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Review Article

Paediatric Surgery Section

Spontaneous Biliary Perforations: An Uncommon yet Important Entity in Children

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ABSTRACT

Spontaneous or idiopathic biliary perforations are an infrequently encountered but an important cause of surgical jaundice in paediatric patients and one of the most common causes of surgical jaundice in infancy. A pre-operative diagnosis with a clinical history and physical findings may not be possible in most of the cases. The exact cause of the perforation remains unclear and the diagnosis is made at the time of laparotomy for an acute abdomen. An early, efficient and an effective surgical management is associated with a good prognosis; however, a delay in the correct diagnosis or an inappropriate management may result in bacterial contamination of the biliary ascites, with an unfavourable outcome. The relative rarity of this condition is reflected by the very few case reports, limited case studies and scarcity of published literature.

Key Words: Spontaneous biliary perforation, Idiopathic biliary perforation, Gall bladder perforation, Common bile duct perforation, Biliary ascites, Choledochal cyst perforation

INTRODUCTION

Spontaneous biliary perforations and the resulting biliary ascites, though rare, are known to affect the extra hepatic biliary tree and occasionally, the intrahepatic ducts. They are in fact, one of the most common causes of surgical jaundice in infancy, the most common being biliary atresia [1]. The exact cause remains unclear in majority of the cases. Caulfield reported the first two cases of biliary peritonitis in infancy. He opined that severe trauma was responsible for all the previous reports. A limited number of cases have been reported thereafter. They are an unusual cause of acute abdomen, are difficult to diagnose pre-operatively and have a potential for a high morbidity and mortality. The rarity of this condition is reflected by the very few case reports, limited case studies and scarcity of published literature. This disorder is infrequently listed as a cause of obstructive jaundice and it may not be recognized till the time of laparotomy.

A literature search was made from Pubmed with the key words, 'spontaneous biliary perforation' and all the available articles were retrieved and analyzed.

EPIDEMIOLOGY

Spontaneous perforation of the common bile duct is commonly encountered in children who are under the age of four years [2,3]. The peak incidence is seen during the first year of life. Banani et al., [4] suggested that the peak age of occurrence was around 6 months of age and that it could range from 25 weeks of gestation to 7 years of age. Both the cases which were reported by Caulfield were full term male infants who were under the age of six months. Spontaneous perforation of the common bile duct and biliary peritonitis have been reported in infancy and as late as 50 months. The sex ratio is almost equal [5]. Perforation of the gall bladder is even rarer in the paediatric age group [6-9]. However, Mirza et al., [10] have paradoxically reported a higher incidence of gall bladder perforations in their series, as compared to the Common Bile Duct (CBD) perforations. They suggested that this might be related to the higher incidence of typhoid fever and its related surgical complications in the developing world. Besides this, typhoid is also known to cause intestinal perforations, bleeding, cholecystitis and pancreatitis [11].

ETIOLOGY AND PATHOPHYSIOLOGY

The aetiopathogenesis of spontaneous biliary perforations is still obscure. Various theories have been proposed to explain the aetiology of biliary peritonitis and they have been summarized in [Table/Fig-1]. Upto 7% of the children with choledochal cysts in the larger series present with spontaneous ruptures [12, 13]. The

Perforation of the Common Bile Duct

- Mural embryopathy/congenital weakness of the wall of the common bile duct
- Congenital anomalies of the pancreaticobiliary junction
- Pancreatitis
- Trauma
  - Delivery trauma
  - Road traffic accident
  - Iatrogenic injury
- Ascariasis
- Distal obstruction of the bile duct (? inspissated bile, etc.)
- Choledocholithiasis
- Stenosis of the common bile duct
- Presence of a diverticulum in the common bile duct
- Abnormal glands in the wall of the common bile duct
- Viral infection of the common bile duct [14]
- Tuberculosis
- Necrotizing enterocolitis
- Rarer causes
  - Erosion of CBD wall by tumor
  - Weakness by previous choledochostomy
  - Localized ischemia of CBD wall due to intramural thrombosis

Perforation of the Gall Bladder

- Calculous cholecystits
- Acalculous cholecystitis
- Typhoid fever
- Perinatal asphyxia and sepsis [15]
- Idiopathic or Spontaneous

[Table/Fig-1]: Factors related to Biliary Perforation

associated anomalous pancreatico-biliary junction and the mural immaturity which is due to infancy lead to constant or recurrent refluxes of the pancreatic enzymes into the common bile duct. The perforations of the cysts are probably related to the associated chronic inflammation which results from the epithelial irritation from the refluxed pancreatic juice; trauma or inspissated bile may be responsible for the perforations occasionally and an abnormal rise in the ductal pressure is seldom responsible. It has been suggested that what is considered as a spontaneous perforation of the Common Bile Duct (CBD) may actually be related to the underlying pancreatico-biliary malunion [4]. However, it is not possible to ascribe all the cases of spontaneous perforations to the same aetiology. This is because most of these cases present for the first time with a biliary problem, the size of the choledochal cyst is small and it gets collapsed after the perforation [16].

