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Spontaneous bile duct perforation with cystic fibrosis and meconium ileus

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ABSTRACT

Spontaneous bile duct perforation in the newborn is rare, with less than 150 cases reported. A term male newborn presented with abdominal distention and respiratory distress on the first day of life. Paracentesis revealed 420 mL of bile without succus or meconium, and laparoscopic washout and drainage was performed. A follow-up contrast enema several days later revealed a microcolon. The second operation confirmed the spontaneous bile duct perforation, but also revealed meconium ileus with ileal perforation and thick meconium. A sweat chloride study was abnormal and the patient was homozygous for the delta-F508 mutation. This is the first reported case of spontaneous bile duct perforation in association with meconium ileus or cystic fibrosis.

1. Case report

A newborn 37 6/7weeks gestation Caucasian male infant weighing 3075 g presented at birth with abdominal distention. He was born via cesarean section with no significant antenatal history and a normal newborn screening panel to a G2P2 mother. Shortly after birth he developed apnea and respiratory distress associated with worsening abdominal distention requiring intubation and chest compressions. He was subsequently transferred for a higher level of care. On exam the patient had severe abdominal distention noted with thin skin and visible veins. He had not passed any meconium. An abdominal radiograph demonstrated non-dilated, centralized lops of bowel with no gas in the expected location of the colon (Fig. 1). Abdominal sonography demonstrated fluid with calcifications.

An urgent bedside paracentesis was done for respiratory compromise and 420 mL of viscous, clear golden–green bile, with no succus or meconium, was drained. The excessive quantity of clear bile lead us to make the presumptive diagnosis of spontaneous bile perforation. He was then taken to the operating room for laparoscopic washout and drain placement.

At laparoscopy, chronic bile peritonitis was encountered as evidenced by bile-stained fibrinous exudate completely covering the bowel and liver which caused the bowel to be completely adherent to itself. As there was no meconium spillage or identification of small bowel perforation, the loops of bowel were not completely separated during this operation. A laparoscopic-guided intraoperative cholangiogram was preformed via the gallbladder with a micro-puncture needle. This study was technically difficult due to the bile peritonitis and a discrete site of leakage was not identified. The puncture site on the gallbladder was closed and the abdomen was irrigated at the end of the procedure. A closed suction drain was left in the right upper quadrant and the patient was transferred back to the neonatal intensive care unit.

Over the next few days there was no bilious drain output and no re-accumulation of fluid by ultrasound. The patient continued to be distended without any clinical signs of improvement. On post-operative day 3 a contrast enema (Figs. 2 and 3) was preformed to evaluate for obstruction. The patient was found to have microcolon as well as increasing pneumoperitoneum suggestive of intestinal perforation. The patient was therefore taken back to the operating room for a full exploratory laparotomy.

Intraoperatively the patient was found to have no significant re-accumulation of bile, but extensive bile-stained adhesions. Additionally, palpation identified very thickened meconium with obstruction as well as a small perforation in the distal ilium. A partial small bowel resection was preformed to include the area of perforation and an ileostomy with a mucus fistula was created. An

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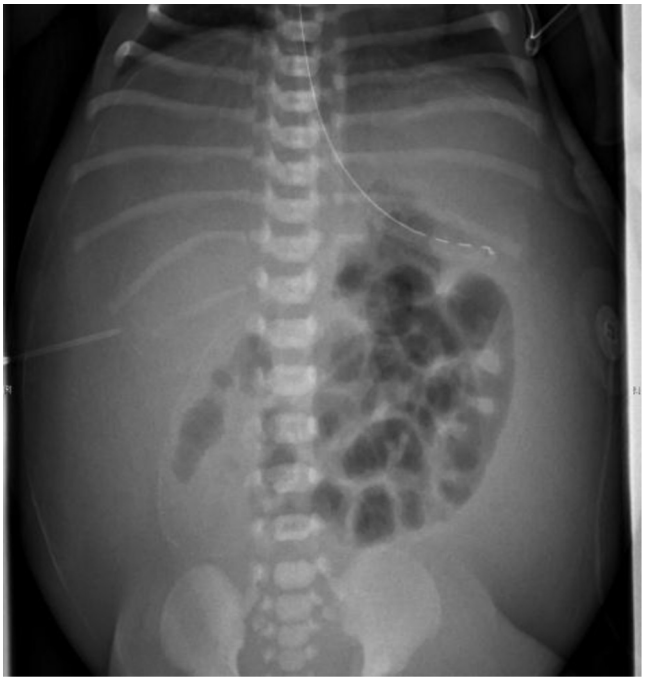

Fig. 1. Presenting KUB demonstrating dilated loops of bowel with no gas in the expected location of the colon.

intraoperative cholagiogram was repeated through the gallbladder to confirm the original diagnosis of spontaneous bile duct perforation in light of the additional pathology. This study nicely demonstrated a small leak from the cystic duct (Fig. 4) confirming the original diagnosis. Another drain was placed under the liver and the patient was transferred back to the NICU.

