ORIGINAL ARTICLE

SPONTANEOUS PERFORATION OF BILE DUCT, CLINICAL PRESENTATION, LABORATORY WORK UP, TREATMENT AND OUTCOME

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Background: Spontaneous perforation of bile duct (SPBD) is a rare and often misdiagnosed entity. Though rare, it is the second most common surgical cause of jaundice in infants, after biliary atresia. This study was planned to determine the clinical presentation, study different diagnostic modalities, treatment and outcome of patients with spontaneous perforation of bile duct. **Methods:** This descriptive case series, comprising 22 patients with spontaneous perforation of bile duct over a period of 24 months. Clinical presentation, biochemical abnormalities, imaging details, treatment options and outcome were studied. **Results:** Total 22 patients (12 Males and 10 Females) between ages of 1.5–36 months were studied. Associated anatomical defects included choledochal cyst in 7 (31.8%) while acquired biliary atresia in 1 (4.5%). Elevated liver enzymes (ALT and AST) were present in 16 patients (72.7%) and 5 (22.7%) had bilirubin above 3 mg/dl. Coagulopathy was seen in 8 (36.6%) patients. Abdominal USG showed presence of ascites in all 22 (100%), hydrocele in 2 (9.0%), inguinal hernia in 1 (4.5%), choledochal cyst in 7 (31.8%) and atretic gall bladder suggestive of acquired biliary atresia in one (4.5%) patient. HIDA scan was diagnostic in all 17 (77.27%) in which it was performed. MRCP was done in 3 (13.6%) patients. Mortality frequency was 3/22 (13.6%); one died of post-surgical sepsis second one was cirrhotic at time of presentation and didn't make It. Two were lost to follow up one which died at home while we lost contact with fourth patient. **Conclusion:** Spontaneous perforation of bile duct can present and should be suspected as an important cause of neonatal biliary ascites or peritonitis. Most patients can be managed with intravenous antibiotics, percutaneous drainage and t-tube insertion while patients with choledochal cysts required cholecystectomy with *roux en y* choledochjejunostomy. Timely recognition and intervention is associated with favourable outcome.

Keywords: Spontaneous perforation of bile duct; clinical features; choledochal cyst; treatment

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INTRODUCTION

Spontaneous perforation of the bile duct (SPBD) affects infants and children and is quite a rare condition. Reported peak age for occurrence of SPBD is around 6 months (range 25 weeks gestation to 7 years).[1]

Biliary ascites is a common presentation of SPBD presents with progressive abdominal distension, jaundice or septic shock, fluctuating course and evidence of cholestasis with mild derangement in liver functions. It is usually missed and not properly diagnosed preoperatively. On paracentesis, presence of bile, signs of peritonitis and no free gas on abdominal x-ray will help confirm the diagnosis. For good outcome, there is need of preoperative recognition so that proper early surgical intervention and proper medical management could be carried out. Although rare, after biliary atresia and choledochal cyst it is the is the third most common cause of surgical jaundice in infants.[2] it was first described in 1882 by Freeland at autopsy.[3,4]

This entity was then described by Caulfield in 1936[5]. It requires early diagnosis and proper management including surgical correction. Bile secreted by the liver, flows through the hepatic ducts, stored into the gallbladder and exits the gallbladder via the cystic duct and flows from the cystic duct into the common bile duct, finally into the small intestine. This disorder is known for almost 80 years when laprotomies were done considering other diagnosis like appendicitis or bowel perforation. The first case report was reported by Dijkstra in 1932.[6] Literature research reveals up to 150 cases.[7]

Despite many theories the cause of bile duct perforation remains speculative. Distal duct obstruction, pancreatic fluid refluxing up the common bile duct, congenital weakness of the common bile duct, or a localized mural malformation of the wall of the common duct and pancreaticobiliary mal-junction have been proposed as possible causes.[8–10]