The relatively high osmotic pressure of the escaping bile leads to a fluid shift from the various peritoneal and intestinal tissues into the peritoneal cavity. The perforation is usually seen as a punched out hole in the common bile duct. The most common site of the perforation is the anterior wall of the common bile duct which is adjacent to its junction with the cystic duct. This site has also been suggested to be naturally prone to mural malformations during embryogenesis [13]. In the cases of underlying choledochal cysts, the most common site of the perforation is controversial. Livesey et al., [17] commented that the anterior wall of the choledochal cyst was the most common site of the perforation and they described two cases wherein the perforation was located in the posterior wall of the choledochal cyst (the junction of the common hepatic duct and the cystic duct were perforated in one of these). This resulted in a constrained leak which occurred preferentially into the lesser sac via the foramen of Winslow. The diagnosis was consequently delayed. Both the cases were also complicated by the development of portal vein thrombosis; one of them developed this several months after a surgery, while the other had portal vein thrombosis without any portal dissection. Some reports suggest that the usual location of the perforation is in the posterior wall of the choledochal cyst [16]. A majority of the case reports on spontaneous bile duct perforations are related to the perforations of the extra-hepatic ducts. Perforations of the intra-hepatic ducts have also been reported. The left intra-hepatic duct is more commonly involved than the right one; this is probably related to the thick liver parenchyma which surrounds the right intra-hepatic duct [18].

A spontaneous or an idiopathic perforation of the gall bladder is rare and the underlying mechanisms are unknown. Perforation of the gall bladder is usually a complication of acute cholecystitis. The reported incidence of gall bladder perforations in adults with acute cholecystitis ranges from 2-18%. Calculus cholecystitis is more commonly related to the subsequent gall bladder perforation as compared to acalculous cholecystitis; the reported incidence is 10-20% [19,20]. The gall bladder perforation following acute cholecystitis has been explained by various hypotheses: a) Bile stasis which occurs due to the obstruction of the cystic duct, dehydration or a total parenteral nutrition may lead to changes in the bile content and concentration; b) a vascular impairment of the gall bladder due to distention of the abdominal viscus as a result of sepsis or shock; and c) ischaemia, necrosis and perforation of the gall bladder wall [21].

The most common site of perforation of the gall bladder is the fundus. Anatomically, the fundus of the gall bladder is the least vascularized gall bladder region. Consequently, the vascular changes and ischaemia are probably the crucial factors in the pathogenesis of the perforation [22]. A gallbladder perforation may also occur at the neck of the gallbladder as a result of the pressure necrosis which is due to impacted stones and/or infection.

CLINICAL PRESENTATION

The clinical picture which is encountered in the cases with isolated traumatic perforations of the gall bladder is not pertinent to the gall bladder perforation. Following the initial injury, there are few symptoms, if any and they are relatively mild in intensity. There is little constitutional upset. The symptoms may regress soon and they are followed by a “period of illusion” in which the patients improve and may be discharged from the medical care. This occurs as there is egress of sterile bile into the peritoneal cavity, leading to chemical peritonitis in cases of blunt abdominal trauma which are associated with ruptures of the gall bladder. The presenting symptoms are usually benign and non-specific, unless there is a superimposed bacterial peritonitis. However, over a period of hours, days or even weeks, the patients gradually develop a progressively increasing abdominal distention, signs of peritonitis and jaundice, which may be associated with acholic stools. The diagnosis of a gallbladder perforation after a blunt injury may also be suspected in the patients with the signs of an acute abdomen and hypotension, which is not explained by blood loss.

Most of the patients who present with spontaneous perforations of the bile duct in infancy are typically previously healthy infants with unremarkable birth and perinatal histories. The presence of jaundice and acholic stools is often mistaken for extra hepatic biliary atresia. However, the clue is provided by the asymptomatic interval between the birth and the clinical presentation.

