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Case Report

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Spontaneous bile duct perforation in a toddler: A rare case

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Abstract

Spontaneous bile duct perforation is rare in all periods of life. It is rarely detected and managed preoperatively. Some of causes are congenital malformations, cholelithiasis, obstruction, and operation. Clinical presentation isn't specific and the management should be based on patient's condition. We hereby report a case of a 3-year-old girl with spontaneous bile duct perforation. She admitted with abdominal tenderness, fatigue, loss of appetite and episodes of vomiting. Radiologic images revealed intra and extra hepatic bile duct dilation with free intraperitoneal fluid accumulation without any extrahepatic obstructive lesion, and which was managed conservatively. Effective management of this condition requires an awareness of its symptoms and radiological findings, appropriate investigation to treat symptoms and site of perforation.

Keywords: spontaneous, biliary duct, perforation, dilatation

1. Introduction

Spontaneous bile duct perforation (SBDP) is a rare clinical occurrence and can be seen in all periods of life. Conditions such as ischemia, congenital malformations, bile tree weakness, cholelithiasis, infection, obstruction, trauma, operation can predispose (1-6). Clinical presentation isn't specific which ranges from acute abdominal symptoms to fever, jaundice, acholic stool (1,3,5,6,7). Early diagnosis is essential as it is associated with high mortality. We report herein a case of spontaneous bile duct perforation in a previously healthy child which was managed conservatively.

2. Case report

A 3-year-old girl presented to the pediatric emergency department with a 3 days history of abdominal pain, fatigue, loss of appetite, and repeated episodes of vomiting. On physical examination, she had abdominal tenderness and symptoms of severe dehydration. Her past medical history was unremarkable. Initial blood investigations demonstrated elevated levels of white blood cells (15,57 $\times 10^3/\mu L$), alanine transaminase (474 U/L), aspartate transaminase (145 U/L) amylase (350 U/L), lipase (527,7 U/L), C-reactive protein (20,52 mg/L), creatine phosphokinase (225 U/L) and gamma-glutamyl transferase (558 U/L).

An abdominal ultrasound (US) was performed subsequently. On US, intra and extrahepatic bile duct dilatation with gallbladder sludge was seen without any extrahepatic obstructive lesion. Magnetic resonance cholangiopancreatography (MRCP) was performed and revealed that intra and extrahepatic bile ducts were very dilated with an abrupt ending of the choledochal duct in the middle part without any obstructive cause (Fig.1). There was massive free intraperitoneal fluid accumulation accompanied by diffuse peritoneal haziness suggesting peritonitis. Moderate bilateral pleural effusion was also noted (Fig. 2).

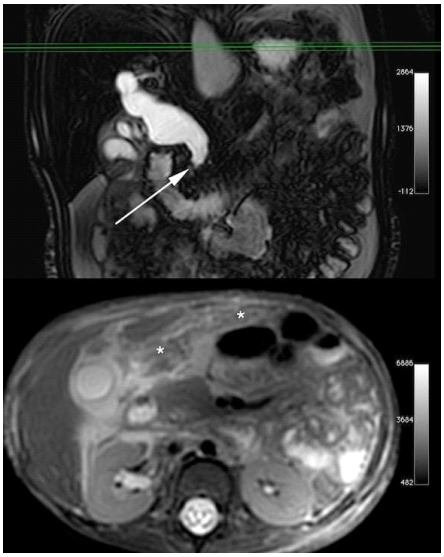

Fig.1. A 3-year-old girl with SBDP.MRCP shown extrahepatic bile duct dilation (long arrow), Axial T2- weighted images reveal peritoneal thickening (*)

The absence of a biliary obstructive cause and findings of peritonitis suggested the diagnosis of SBDP and associated biliary peritonitis. Subsequent peritoneal fluid aspiration under US guidance yielded bilious fluid and biochemical fluid examination confirmed the diagnosis which showed elevated total bilirubin (11.66 mg/dl bilirubin), amylase (2946 U/L), and lipase (12437 U/L) levels, and elevated white blood cell count (40.649 cells/μL). Pediatric surgery was consulted for the

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patient's condition, but surgery was not considered, and a conservative treatment strategy that included broad-spectrum antibiotics, ursodeoxycholic acid, octreotide was preferred. During hospitalization, an endoscopic retrograde cholangiopancreatography (ERCP) was performed, which showed intrahepatic bile duct and choledochal dilatation. A biliary sphincterotomy was carried out and yielded debris. During the follow-up with conservative treatment, the clinical condition of the patient improved and radiological findings like biliary dilatation and findings of peritonitis regressed on control MRCP.