After an initial jaundice free interval re appearance of jaundice and any one of these signs and symptoms, i.e., dark urine, acholic stools, ascites, hydroceles and inguinal hernias should raise suspicion of SPBD. Staining with bile of umbilicus and scrotum may be present in late cases. Main stay of diagnosis remain, evidence of bilirubin in ascitic fluid, ultrasound, CT scan and MRCP. It is confirmed by bilious abdominal paracentesis and HIDA scan showing

leaking of radio labeled material in abdominal cavity and absent free gas on x-ray. Hypothesis of developmental weakness of the wall of the duct is supported by many factors, after a certain intra-ductal pressure is reached that perforate the duct. The most common site of perforation is close to the junction of the cystic duct and this has been postulated by Peterson[11], in 1955 followed by Johnston in 1961[12]. There is evidence that is reported in literature suggesting that SPBD and choledochal cysts are interrelated entities and underlying pathogenic factor is also common.[12] Spontaneous perforation of bile duct is diagnosis of exclusion and labelled once other causes of biliary ascites, like trauma, choledochal cyst, stone diseases & distal atresia of the bile duct, are excluded. Surgical intervention has a definitive role including from percutaneous tube placement to complicated biliary procedures.

The objective of this study was to determine the clinical presentation, treatment and outcome of patients with spontaneous perforation of bile duct

MATERIAL AND METHODS

This descriptive case series was carried out in the department of Paediatric Gastroenterology, Hepatology and Nutrition, the Children's Hospital & the Institute of Child Health, Lahore over a period of 20 months, from 1st January 2013 till 30th October 2015.

A total number of 22 patients, confirmed cases of bile duct perforation were included in the study by consecutive non-probability sampling. After informed consent from the parents, data including age, gender, presence of jaundice, abdominal distension, hepatosplenomegaly, stool colour was collected on preformed *pro forma*. Complete blood count, Liver function tests and PT, APTT of all patients were done from hospital laboratory. Diagnostic ascitic tap of all patients was done under full aseptic precautions and appearance of ascitic fluid was noted. Ascitic fluid was analysed from hospital laboratory for protein, glucose, bilirubin and cytology, as well as dipstick was done on bed side. High suspicious of BDP was raised on presence of bile in ascites. Ultrasound abdomen of all patients was done by consultant radiologist. CT scan, HIDA scan and MRCP was done to establish diagnosis where ever necessary. All patients were managed with broad spectrum IV antibiotics and supportive care. Indwelling peritoneal catheter was placed and ascitic fluid was drained in daily increments amounting to 10-20 ml/kg/day with controlled monitoring. Surgical consultation was taken depending on individual case scenario wherever required.

SPSS-19 was used to analyse the data. Qualitative variables such as gender, clinical features, aetiology and outcome were described as frequency. Quantitative variables such as age, haemoglobin, serum bilirubin, albumin, ALT, PT etc were presented as mean, standard deviation and range. Charts and tables were constructed where appropriate.

RESULTS

Our study included 22 patients having age range between 1.5–36 months with a mean age 7.87±7.60 months. Maximum number of our patents was less than 6 months old (Figure-1). Male to female ratio was 1.2:1 (Figure-2). All 22 (100%) patients had abdominal distension and ascites, jaundice was seen in 15 (68.2%), vomiting in 6 (27.3%), fever in 6 (27.3%) and abdominal pain in 6 (27.3%). Constipation, steatorrhea and hydroceles testis was present each in 2 (9.09%) patients. Acholic stools in (5) 22.7%, bilateral inguinal hernia in 3 (13.6%), signs of peritonitis in 3 (13.6%), choledochal cyst was associated in 7 (31.8%) while biliary atresia was associated in (1) 4.5% recently diagnosed patient (Figure-3).

Serum bilirubin and liver enzymes were either normal or only mildly elevated ALT was raised above upper limit of normal in (16) 72.7%. AST was raised above upper limit of normal in (16) 72.7%. Bilirubin was raised above 3 mg /dl in (5) 22.7% and less than 3 mg/dl in 17 patients PT was prolonged in (8) 36.6% patients while it was normal in (14) 63.6% patients (Table-1).

Ascitic fluid tap was performed in all and dip stick showed raised bilirubin in fluid laboratory investigations also confirmed high bilirubin level in ascitic fluid. In most cases ascitic fluid bilirubin concentration was more than serum bilirubin level.