Chardot et al., [3] initially summarized that the spectrum of presentation of the common bile duct perforation can be grouped into three broad categories viz., acute peritonitis, localized peritonitis and with bile duct stenosis. However, the following four patterns of clinical presentations have become recognized over time.

Antenatal Presentation: Chilukuri et al., [23] reported the first case of an antenatal spontaneous perforation of the extra hepatic biliary tree in a 16 years old pregnant young lady. The sonogram at 16 weeks of gestation revealed the presence of ascites in the foetus. The postnatal hepatoiminodiacetic acid scan revealed a spontaneous perforation of the common bile duct after delivery at 32 weeks of gestation.

Contained Biliary Peritonitis: The presentation in this situation is classically subacute or chronic. The usual presenting symptoms include non-bilious vomiting, mild fluctuating jaundice, normal to acholic or clay coloured stools, poor weight gain or a failure to thrive, a gradual onset of ascites, irritability and occasionally and the development of inguinal or umbilical hernias. The hydrocele fluid or the umbilicus may be bile stained. Fever is usually absent and so are the signs of a peritoneal irritation.

Generalized Biliary Peritonitis: An acute onset of generalized biliary peritonitis is seen in 20% of the patients and it manifests itself as a distention of the abdomen, which is associated with pain and vomiting, which might be non-bilious initially or bilious from the outset, constipation or obstipation, high grade fever and ascites. Jaundice is usually absent or it may be mild. The

examination reveals varying degrees of dehydration which occur due to third space losses and a distended abdomen with signs of peritonism or peritoneal irritation. An abdominal mass may become palpable due to the formation of a pseudocyst. The laboratory evaluations are not pathognomonic. The liver function tests are usually normal or mildly deranged, although the serum conjugated bilirubin levels may be an exception. The alkaline phosphatase levels may be raised. While this type of presentation is encountered in every fifth patient of biliary perforation, biliary peritonitis is overlooked from the differential more- often- than -not and it is usually diagnosed at the time of laparotomy. Carubelli et al., [24] also reported an infant with a similar presentation. The child presented to the emergency department with an acute abdominal distention, ascites, haematochezia and shock, and was subsequently found to have a spontaneous perforation of the common bile duct.

Secondary Bile Duct Stenosis: Charcot et al., [3] described 5 cases in which perforations of the common bile duct were associated with stenosis and required biliary reconstructions. The seepage of bile into the intraperitoneal space leads to a severe inflammatory reaction at the porta hepatis and a possible collection of the inspissated bile around the common bile duct, resulting in stenosis or a complete occlusion. This presentation has also been reported by Davenport et al., [25].

THE DIAGNOSIS OF SPONTANEOUS BILIARY PERFORATIONS

Spontaneous biliary perforations account for a small number of cases of the whole spectrum of peritonitis. They are rarely diagnosed pre-operatively and are associated with a high risk of morbidity and mortality. Surgery is usually performed for diffuse peritonitis like acute appendicitis.

Ong et al., [26] reported six cases of gall bladder perforations in adult patients and the diagnoses in all of these could be made only at the time of laparotomy. The clinical manifestations in the patients with acute gall bladder perforations may be akin to acute cholecystitis. This leads to an incorrect diagnosis, a delayed diagnosis and the high morbidity and mortality which is associated with this condition. It is for this reason that Addison and Finan [27] advocated an 'urgent and early' cholecystectomy for acute gall bladder disease in a retrospective study which was done on 645 adult patients.

The presence of cholestatic jaundice with near normal liver function tests does not go a long way in making the diagnosis of a biliary perforation. Abdominal paracentesis may reveal the presence of bilious ascites with highly elevated bilirubin levels in the ascitic fluid. It has been suggested that the presence of bile in the peritoneal cavity, which is associated with obstructive jaundice in the absence of derangements in the liver functions, may be considered as pathognomic for the biliary perforation. The combination of mild fluctuant jaundice with acholic stools and the absence of urobilinogen in the urine are usually suggestive and indicative of the fact, that in the presence of normal bile production, the bilirubin is neither reaching the gut nor the blood stream in quantity but that it is escaping elsewhere. The abdominal distention with shifting dullness and the scrotal swellings which contain fluid suggest the site of the collection.

