Paediatric infant presenting with an atypical spontaneous biliary perforation

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Spontaneous biliary perforation (SBP) is a life-threatening condition. Although rare, it is the second most common surgical cause of jaundice in infancy after biliary atresia. SBP is theorized to occur due to a localized injury to the embryological developing biliary tree, predominantly at the junction of the cystic and the common ducts. The weakness usually develops on the anterior wall of the junction of the common and the cystic ducts. Insults to the wall, such as from infection, trauma or increased biliary pressure, can subsequently lead to perforation. We report a case of an atypical SBP in a 3-week-old girl presenting with a biloma. *Ann Pediatr Surg* **10:14–16** © **2014 Annals of Pediatric Surgery.**

Annals of Pediatric Surgery 2014, **10**:14–16

Keywords: biloma, paediatric, spontaneous biliary perforation, surgery

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Received 21 April 2013 **accepted** 26 October 2013

Introduction

Spontaneous biliary perforation (SBP) is a rare condition, yet is the second most common surgical cause of jaundice in infancy. It occurs due to an embryological insult to the developing biliary tree, most commonly at the anterior junction of the common and the cystic ducts. The injury leads to weakness of the wall, which can result in perforation. We report a case in which an infant was operatively diagnosed with a biloma secondary to an atypical SBP – located on the posterior wall of the distal cystic duct.

Case report

A 3-week-old girl presented to the emergency department with a 1-week history of increasing abdominal distension, reduced feeding and progressively acholic stools. There were no complaints of fever, vomiting, jaundice or abdominal trauma. The patient had normal antenatal scans and was an induced vaginal delivery at 38 weeks with no other postnatal complications. On examination, the child appeared grey with distress on handling, but was hemodynamically stable and afebrile. There was evidence of a moderate tracheal tug with increased respiratory effort when the patient was supine, which improved with positioning. Abdominal exam revealed a soft, yet grossly distended abdomen, with dullness to percussion 10 cm inferior to the right costal margin. Laboratory results revealed a lymphocytic leucocytosis (lymphocyte 12.6×10^9/l, white cell count 23.8×10^9/l), hyperbilirubinemia 53 μmol/l (≤ 14) and elevated γ-glutamyl transferase 436 U/l (5–26 U/l). Electrolytes and coagulation studies were unremarkable. Ultrasound indicated a large simple cyst lying within the parenchyma of the right lobe of the liver (Fig. 1). Magnetic resonance cholangiopancreatography (MRCP) demonstrated a large complex unilocular cystic structure with significant mass effect appearing to be closely related to, but not arising from, the liver, suggesting a large choledochal cyst (Fig. 2).

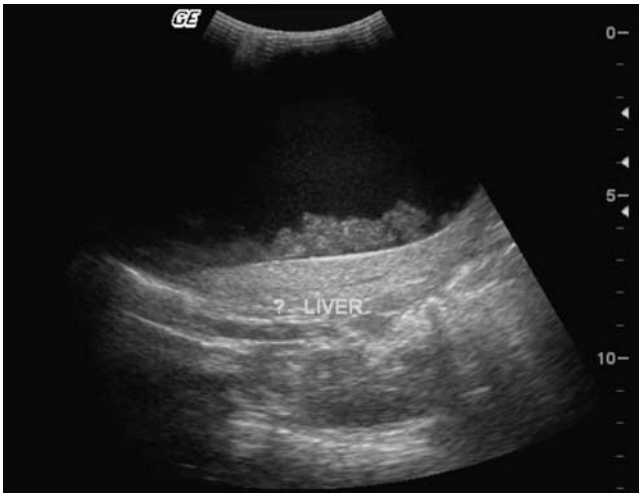
Laparotomy was planned with a provisional diagnosis of a congenital hepatic cyst. Intraoperatively, we found a cystic cavity filled with bilious fluid, with a pseudocapsule formed laterally by the abdominal wall, posteroinferiorly by the liver, medially by the falciform ligament and anteriorly by the peritoneum (Fig. 3). A bile leak due to perforation of the posterior wall of the distal cystic duct was identified. Cholecystectomy was performed with the cystic duct transfixed. An intraoperative cholangiogram indicated normal intrahepatic and extrahepatic biliary systems, with no extravasation (Fig. 4). The patient recovered well; she was extubated on day 1 postoperatively and discharged home on day 6. Outpatient follow-up was unremarkable, with the child gaining weight well.