Abdominal USG was available for all patients and showed presence of ascites in all 22 (100%), hydrocele in 2 (9.0%), inguinal hernia in 1 (4.5%), choledochal cyst in 7 (31.8%) and atretic gallbladder signifying acquired biliary atresia in one (4.5%) patient. Ct scan was available for 3 (13.6%) patients, which confirmed the presence of fluid in abdomen and also confirmed localization of loculated fluid in 2 (9.09%) patients. HIDA scan was done in 17 (77.27%) patients showing tracer activity in peritoneum. MRCP was done in 3(13.6%) patients. Four (18.18%) patients couldn't make it 2 owing to surgical complications while 2 died cause of underlying cirrhosis.

In present study, 5 (22.7%) patients got well with percutaneous drainage of cyst and did not require any further treatment. Ten patients required t-tube insertion and in 7 Cholecystectomy and *rouex en y* procedure was done. Mortality frequency was 3/22 (13.6%); one died of post-surgical sepsis second one was cirrhotic at time of presentation and didn't make It. 2 were lost on follow up one which died at home while we lost contact with fourth patient.

Table-1: Laboratory variables:

Parameters	Mean value	±SD	Range
ALT (iu/ml)	120.2	99.6	17–378
AST (iu/ml)	79.86	60.17	15–292
S.BILIRUBIN (mg/dL)	9.16	6.92	0.8–24.5
PT (sec)	30.3	12.3	12–120
ALBUMIN (g/dL)	3.33	0.7146	1.5–4.2

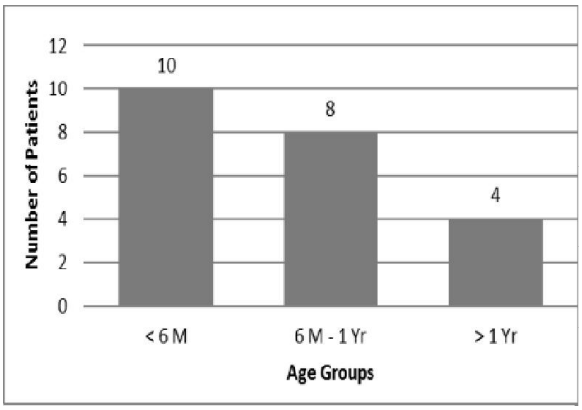

Figure-1: Age distribution

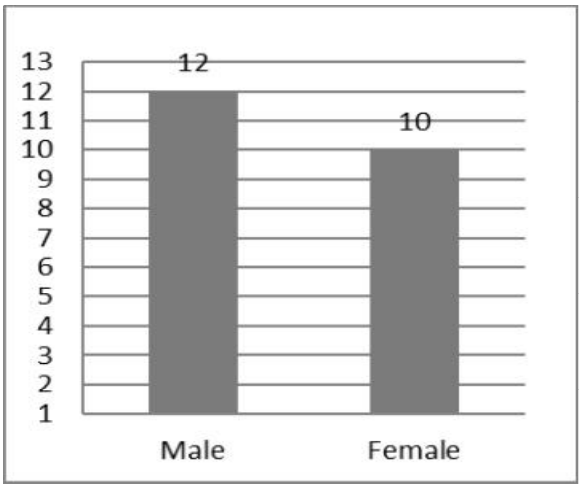

Figure-2: Gender distribution

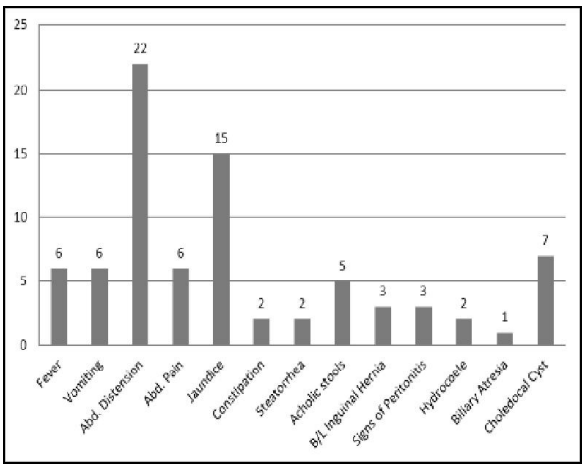

Figure-3: Clinical features

DISCUSSION

Review of extensive literature shows that Spontaneous perforation of bile duct is a rare[14], frequently missed less understood entity and reported cases are a few[15].