Imaging studies have limited use in the diagnosis of biliary perforations. Several studies have reported a high sensitivity in the preoperative identification of free intraperitoneal fluid or 'pericholecystic fluid' with the use of real time sonography in the patients with gallbladder perforations. The real time transducer can be manipulated to find the appropriate longitudinal section which is needed to demonstrate the site of perforation. Distention of the gallbladder and oedema of the walls may be the earliest signs of the impending gallbladder perforations which are detected on sonography. Once the perforation has occurred, the typical radiological signs include the 'hole sign' i.e., a defect in the gallbladder wall, which bears a striking resemblance to a defect in a perforated balloon or a pericholecystic fluid collection and the formation of an abscess [28]. The most specific finding of a perforation is the defect in the gallbladder wall, which has been described in 38.4% of the cases on USG and in 69.2% cases on CT Scan [29]. The features are related to the time which elapses between the perforation and the sonographic evaluation. Various conditions such as ascites, pancreatitis and peritonitis which occur due to other reasons are known to simulate gallbladder perforations on sonography. It has been suggested that scanning the gallbladder in the upright or the decubitus position may help in displacing the free ascitic fluid from the region of the gallbladder. The extrapancreatic fluid collection in case of pancreatitis may sometimes get accumulated in the right pararenal space instead of the left side and it may mimic a pericholecystic process. The gallbladder bed serves as a receptacle for the intraperitoneal abscess formation in cases of peritonitis.

In cases with spontaneous perforations of the bile duct, the ultrasound evaluation will show a free or loculated intraperitoneal collection, with normal intrahepatic and extrahepatic ducts [30].

The ability of a Computed Tomography (CT) scan to identify the gall bladder wall defects is limited by the need of an intravenous contrast medium to allow the evaluation of the gallbladder enhancement and the mural contour. Paracentesis may be performed under sonography or CT scan guidance.

Hepatobiliary scintigraphy can provide useful information about the liver function tests, patency of the biliary tree, the site of perforation and any free leak of the bile into the peritoneum. However, it lacks anatomical information and the radiation dose in not insignificant. It is relatively insensitive for the detection of subacute or chronic gall bladder perforations pre-operatively. In addition to this, it is almost impossible to perform this study without any prior suspicion of a gall bladder perforation. The cholescintigraphic findings in gallbladder perforations may be broadly grouped into the following four categories:

1. Direct visualization of the radionuclide extravasation from the gallbladder fossa during cholescintigraphy is pathognomonic of the perforation and it is probably the only method which can be used to establish the diagnosis pre-operatively. The pre requisite for a radionuclide extravasation is that the cystic duct should be patent and that the pressure within the gallbladder lumen should be less than the filling pressure in the biliary ducts.
2. The visualization of the gallbladder is unusual and it is related to 'resealing' of the perforation by inflammatory adhesions, without a concomitant return of the increased intraluminal pressure in the absence of a cystic duct obstruction [31].
3. Non visualization of the gall bladder is probably the most common observation but is not specific for a gallbladder perforation.

4. An indirect evidence of a gallbladder perforation [32] may be provided by the 'pericholecystic hepatic activity sign' or the 'photon deficient gallbladder fossa sign'.

For spontaneous bile duct perforations, hepatobiliary scintigraphy has been shown to be a highly sensitive and a specific modality and it is highly recommended when this pathology is suspected [33]. It has been suggested that in infancy, this is the most reliable method for visualizing the extravasation of bile into the peritoneal cavity from the biliary tract [34, 35]. The site of perforation may also be indicated by the accumulation of a radiotracer at the perforation site before the peritoneal soiling.

Magnetic Resonance Imaging may be a better imaging tool, especially in children, including infants, due to its superior soft tissue resolution and multiplanar capability [36, 37]. A loculated fluid collection or a pseudocyst formation can be more easily visualized on Magnetic Resonance Cholangio Pancreaticography (MRCP) than on ultrasonography. It is able to demonstrate the wall of the gall bladder, the defect and the biliary tree to a much better advantage and more convincingly. It has also been found to be useful in the evaluation of the pancreaticobiliary junction anomalies. The effectiveness of a dynamic MRCP with secretin stimulation [38] in the evaluation of the pancreaticobiliary malformations has recently been reported. However, it has limited use in the diagnosis of spontaneous biliary perforations in patients with fluid collections in locations such as the perirenal space or the lesser sac.

