount of diffuse accumulating activity within the abdomen that appeared to lie within the ascitic fluid.

most common site of perforation is at the junction of the cystic duct and common bile duct [1]. A second possible etiology is a pancreaticobiliary malunion near the sphincter of Oddi that would allow reflux of pancreatic juices into the bile duct, leading to acute inflammation and biliary obstruction with increased intraluminal pressure causing perforation [5,6]. Lastly, Moore et al. suggested that SBP seemed to exist on a continuum with choledochal cyst as a form of acquired biliary atresia [7]. This opinion is supported by the observation that SBP seems to occur in children two weeks to five months of age, and perforated choledochal cysts usually present six months to seven years [8,9].

While the presentation of SBP is variable, the most common symptoms are abdominal distension, jaundice, acholic stools, and nonbilious emesis [1]. Evans et al. also noted that hernias and hydroceles were seen in 16% of SBP cases, presumably from increased intra-abdominal pressure following perforation [2]. When presented with a neonate or infant that has jaundice and acholic stools, the surgical differential should include biliary atresia, SBP, and spontaneous perforation of a choledochal cyst. An asymptomatic interval between birth and presentation should decrease the suspicion for biliary atresia, however, SBP and perforation of a choledochal cyst usually present after an asymptomatic interval. It is more difficult to differentiate SBP and perforated choledochal cyst. As noted above, the age of SBP presentation is usually younger than perforated choledochal cysts [8,9].

Imaging is frequently helpful to elucidate the etiology. Abdominal ultrasound has been shown to be a useful diagnostic tool identifying ascites, ductal dilatation, a fluid collection, or a pseudocyst in cases of SBP [2]. Jeanty et al. noted that RUQ bilomas found during a laparotomy had often been mistaken for an enteric duplication cyst, pancreatic pseudocyst, or choledochal cyst. A HIDA scan is highly sensitive and specific for SBP and is virtually diagnostic when it shows extravasation into the peritoneal cavity with no activity in the duodenum [10]. Over the past several years, magnetic resonance cholangiopancreatography (MRCP) has gained more recognition as a diagnostic tool for SBP. A recent study by Goel et al. stated MRCP may even be better than ultrasound at visualizing a loculated fluid collection or pseudocyst due to its superior soft tissue resolution and multiplanar capability [11].

The optimal management strategy of SBP has been debated. However, nearly all studies agree that an intraoperative cholangiogram should be performed to determine the location of the perforation, to detect any ductal abnormalities, and to exclude distal obstruction [2,12]. If a perforated choledochal cyst is discovered, a single-stage biliary cholecystectomy with a Roux-en-Y hepaticojejunostomy is frequently performed to minimize the risk of developing a cholangiocarcinoma later in life [13]. In the management of SBP, three interventions have been advocated: 1) non-operative management with antibiotics and drainage, percutaneous or ERCP; 2) surgical biliary drainage utilizing external drains, a cholecystostomy tube, or a common duct T-tube; 3) biliary reconstruction via primary repair of the perforation or Roux-en-Y hepaticojejunostomy. Overall, previous