Discussion

The first reported case of SBP was by Dijkstra in 1932 [1]. Since then, there have been less than 160 reported cases in children. Although it is a rare condition, it is described as the second most common surgical cause of jaundice in infancy after biliary atresia [2]. SBP is theorized to occur after insult to a weakened segment of bile duct wall. The currently accepted theory for the weakened bile duct wall is a congenital localized mural malformation that occurs during embryogenesis [3–5]. The weakness is commonly located anteriorly at the junction of the common and the cystic ducts. It has subsequently been hypothesized that if this area of weakness is large, it may gradually dilate and form a choledochal cyst rather than perforate [6,7]. Suspected insults to the weakened wall include cholelithiasis, infection, trauma, pancreatitis and congenital pancreaticobiliary anomalies. Whether ductal obstruction (secondary to stones, sludge or stenosis) leads to perforation or whether it results after perforation is currently unknown [6]. Currently, there are no reported cases of perforation in the posterior wall of the cystic duct.

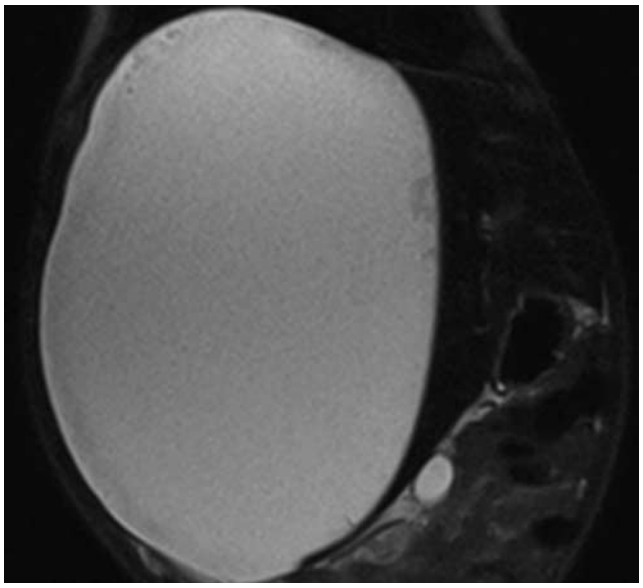
1687-4137 © 2014 Annals of Pediatric Surgery DOI: 10.1097/01.XPS.0000438126.32652.87

Fig. 1



Ultrasound imaging indicating a large simple cyst within the right lobe of the liver. The cyst has mildly hypoechoic peripheral nodular debris and is very well marginated.

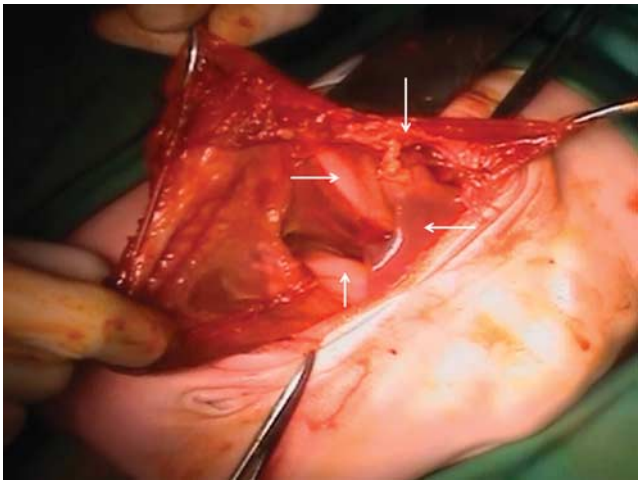
Fig. 2



Magnetic resonance cholangiopancreatography revealed a large complex unilocular cystic structure displacing and compressing the liver but not arising from within it. At the posterior/inferior margin, there is a tubular tail extending posteromedially to the region of the porta hepatis.