Gd-BOPTA and Gd-EOB-DTPA allow a hepatic dynamic imaging and are eliminated by the biliary system. They provide both functional and morphological information by showing the bile leakage directly. They increase the signal intensity of the bile ducts during the hepatobiliary elimination and the leakage of the contrast agent into the gallbladder and to the pericholecystic area can be easily detected in the delayed phase post contrast T1W images [39-41]. However, the safety of these contrast agents has not been established in children who are younger than 18 years [42].

MANAGEMENT

This condition is associated with high mortality unless it is managed well in time. An early surgical treatment after a pre-operative stabilization is highly recommended, to prevent the setting in of a superimposed infection.

There is no single procedure which can be recommended for all the patients with biliary perforations. The treatment has to be tailored according to the age of the patient, the general condition of the patient, the severity of the peritonitis, the site of perforation, the associated intra-operative findings and the findings on a per-operative cholangiogram. Controversy exists regarding the optimal surgical modality, which is probably related to the relative lack of understanding of the cause of the spontaneous perforation.

Biliary ascites is evident on the opening of the peritoneum. However, the site of the biliary perforation is not always easily discernible. There may be an inflammatory mass which may extend from the porta hepatis to the second part of the duodenum. A pseudocyst is formed, which might be confused with a choledochal cyst. However, the wall of the pseudocyst is much thinner as compared to that of the choledochal cyst. The common bile duct is usually identified after a careful dissection into the pseudocyst. The site of the bile leak may then be identified. An intra-operative cholangiogram may be helpful for confirming the presence of the biliary leak into the false cyst and to rule out a distal obstruction or a pancreaticobiliary malunion.

A cholecystectomy is curative in most of the cases, with the perforation being located in the gall bladder or in the cystic duct. Good results with simple repairs of the gall bladder perforations have also been reported during infancy [15]. Cholecystectomy is also recommended for perforations of the cystic duct, provided a distal obstruction to the flow of bile has been ruled out.

The available surgical options for spontaneous perforations of the bile ducts include:

a. A simple drainage with or without a cholecystectomy
b. A primary repair of the perforation with or without an external biliary drainage (cholecystostomy or T tube drainage)
c. Excision of the choledochal cyst, followed by a biliary reconstruction as a primary or a secondary procedure
d. A hepaticojujenostomy, especially if there is a pancreaticobiliary malunion or a distal obstruction
e. Diagnostic laparoscopy and a simple percutaneous drainage
f. ERCP and biliary stenting [43]
g. For the cases wherein the wall of the cyst is sloughed off due to necrosis, a T-tube may be secured with a purse-string in the proximal and the distal stumps, for external drainage of the bile via the third limb [44].

In the absence of a distal stenosis of the common bile duct, simple drainage procedures in the region of the perforation with or without a catheter drainage of the common bile duct via the perforation or a simple suture repair of the perforation may be sufficient.

A distal obstruction of the common bile duct may be related to the presence of the inspissated bile plugs. A spontaneous clearance may be anticipated over a variable period of time after the episode of the bile duct perforation has subsided. Flushing of the distal common bile duct with a fine catheter which is passed from the site of perforation has also been recommended, to remove the bile plug. Cholecystectomy may be added as a preliminary procedure, if the distal common bile duct remains obstructed and it may be dealt with at a later stage with various 'short circuiting' procedures. A hepaticojujenostomy may sometimes be contemplated as a primary procedure, especially if there is minimal inflammation.

Even in the presence of a choledochal cyst or a pancreaticobiliary malunion, the initial treatment which is recommended is drainage of the peritoneal cavity and the bile duct, to tide over the emergency situation. Subsequently, as and when the sepsis resolves and after allowing sufficient time for the inflammation to settle down, a biliary reconstruction should be contemplated.

LONG TERM OUTCOMES

A spontaneous biliary perforation is a rare but important cause of surgical jaundice. The overall prognosis of the condition is good, provided an early surgical management is instituted. A spontaneous biliary perforation and the resulting biliary peritonitis are associated with a marked degree of inflammatory reactions and they may result in narrowing of or a stricture in the common bile duct, or even an "acquired" biliary atresia in infants [25]. Stenosis of the bile duct is probably the most common complication which is reported in the patients who are managed with a simple drainage

[3]. Portal vein thrombosis has been reported in the patients who develop biliomas or pseudocysts [14]. Yano et al., reported three patients with idiopathic perforations of the biliary tract, who had been treated by a simple drainage procedure and subsequently developed choledochal cysts during 2-8 years of follow-up [45]. It has been suggested that the biliary perforation in childhood and the choledochal cysts, both were related to the pancreaticobiliary junction anomaly [45].

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