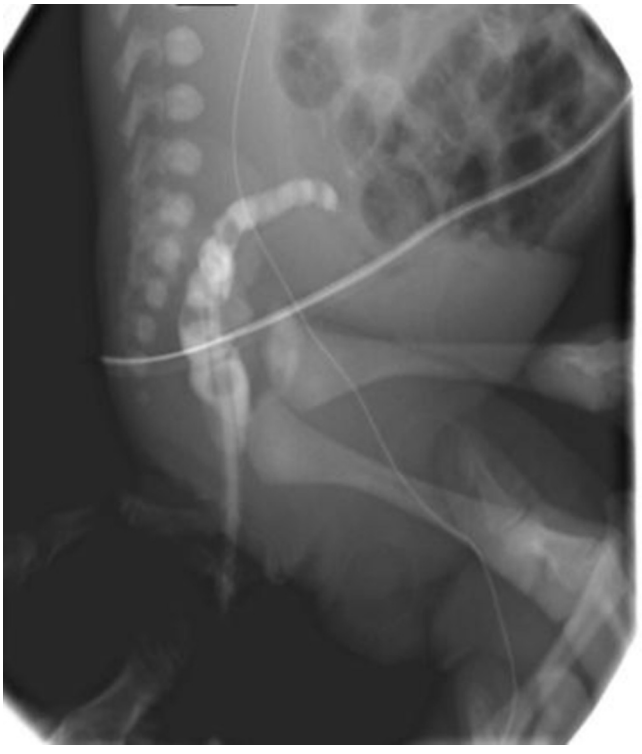

Fig. 2. Lateral contrast enemas demonstrating small filling defects and microcolon.

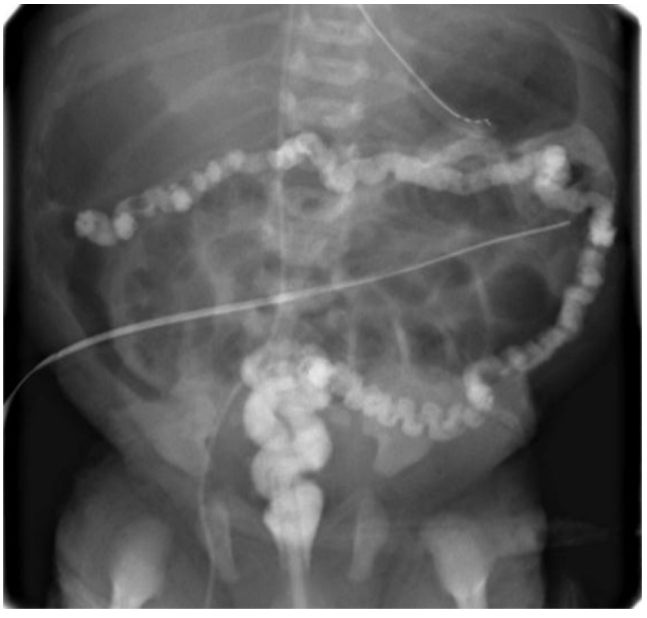

Fig. 3. Contrast enemas POD #4 demonstrating microcolon and free air suggestive of perforation.

Postoperatively the mucus fistula, nasogastric tube and ostomy were flushed with mucomyst to facilitate clearing the meconium obstruction.

Sweat chloride test and cystic fibrosis genetic mutation studies were sent and both came back confirmatory for cystic fibrosis. He was found to be homozygous for the delta-F508 mutation.

2. Discussion

Spontaneous bile duct perforation (SBDP) is a rare disease with less than 150 cases reported in the literature. Medline reveals no prior cases of spontaneous bile duct perforation associated with cystic fibrosis or meconium ileus. SBDP typically presents in early infancy but presentation can range anywhere from prenatal (25 weeks of gestation) to 7 years, with a peak incidence around 6