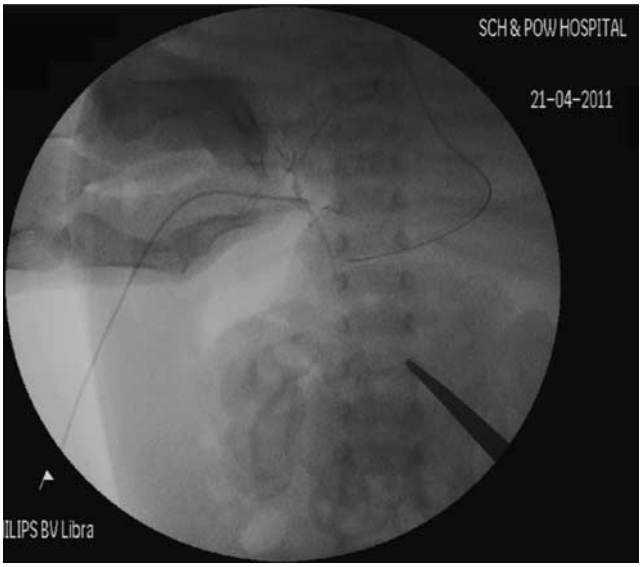
SBP most commonly occurs within the first year of life, with a median age of 4 months. There have been cases reported as early as 25 weeks gestation and as late as 15 years of age [8–10]. The typical presentation is of a previously well infant with an unremarkable antenatal history, who presents with abdominal distension and/or jaundice. Other symptoms include acholic stools, vomiting, fever and poor weight gain [11–13]. Although the constellation of symptoms might raise concern of biliary atresia, the previously asymptomatic history should rather cause suspicion of a SBP.

Fig. 3



An intraoperative picture with arrows depicting important structures (clockwise from top: pseudocapsule, liver, stomach and falciform ligament).

Fig. 4



Intraoperative cholangiogram through the perforated cystic duct indicating normal intrahepatic and extrahepatic biliary systems.

There are no pathognomonic laboratory results to aid in diagnosis. There can be mild leukocytosis, hyperbilirubinemia and elevated serum transaminases. Serum inflammatory markers can be elevated. These test results can differentiate SBP from neonatal hepatitis and biliary atresia [14]. If an ascitic tap is performed, bilirubin concentration in the fluid is higher than in the serum; however, paracentesis is unnecessary if imaging confirms biliary leakage [12].

Ultrasound is the common first step for imaging and can demonstrate intraperitoneal fluid and the biliary system. However, misdiagnosis is common: a dilated pseudosac can be misdiagnosed as a choledochal cyst in 45–60% of cases. [15] MRCP is very beneficial as a noninvasive

investigative tool. Loculated fluid collections/pseudocyst formations adjacent to the perforation site and associated bile duct anomalies are hard to visualize on ultrasound; MRCP can easily display these pathologies. MRCP is limited to diagnosing SBP if fluid collections are located in the lesser sac or the perirenal space [16]. If the patient is stable, the preoperative imaging of choice is radionuclide hepatobiliary scan. It is highly sensitive and specific for SBP and has the additional advantage of being able to detect the exact site of leak secondary to radiotracer accumulation at the perforation site [17].

Current literature advocates early surgical intervention [3,6]. One case has been reported in which a child was managed nonoperatively and survived [18]; however, the patient did not have confirmation of an infrahepatic bile collection, nor was any perforation site identified. Intraoperative exploration allows definitive localization of the perforation, and cholangiogram can evaluate the entire biliary system [10,14,19,20]. Nonobstructed biliary perforation can be managed with the placement of an external drain adjacent to the perforation with nil need for formal closure of the perforation [3,12,19–21]; most perforations will close spontaneously with decompression. In perforations with distal obstruction not resolved by a cholangiogram, a biliary-intestinal anastomoses is commonly recommended [14,19–21]. T-tube drainage is not recommended, nor is primary closure of the perforation, because of the risk of developing biliary stricture [12,20]. Some authors do not recommend exploration of the porta hepatis as inflammation significantly increases technical difficulty [21]. Endoscopic retrograde cholangiopancreatography has been used in both diagnostic and therapeutic modalities [11,22]. However, there are significantly limited resources and qualified personnel capable of investigating and treating paediatric SBP with endoscopic retrograde cholangiopancreatography.

Potential postoperative complications include bile leak, cholangitis, portal venous thrombosis and portal hypertension [12]. The risk of cholangitis can be reduced by the introduction of broad-spectrum antibiotics postoperatively [3,6]. Only a few cases have required a return to the theatre for causes such as biliary revision or portosystemic shunting [18]. Overall, paediatric SBP has low morbidity and mortality if there is early recognition and surgical intervention.

Conclusion

We present an atypical paediatric biloma, caused by a spontaneous perforation on the posterior wall of the distal cystic duct. The chosen imaging modalities, ultrasound and MRCP, did not diagnose the patient's pathology accurately. Previous authors recommend radionuclide hepatobiliary scans over MRCP because of its greater sensitivity and specificity; however, the erroneous provisional diagnoses did not result in any change of management.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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