International Journal of Medical Science in Clinical Research and Review
Online ISSN: 2581-8945
Available Online at http://www.ijmscrr.in Volume 7|Issue 01 (January-February)|2024 Page: 111-113

Case Report

Spontaneous Bile Duct Perforation: Two Case Reports and a Literature Review

Authors:
Reda Belbouab, Amina Khabtani, Hayet Benalioua, Rachida Boukari
Mustapha Bacha University Hospital Centre, Pediatrics Department Algiers, Algeria
Corresponding Author:
Reda Belbouab, Mustapha Bacha University Hospital Centre, Pediatrics Department Algiers, Algeria

Article Received: 30-December-2023, Revised: 19-January-2023, Accepted: 08-February-2024

ABSTRACT:
Spontaneous perforation of the biliary ducts is a rare disorder in infants. Early diagnosis of this entity is important because it can be treated surgically. The authors report on two patients with spontaneous biliary perforation who were treated during the year 2020 and 2022. The diagnosis was made in two children aged 2 and 4 months presenting progressive abdominal distension associated with jaundice, acholic stools and elevated bilirubin in the ascites fluid. Surgical exploration by laparotomy confirmed biliary duct perforation. Cholecystectomy were performed including a suture of the perforation in one patient and an external surgical drainage in the second. Perforation of the bile ducts should be suspected in every neonate and infant with ascites and jaundice. Confirmation is obtained by paracentesis and bilirubin measurement in the ascitic fluid or by combination of sonography and scintigraphy.

Keywords: cholestasis, *spontaneous perforation of biliary duct, biliary ascites, Surgery, Infancy*

INTRODUCTION:

Spontaneous bile duct perforation (SBDP) is a pathological entity in which the wall of the extrahepatic or intrahepatic bile ducts is perforated without any traumatic injury. Although a very rare condition, it is the second most common cause of jaundice requiring surgical correction in early childhood, after biliary atresia. Occurring during early childhood and mainly in infants, it can manifest acutely as biliary peritonitis or subacutely as cholestasis secondary to stenosis at the initial site of the perforation. Its diagnosis remains difficult and it can be pre- or intraoperative. The rarity of this condition prompts us to report two clinical cases documented in our department in the years 2020 and 2022.

CASE REPORTS:

CASE: 1.

A 4-month-old girl, born at term to consanguineous parents, without significant medical history, who presented with progressive abdominal distension associated with jaundice, acholic stools and dark urine over the last two months. On examination, positive findings included: generalized jaundice with pallor, a tight and distended abdomen, with shiny skin, visible veins and palpable umbilical and inguinal hernias: both reducible and painless. Laboratory results included an elevated total bilirubin of 34 with a conjugate value of 32 mg/l, serum alkaline phosphatase level of: 617 U/I and GGT of 803 U/l, Serum liver enzymes were within normal limits (AST is 49 and ALT is 16 IU/l). Abdominal ultrasound revealed: massive non-septate ascites, without organomegaly or signs of PH.

A paracentesis was performed, and the aspirated liquid was found to have a xanthochromic appearance with a total bilirubin of 162g/l with a conjugate value of 96g/l, thus confirming the diagnosis of bilious ascites. The child was referred for surgery, the decision was made to perform an ascitic tap yielding approximately 1200 ml of bile fluid. Followed by laparotomy through a right supraumbilical transverse incision, revealing extensive inflammatory adhesions at the level of the porta hepatis. After careful dissection and minimal release, exploration identified a non-distended gallbladder embedded in the liver. Subsequently, a warm saline injection into the common bile duct (CBD) was performed, exposing a perforation adjacent to the porta hepatis. The perforation was quickly sutured with two stitches. Cholecystectomy, including removal of the cystic duct, was performed. Peritoneal washing and closure with lamellar drainage

were performed. Postoperative convalescence was uneventful. After 1 month, 9 months and 3 years of follow-up, the patient was in perfect health.

CASE: 2.

