Spontaneous bile duct perforation in an infant, managed with simple drainage: a case report

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Spontaneous bile duct perforation is a very rare but important cause of surgical jaundice in pediatric patients and one of the most common causes of surgical jaundice during infancy after biliary atresia. Preoperative diagnosis may not be possible in most of the cases. The exact cause of the perforation remains unclear. The diagnosis is usually made at the time of laparotomy. We report a case of spontaneous bile duct perforation in an infant and showed the successful minimal surgical intervention with T-tube insertion as the sole management. ***Ann Pediatr Surg*** **11:153–155 © 2015 Annals of Pediatric Surgery.**

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Introduction

Idiopathic perforation of the nondilated extrahepatic bile duct is rare. Since its first description by Caulfield [1], more than 150 cases have been reported in the literature. The presentation ranges from obstructive jaundice to biliary peritonitis. The diagnosis is usually made at the time of laparotomy, except if there is a high index of suspicion [2]. The management ranges from simple drainage to biliary diversion. We report a case of spontaneous bile duct perforation (SBDP) in an infant and showed the successful management with T-tube drainage.

Case report

An 80-day-old female infant was referred to our hospital from a maternity and children's hospital as a case of obstructive jaundice (with a suspicion of choledochal cyst). She presented at the age of 7 weeks with jaundice, dark urine, and pale stool, but with no incidence of history of fever, diarrhea, or feeding problems. She was the result of a full-term, vaginal delivery, after an uneventful pregnancy. Clinically, she suffered from jaundice but was active and looked well. The abdomen was distended but soft and lax, with no evidence of peritonitis. The liver was 2 cm below the costal margin with sharp edge. Her stool was clay colored. Blood workup showed hemoglobin of 9.7, white blood cell of 14.2, platelet count of 664, total bilirubin of 156 μmol/l, direct bilirubin of 133, alkaline phosphatase of 701, γ-glutamyltransferase of 2257. Ultrasound examination showed dilated common bile duct (CBD) with suspicion of choledochal cyst (Fig. 1). Hepatobiliary scintigraphy showed good liver uptake and excretion with a dilated CBD, which looked similar to a choledochlal cyst. Magnetic resonance cholangiopancreatography showed the same findings. The impression was either a choledochal cyst or SBDP. The latter was more likely, given the history of the patient, the imaging studies, and reviewing the literature. Exploration laparotomy was performed, and it revealed large perforation of the CBD just distal to the cystic duct insertion with inflamed edges. The perforation was contained and localized around the CBD (Fig. 2). It was seen on ultrasound as dilated CBD, although it was in fact a contained perforation. There was no evidence of spillage of generalized peritonitis. A size of 7 Fr T-tube was inserted, and intraoperative cholangiography showed free flow of contrast to the duodenum and demonstrated the biliary tree, which was of normal diameter of 3 mm.There was no evidence of any CBD stone or distal biliary obstruction (Fig. 3). After a generous wash, a drain was left in the porta hepatis, in addition to the T-tube. Postoperatively, she recovered well, with return of liver function to normal and resolution of her jaundice. The T-tube was removed on the 10th day after demonstration of normal T-tube cholangiography and normal CBD diameter of 3 mm. She was discharged home in good health. Upon follow-up in the clinic, she was clinically jaundice-free with normal liver function. After 2 years she remains the same. Follow-up ultrasound examination showed normal biliary anatomy and CBD of normal caliber.

Discussion

Spontaneous biliary perforation is a rare entity in children. It is the second most common cause of surgical jaundice after biliary atresia. More than 150 cases have been reported in the literature. The peak age of occurrence was around 6 months, but it ranges from 25 weeks gestation to 7 years [2]. The exact etiology is poorly understood, but congenital mural weakness at the junction of the cystic duct and the CBD makes it the most common site, as it was in our case. Transient increase in intrabiliary pressure causing a 'blowout' is a possibility. Congenital embryonic weakness of malformation of the bile duct wall and ischemia of the CBD because of thrombosis in the microcirculation are also put forward as etiology. Associated factors include inspissated bile, necrotizing enterocolitis in the context of neonatal susceptibility to ischemia, distal bile duct stenosis in a pre-existing choledochal malformation, and anomalous pancreatobiliary junction with long common channel causing pancreatitis [3,4]. In cases of posterior perforation of the bile duct or hepatic duct, the diagnosis of the exact site is difficult. Patients with AIDS or lymphoma on chemotherapy can also develop SBDP. The clinical presentation is usually insidious with increasing abdom-

