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Spontaneous biliary peritonitis: Is bed side diagnosis possible?

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ABSTRACT

Background: Spontaneous biliary peritonitis is a rare cause of acute abdomen. In spontaneous biliary peritonitis there is perforation in the wall of the extra-hepatic or intra-hepatic duct occurs without any traumatic or iatrogenic injury and have been described more often in neonates. The symptoms may be acute or insidious delaying the diagnosis. Present manuscript deals with diagnosis and management of these cases. **Materials and Methods:** This is a prospective study and all patients of suspected biliary peritonitis presented during Dec 2010 to Feb 2012 were included in the study. After preliminary investigations in all patients abdominal paracentesis was done and in cases where intra-abdominal fluid bilirubin level was several fold higher than serum bilirubin level were subjected to exploratory laparotomy. Further investigation like T-tube cholangiogram and magnetic resonance cholangiopancreatography (MRCP) was done to rule out choledochal cyst before leveling these cases as SPBD. **Results:** A total of 6 patients were included in present series commonest presenting symptom was progressive abdominal distension without signs of overt peritonitis followed by progressive jaundice, fever and abdominal pain. On exploration site of perforation was observed in 50% of cases and in 50% of cases bile duct was not dilated. Second surgery was not required in 34% of cases. There was no mortality or significant morbidity in our series. **Conclusion:** Spontaneous perforation of bile duct is rare disease and high index of suspicion is required for diagnosis. Simple bed side test can help in diagnosis but T tube cholangiogram or MRCP are must to rule out choledochal cyst.

Key words: Bile duct, choledochal cyst, perforation, peritoneal drain

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INTRODUCTION

Spontaneous perforation of the bile duct (SPBD) is a disease in which perforation occurs in the wall of the extra-hepatic or intra-hepatic duct without any traumatic or iatrogenic injury and the cause is idiopathic once trauma and choledochal cyst are ruled out. Most common etiologic factors for this entity are: Common bile duct stones and/or cysts, blunt or penetrating abdominal traumas, hepatobilliary operations and instrumentations.[1] Only few cases of spontaneous rupture of bile duct had been reported in literature.[2-5] Despite the fact that spontaneous biliary peritonitis occurs almost invariably following biliary tract perforation, none could be easily identified with contemporary imaging. The diagnosis of this entity is difficult because of its non-specific presentation and rarity of the diseases; we are sharing our experience on this entity with a clue for its bed side diagnosis.

MATERIALS AND METHOD

We have treated 6 cases of suspected SPBD during Dec 2010 to Feb 2012 at the department of Pediatric Surgery in a tertiary care in center India. All suspected cases of biliary peritonitis with no obvious history of trauma were included in study. Patients were evaluated clinically and then subjected to preliminary hematological, biochemical examination and ultrasound of the abdomen. Once preliminary investigation and clinical examination are suggestive of biliary peritonitis abdominal paracentesis was done and fluid bilirubin level was compared with the serum bilirubin level. In cases where intra-abdominal fluid was several fold higher than serum bilirubin level, our index suspicion for spontaneous biliary peritonitis was very high. After emergency laparotomy and proper peritoneal lavage, depending on the size of perforation dilatation of CBD the decision for placing T-tube was made. In cases where site of perforation was large (large adequate to insert T- tube) and CBD was dilated we place the T-tube (the

back of the T tube was split opened and T tube was fixed by suture). In cases where site of perforation was very small or bile duct was not dilated, cholysystostomy was done. In cases where site of perforation was not localized after thorough lavage of sub hepatic drain was placed. Pelvic drain (if placed) and subhepatic drain were removed sequentially once the drainage was nil and ultrasonography showed no intra-abdominal collection. In patients, where site of perforation was not localized on exploration nuclear scan was done to rule out ongoing biliary leak [Figures 1 and 2] and latter MRCP was done to delineate the pancretiobiliary anatomy. In patients who had T tube in place Cholangiogram was done to delineate the anatomy. Definitive surgery was done after 6 - 8 weeks depending on the recovery of the patients.