A 2-month-old male infant, born at term to consanguineous parents, without any particular pathological history, presented with progressive abdominal distention and cholestasis lasting 4 weeks. On examination: the infant was hemodynamically stable, well hydrated, afebrile, slightly icteric and pale, the abdomen was distended, the liver and spleen could not be assessed clinically. Laboratory studies revealed anemia (hemoglobin level of 9.5 g/dL) and mild cholestasis (serum bilirubin level of 71 with a conjugate value of 43 mg/l, serum alkaline phosphatase level of 640 IU /L), liver enzymes and other parameters were within normal limits. An abdominal ultrasound revealed the appearance of a loculated peritoneal effusion with septations and hyperechoic images within, a liver and spleen of normal size and echo structure. Paracentesis yielded bilious fluid with a total bilirubin of 321 mg/l with a conjugate value of 207 mg/l, confirming its bilious nature. The infant was referred for surgery for laparotomy, upon opening a copious amount of bilious fluid was aspirated, a sample was sent for laboratory examination, exploration found a liver and gallbladder of normal appearance, instillation of warm saline solution via the gallbladder confirmed biliary puncture perforation at the level of the bile duct just below the hepatocyst junction, Bile ducts appeared impermeable: possible stenosis, requiring further examination, drainage External surgical procedure was performed without suture with cholecystectomy, the patient may need a hepaticojejunostomy possibly depending on postoperative follow-up. The immediate postoperative period was clinically uneventful, with a gradual decrease in the volume of fluid drained and a normalization of the color of urine and stools. During follow-up after 1 and 3 months, the infant was asymptomatic with strictly normal biological results, US showed dilation of the common bile duct with a diameter of 1.3 mm and 2.6 mm respectively.

DISCUSSION:

Spontaneous perforation of the bile ducts is a rare pathology, with an incidence of 1.5 per 1,000,000 live births and can be observed from 25 weeks of gestation up to the age of 7 years with a median age of 2. 4 months according to recent literature reviews [1,2]. The disease usually manifests in previously healthy infants with unremarkable prenatal and postnatal histories [3]. The elective site of perforation is the anterior wall of the common bile duct near its junction with the cystic duct, as in our second case. At present, the etiopathogenesis of SBDP remains unclear, and it is likely multifactorial, and may include the following conditions in isolation or in combination: congenital weakness of the biliary wall due to ischemia of the anterior common bile duct related to a posterolateral arterial supply, a pancreaticobiliary malunion near the sphincter of Oddi which would allow reflux of pancreatic enzymes into the bile duct, leading to acute inflammation and biliary obstruction with increased intraluminal pressure causing perforation, Moore et al. suggested that SBDP appears to exist on a continuum with choledochal cyst as a form of acquired biliary atresia [4].

Clinically, the presentation of SBDP in children can be acute or insidious, the latter being the most common, accounting for 80% of cases. The insidious form presents in the form of chronic biliary ascites with fluctuating cholestasis composed of mucocutaneous jaundice often mild to moderate, dark urine and normal or acholic stools, progressive painless abdominal distention, often associated with anoraexia, growth retardation, fever and the development of umbilical, inguinal or scrotal hernias and hydroceles. Acutely, there may be fever, vomiting, abdominal pain and signs of severe biliary peritonitis [7], sepsis and shock. Preoperative diagnosis in this case is much more difficult because jaundice is rarely present and liver function may be strictly normal.

Laboratory evaluations are not pathognomonic. Conjugated bilirubin and alkaline phosphatase levels may be elevated, hepatic transaminase levels are often normal.

Abdominal ultrasound shows free or loculated peritoneal effusion and there is usually no pneumoperitoneum on erect abdominal X-ray [5].

Paracentesis is an extremely useful tool in resource-limited settings, as it is the case in our country. An ascitic tap yielding xanthochromic ascitic fluid with bilirubin levels that are elevated above the normal range and above the serum bilirubin level would be highly suggestive of a biliary leak. The normal bilirubin level in ascitic fluid is 7 to 8 mg/L. An ascitic fluid bilirubin concentration of > 60 mg/L is highly suggestive of choleperitoneum [6].