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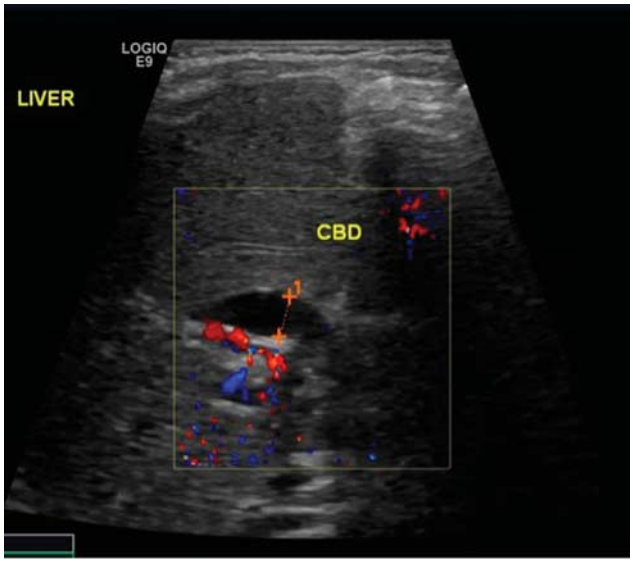
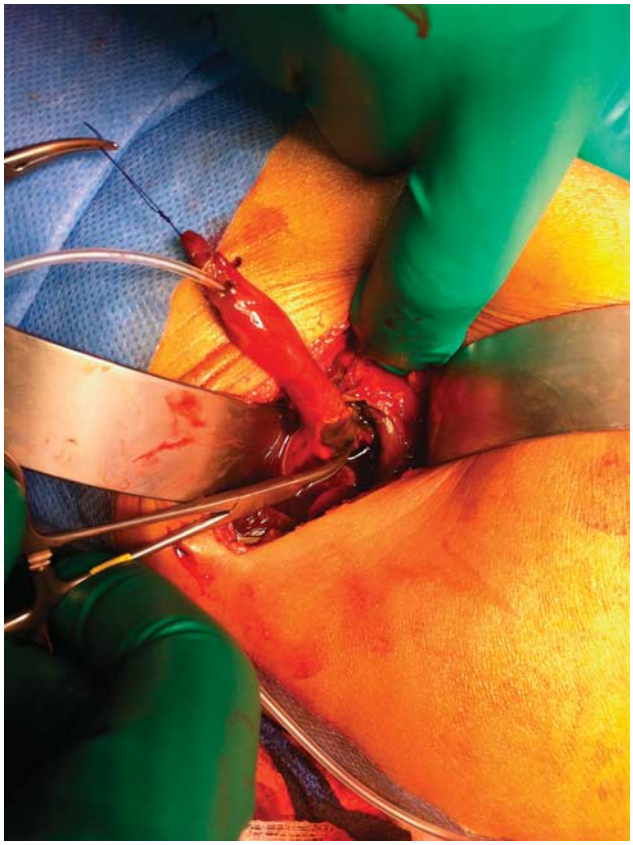
Fig. 1



Ultrasonography on the upper outer quadrant of the abdomen showing biliary leak.

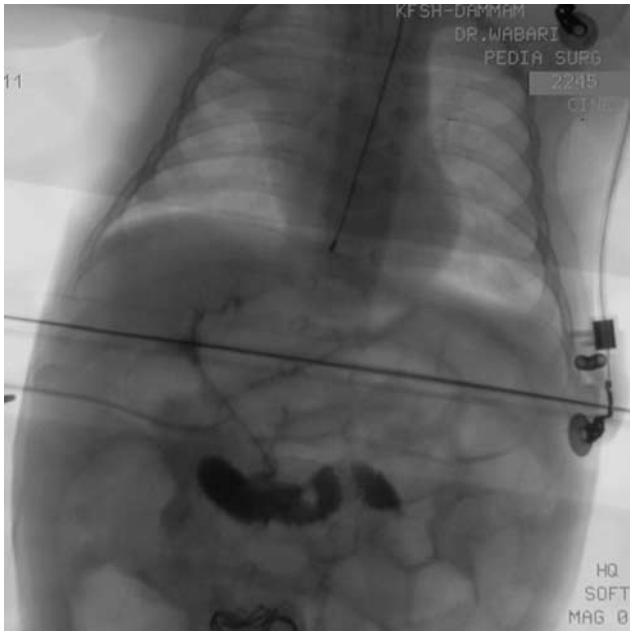
Fig. 2



Site of perforation.

inal distention due to biliary ascites, jaundice, and pale stool. Acute presentation without jaundice is rarer. The triad of bilious abdominal paracentesis, peritonitis, and absent free gas on radiography indicates SBP. Biliary scintigraphy is a good investigation tool [5,6]. Perforation of a pre-existing choledochal cyst has to be ruled out with either biliary scintigraphy or magnetic resonance cholangiopancreatography [7,8].

Fig. 3



Contrast study confirming biliary leakage.

Management is usually tapered to each case [9,10]. It ranges from simple drainage of bile to optional sphincteroplasty with or without primary closure of perforation, which is also an option. Bilioenteric diversion is controversial as a primary procedure as simple drainage with a T-tube may be sufficient. It may be indicated when perforation is large, or when distal obstruction cannot be corrected [10]. The incidence of bile duct stricture after simple drainage is not known; hence, the controversy exists whether to perform simple drainage or biliary diversion [11]. It seems that simple biliary drainage is a good primary option, but follow-up is mandatory to rule out development of stricture as the perforation heals [12]. Further studies are required to show the long-term results of simple biliary drainage [13].

Conclusion

SBDP is a very rare but important cause of surgical jaundice in children. High index of suspicion is required. Conservative management with simple drainage is usually sufficient.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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