Age range in our patients was between 1½ months to 36 months with most patients in infantile age. These figures are concordant with existing literature where age at presentation ranges from 25 weeks of gestation to 15 years.[1,16]

Most common age at presentation was the first year of life with median age of presentation being 4 months.[7] Our study included 22 patients. And it showed same pattern of slight male preponderance. Out of which 12 were Males and 10 were Females, with a ratio of 1.2:1 with almost same sex ratio. Published literature also show similar male to female ratio.[17]

In our study, 8 patients had pre-existing disorder with SPBD presenting as a complication; i.e., acquired biliary atresia in 1 (4.5%) patient and choledochal cyst in 7 (31.8%) patients. Various publications have shown associations of SPBD with different disorders like Ivemark syndrome[18], HIV with stage IV Hodgkin lymphoma[19] and necrotizing enterocolitis[20] in the first week of life. There are Case reports of SPBD associated with acquired biliary atresia[21] and choledochal cyst[22] in literature. Spontaneous perforation of bile duct has been suggested to be a form of acquired biliary atresia.[21]

Pattern of presentation in all of our patients was in the form of abdominal distension and ascites. Aspiration of ascites revealed bile stained fluid which was confirmed to be bilious on dipstick examination for bilirubin and laboratory evaluation of fluid. Out of all, 03 patients had tense tender abdomen, cytology of aspirated ascitic fluid revealed high white cell count and predominant Neutrophils, these cases were treated as biliary peritonitis.

This observation was also in close concordance with previous studies which endorse the most common presentation to be generalized or localized ascites which may be complicated secondarily by bacterial peritonitis; a lethal complication.[22–24]

Abdominal ultrsonography was done in all 22 (100%) patients with ascites being the most consistent finding. In 2 (9.09%) patients has loculated intraperitoneal collection and 20 had frank ascites.

It has been documented that in cases with spontaneous perforation of bile duct abdominal ultrasonography will show normal intrahepatic and extra hepatic ducts along with a free or loculated intraperitoneal collection.[25]

The CT scan was done in 3 patients. It confirmed the presence of free fluid and dilated gall bladder in one patient while other 2 cases showed loculated collection it confirmed the site of loculation

which were collections of bile as masses around the common bile duct. The ability of CT scan to recognize the gall bladder wall defects is limited by the need of an IV contrast medium.[26]

MRCP was done in 3 of our patients and it helped in distinguishing anatomy and also revelled site of leakage, site of leakage in all these patients were from the posterior wall of the common bile duct. MRCP is able to demonstrate the wall of the gall bladder, the defect and the biliary tree with much better accuracy. It has also been found to be useful in the evaluation of the pancreaticobiliary junction anomalies.[27,28]

Seventeen of our patient underwent HIDA scan and all showed tracer activity in peritoneum. Hepato biliary scintigraphy (HIDA scan) in infancy is considered the most reliable method foe seeing extravasation of bile in peritoneal cavity in cases of spontaneous perforation of bile duct.[29,30]

HIDA scan is Highly recommended For spontaneous bile duct perforations as it is shown to be highly effective when it comes to sensitivity and specificity this pathology is suspected.[31,32]

Most authors recommend a conservative approach of draining the abdomen to decompress the biliary tree; once the biliary tree is decompressed spontaneous closure is typical even with distal obstruction.[33]

Early recognition and treatment yield a good prognosis, whereas lack of surgical treatment results in universal mortality.

CONCLUSION

Spontaneous perforation of bile duct can present and should be suspected as an important cause of neonatal biliary ascites or peritonitis. One third of the patients have associated choledochal cyst. Most patients can be managed with intravenous antibiotics, percutaneous drainage and T tube insertion while patients with choledochal cysts required cyst excision and with *roux en y* hepaticojejunostomy. Timely recognition and intervention is associated with favourable outcome.

AUTHOR'S CONTRIBUTION

HSM: Conceptualization of study, data collection, write up. HAC: Supervision, Literature Review, Proof reading. MAH: Data analysis, proof reading. AP, NW, ZF, IM, NA: Data Collection

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