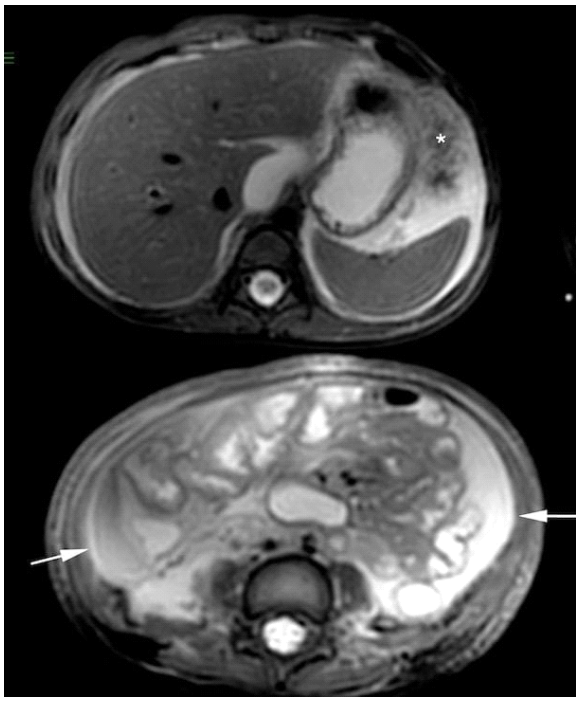

Fig.2 Axial T2- weighted images reveal peritoneal thickening (*) and intraperitoneal free fluid suggesting perforation(arrows)

The patient was then discharged and did not have any complaints in the first year follow-up.

3. Discussion

Perforation of the biliary tree without trauma or iatrogenic injury is very rare (1,2,4,5). The pathogenesis of spontaneous perforation of the bile duct is poorly understood but there are some postulated mechanisms to clarify: intramural infection that decreases the residence of duct wall; increased intraductal pressure of the sphincter of Oddi due to reflux spasm or obstruction of stones, or both; congenital weakness; congenital malformations such as choledochal cysts and biliary diverticulum; necrotizing enterocolitis; iatrogenic causes; delivery trauma; thrombosis of a mural vessel causing to necrosis of affected area of bile duct wall; reflux of pancreatic secretions that results in autodigestion, lymphoma, chemotherapy, AIDS (1-6,8).

Preoperative diagnosis isn't possible in most cases and the exact cause of the perforation remains unclear as in our case. In our case, MRCP findings were suggestive of the diagnosis although perforation itself was not seen apparently and aspiration of peritoneal fluid confirmed the diagnosis. According to a study, the meantime is 4.2 days between the onset of symptoms and the diagnosis of bile leak (7). Our patient was diagnosed four days after the onset of symptoms.

Spontaneous biliary perforation is highly associated with inflammatory reactions and peritonitis. The triad of peritonitis, bilious abdominal free fluid on paracentesis without free gas on radiograph suggests SBDP (8). According to a systematic review that revealed 60 cases of SBDP in infants and children, the most common presentation was abdominal distention and/or jaundice and the median age was 4 months (2). Other clinical symptoms included findings of acute abdomen, abdominal distention, ascites, acholic stool, non-bilious vomiting, dark urine, fever, lethargy, inguinal, umbilical hernia (1-8). Our patient also had nonspecific clinical symptoms.

Early diagnosis is very important and radiological findings are very helpful to suggest the diagnosis. As the first-line imaging modality, abdominal US is the best option to evaluate the biliary system and can identify ascites. (2,3,4). MRCP better delineates the biliary ductal system and can show ductal and junctional anomalies owing to the inherent contrast of water in the bile. ERCP can be used for both the diagnosis and treatment, however, it is an invasive method and MRCP has replaced ERCP for diagnostic purposes.