Multiple radiological tools can be used to document the presence of a biliary leak and its site in order to confirm the diagnosis and guide the treatment. Hepatobiliary scintigraphy (HIDA scan) is recommended as it is highly sensitive and specific for SBDP and is virtually diagnostic when it shows extravasation into the peritoneal cavity with no activity in the duodenum [4], it is also useful in the follow-up of treated patients. CP-MRI has gained recognition in recent years as an interesting diagnostic tool. While endoscopic retrograde cholangiopancreaticography (ERCP) has an additional

interventional advantage, as stenting can be performed, and pancreatitis can simultaneously be ruled out [3].
Early treatment combined with adequate preoperative resuscitation and IV antibiotics reduce morbidity and mortality. Intraoperative cholangiography should always be performed to determine the location of the perforation, detect any ductal abnormalities, and exclude distal obstruction. In the management of SBDP, three interventions have been advocated: non-operative management with antibiotics and drainage, percutaneous or ERCP; surgical biliary drainage utilizing external drains, a cholecystostomy tube, or a common duct T-tube; biliary reconstruction via primary repair of the perforation or Roux-en-Y hepaticojejunostomy [4].
Both our cases had a subacute onset, with chronic biliary ascites and mild cholestatic jaundice. Ultrasound and laboratory evaluations were none-specific. Paracentesis was highly suggestive of chole-peritoneum. The diagnosis was, however, confirmed intra-operativaly and surgical biliary drainage with a cholecystectomy was chosen for both patients with good post-operative follow-up.[7]

CONCLUSION:

Awareness of this fairly rare condition will make it possible to consider it in the face of any picture of cholestatic jaundice associated with progressive abdominal distention in previously healthy infants, in order to make an early diagnosis and allow adequate surgical management. The common postoperative complications include stenosis of CBD and portal vein thrombosis rendering long-term follow-up with serial ultrasound essential. Overall, prognosis remains excellent with prompt diagnosis and adequate treatment.

Disclosure:

The authors declare no competing interest.

Authors' Contributions:

All authors contributed equally to the write up of this article. They also read and approved the final version of the manuscript.

REFERENCES:

1 - Journal of Pediatric Surgery Case Reports - Volume 38, November 2018. Spontaneous bile duct perforation in a neonate. Leslie Hopper, Scarlett B. Hao, David Rodeberg, Shannon Longshore.

2 - Journal of Pediatric Surgery Case Reports - Volume 39, December 2018. Spontaneous biliary perforation in a 3-month old. Vallery Logedi, Erik N. Hansen.

3 - Malays J Med Sci. Jan-Mar 2012. Management of SPBD in an Infant in a Semi-Urban Setup: A Case Report. Satish Jain, Monica Jain, Dalbir Kaur, Loves ShuKla.

4 - Journal of Pediatric Surgery Case Reports - Volume 38, November 2018. Spontaneous bile duct perforation in a neonate. Leslie Hopper, Scarlett B. Hao, David Rodeberg, Shannon Longshore.

5 - Journal of Pediatric Surgery Case Reports - Volume 77, 2022. Spontaneous perforation of the cystic duct in an infant. Workye Tigabie, Goytom Knfe, Tesfay Yeneneh Yirga.

6 - Frontières de la pédiatrie. Volume 10, mars 2022 - Analyse des caractéristiques cliniques de la perforation spontanée des voies biliaires chez l'enfant. Xueqiang Yan, Nannan Zheng, Jinfu Jia, Houfang Kuang, Haiyan Lei, Hongqiang Bian, Xinke Qin, Xuan Sun, Xufei Duan et Jianghua Zhan Département de chirurgie générale, Hôpital pour enfants de Wuhan

7 - Journal of Pediatric Surgery (2012), Prise en charge conservatrice de la perforation spontanée des voies biliaires chez l'enfant : étude de cas et revue de la littérature. Maria Virginia, Pereira E Cotta, Jennifer Yan, Mina Asaid, Peter Ferguson, Thomas Clarnette.