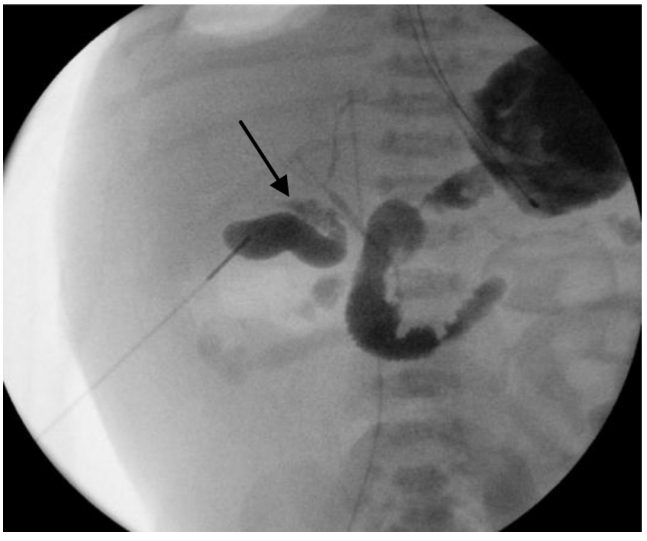

Fig. 4. Intraoperative cholangiogram via gallbladder during second operation showing bile leak arising from junction of cystic and common bile duct.

months of age [1]. The etiology is unknown, but likely variable in nature with suspected causes including: congenital weakness at the junction of the cystic and common bile duct, ischemia, viral infection, distal obstruction and pancreaticobiliary malunion [2]. The possibility of obstruction distal to the perforation, which most commonly occurs at the junction of the cystic duct and common hepatic duct, has been suggested in some cases, but healing without reconstruction suggests this is not common [2].

Typically, patients present with subacute symptoms such as fluctuating jaundice, pale or acholic stools, failure to thrive, vomiting and irritability [3,4]. Only 20% of patients present with acute onset severe abdominal distention, emesis and respiratory distress as was seen in our patient [3]. While elevated bilirubin and liver enzymes may be helpful in suggesting a diagnosis, labs may also be normal in some patients [1,2]. The most common initial diagnostic modality is ultrasound which identifies free abdominal fluid and usually dilation, obstruction or other abnormality of the biliary tree [2]. Ultrasound-guided paracentesis typically demonstrates either frank bile (as in our case) or ascites with a bilirubin level greater then the serum level [4].

Prompt drainage should be undertaken in most cases [3]. Drainage may be approached in several ways depending on the patient's clinical picture. Options include percutaneous drainage, laparoscopic drainage, laparotomy, internal drainage using endoscopic retrograde pancreaticoduodenoscopy (ERCP) with biliary stent placement, or percutaneous transhepatic cholangiography with stent placement [3,5,6]. Formal repair of the biliary leak is only performed in selected cases, as small perforations typically seal. Repair of an inflamed, small duct in a newborn may not be not be feasible [5–7]. In the rare cases of ductal obstruction or severe disruption, repair options depend on the clinical situation [5–7]. Described techniques include suture closure with or without a t-tube, cholecystostomy and drainage, transhepatic biliary stent placement, ERCP with biliary stent placement and roux-en-y reconstruction if a large perforation is encountered [5–7]. The goal of treatment is to confirm patency of the distal bile ducts and to provide adequate biliary drainage. Intraoperative cholagiogram should be done to evaluate for distal obstruction or biliary abnormality indicating the need for biliary reconstruction [6]. If distant obstruction is not identified, laparoscopic drainage alone is adequate and even preferable in some cases [1–7]. Without repair and no evidence of distal obstruction, bile duct leaks have been shown to close between 14 and 24 days [2].

Previously reported conditions associated with SBDP include choledochal cysts, necrotizing enterocolitis, biliary atresia, and portal vein thrombosis (possibly secondary to chemical irritation and inflammation around the vein) [8]. SBDP, however, has never previously been linked to cystic fibrosis or meconium ileus [4,7]. While patients with long standing CF may have biliary complications associated with their disease including cholestasis and strictures leading to cirrhosis and portal hypertension [9], SBDP has not been reported. Liver disease in older CF patients has been attributed to several factors, one of which is abnormal regulation of membrane chloride channels leading to thick secretions and hepatic congestion of bile, although predominantly intrahepatically [10]. The precise etiology of this patient's biliary perforation is unclear.

3. Conclusion

Spontaneous bile duct perforation in infants is a rare condition. Most cases are associated with neonatal jaundice and a subacute course, but fulminate presentations such as this case do occur. The pathophysiology of SBDP is unknown. The association of SBDP with meconium ileus should alert the clinician to possible multiple concurrent surgical processes in these rare cases. While SBDP may often be managed with drainage alone, the association with meconium ileus required additional surgical intervention in this case. While this is the first reported case of SBDP in a patient with meconium ileus and cystic fibrosis, the potential for a relationship between meconium ileus and SBDP must be considered in similar clinical settings.

Disclosure statement

The authors have nothing to disclose.

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