Management of SBDP should be based on the patient's age, general condition, the severity of peritonitis, side of perforation, imaging findings. The management has 3 options: 1) nonoperative strategy with antibiotics and percutaneous drainage or ERCP; 2) surgical biliary drainage including external drains placed at the porta hepatis, cholecystostomy tube, and T-tube insertion; 3) biliary reconstruction via primary repair of the perforation or Roux-en-Y hepaticojejunostomy (3,9). If the patient is stable without distal biliary tract obstruction and symptoms of peritonitis, conservative treatment can suffice (6). Simple biliary drainage is an advisable primary step, but follow-up is essential to rule out the prognosis of stricture while the perforation heals (8,9). ERCP can be used for biliary stenting for strictures and perforation, pancreatic and biliary drainage, bile duct stone removal, balloon dilation of the duodenal papilla or ductal strictures, sphincterotomy in cases of the sphincter of Oddi dysfunction or stenosis, biliary tissue sampling from the pancreaticobiliary ductal system, (1,9,10). Surgery may be indicated when a distal obstruction, cholelithiasis, congenital anomaly, gallbladder perforation, cystic duct perforation occurs.

In conclusion, SBDP is a rare and rather unknown entity with nonspecific symptoms and clinical findings. Awareness of the condition along with radiological findings can raise the suspicion for diagnosis and prevent serious consequences. Although there is no definitive care for the perforation, conservative management can be a successful treatment option.

Conflict of interest

The authors declared no conflict of interest.

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Authors' contributions
Concept: E.K, İ.Ç., Design: E.K, İ.Ç., Data Collection or Processing: E.K, İ.Ç., Analysis or Interpretation: E.K, İ.Ç., Literature Search: E.K, İ.Ç., Writing: E.K, İ.Ç.

Ethical Statement
Ethic committee permission is not required for this study.

References

1. Duminda Subasinghe, Edippuli Arachchige Don Udayakumara, Upul Somathilaka, Milinda Huruggamuwa, "Spontaneous Perforation of Common Bile Duct: A Rare Presentation of Gall Stones Disease", Case Reports in Gastrointestinal Medicine, vol. 2016, Article ID 5321304,3pages,2016. https://doi.org/10.1155/2016/5321304
2. Evans K, Marsden N, Desai A. Spontaneous perforation of the bile duct in infancy and childhood: a sys tematic review. J Pediatr Gastroenterol Nutr. 2010 Jun;50(6):677-81. doi: 10.1097/MPG.0b013e3181d5eed3. PMID: 20400907.
3. Leslie Hopper, Scarlett B.Hao ,David Rodeberg,ShannonLongshore.Spontaneous bile duct perforation in a neonate. Journal of Pediatric Surgery Case Reports. Volume 38, November 2018, Pages 57-60
4. Kobayashi, Katsutoshi, et al. Bile peritonitis due to spontaneous perforation of the left hepatic duct: A case report. Japan Medıcal Associatıon Journal 48.8 (2005): 422.
5. Sharma, S.B., Sharma, S.C. & Gupta, V. Spontaneous biliary perforation: a rare entity in late infancy and childhood. Indian J Pediatr 70,829–831(2003). https://doi.org/10.1007/BF02723808
6. Mohamad Halabi,Neil Merritt,Dhandapani Ashok, Emergency case of spontaneous common bile duct (CBD) perforation. Canadian Liver Journal, Volume 1 Issue 4, Fall 2018, pp. 256-260
7. McLindon, J., England, R. & Martin, D. Causes, clinical features and non-operative management of bile leaks. Eur Radiol 8, 1602–1607(1998). https://doi.org/10.1007/s003300050595
8. Alwabari, Abdullah; Abbas, Gammal Spontaneous bile duct perforation in an infant, managed with simple drainage, Annals of Pediatric Surgery: April 2015 - Volume 11 - Issue 2 - p 153-155 doi: 10.1097/01.XPS.0000462926.99853.97
9. .C. Jeanty, S.C. Derderian, S. Hirose, H. Lee, B.E.-Padilla Spontaneous biliary perforation in infancy: management strategies and outcomes-J Pediatr Surg, 50 (2015), pp. 1137-1141.https://www.sciencedirect.com/science/article/pii/S0022346814004758
10. Meseeha M, Attia M. Endoscopic Retrograde Cholangiopancreatography. [Updated 2020 Aug 11]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2021 Jan-.Availablefrom: https://www.ncbi.nlm.nih.gov/books/NBK493160/