RESULT

Six cases of suspected SPBD were managed during this period representing around 20% of all (30 cases) hepatobiliary cases managed at our center. Age ranged from1 month to 6 years and median age 18months. Male to female ratio was 2:1. All patients presented with history of abdominal distension, clinically evident jaundice and signs of dehydration. All except one had history of clay colored stool with recent change in color of stool [Table 1] which has been operated and cystojenunostomy was done for perforated choledochal cyst at some other hospital. One patient had a history of recurrent episode of pain, fever and jaundice and had been diagnosed as choledochal cyst [Figure 3] Provisional diagnosis of biliary peritonitis was made by history, clinical examination, preliminary investigation and comparing the serum bilirubin and peritoneal fluid bilirubin (several fold higher bilirubin in peritoneal fluid in comparison of serum bilirubin level) Table 2. On exploration, site of perforation was seen in four cases and in two cases site of perforation could not be located [Table 1]. All had diffused peritoneal collection; none had localized collection at porta hepatis or lesser sac. Out of 4 suspected cases of SPBD only two have normal caliber common bile duct and on follow up of 14-24 months they were doing well and did not require any surgical intervention, so by definition only these 2 are the true cases of SPBD.

DISCUSSION

Spontaneous perforation of the common bile duct in infants was first described by Dijkstrat in 1932. Age of presentation for non-traumatic perforation of bile duct with no evidence of choledochal cyst is usually between 2 weeks and 2 months of life whereas those with choledochal cyst can present even in adulthood.

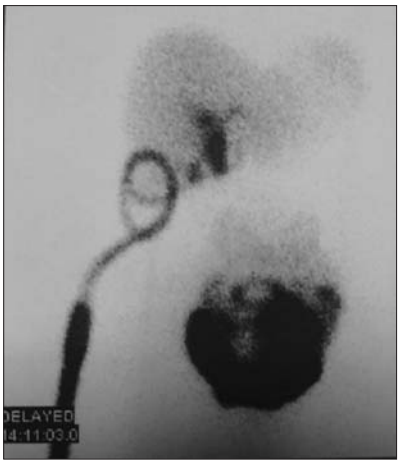


Figure 1: Radiocuclide hepatobiliary scan using TC-99m mebrofenin showing collection in CBD and T tube in situ

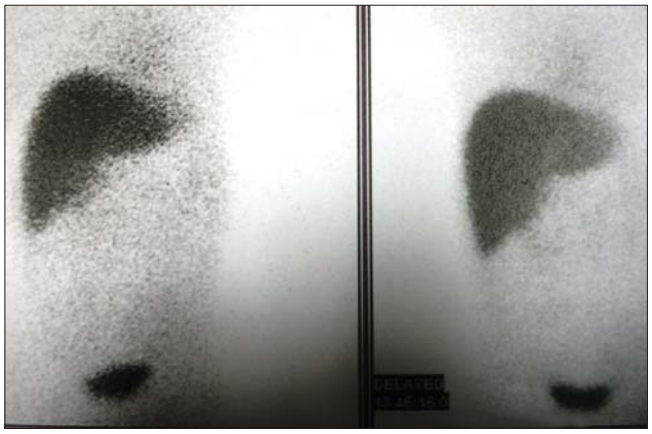


Figure 2: Radiocuclide hepatobiliary scan using TC-99m mebrofenin showing normal scan



Figure 3: MRCP Showing choledochal cyst (scan done before rupture of cyst)

Usually these patients have un-complicated birth histories and develop normally until the onset of jaundice, acholic stools or dark urine. Signs of pyrexia or peritonitis are usually absent. Progressive abdominal distension, jaundice, ascites, with no signs

Table 1: Patient profile and clinical features at time of presentation and management

Patient	Age/sex	Clinical presentation	Delay in presentation	Intraoperative finding	Management	Follow up
1	2 m /M	Abdominal distension, pain, vomiting, clinical jaundice, low grade fever, acholic stool,	10 days	No perforation seen and CBD not dilated	Laparotomy, lavage and subhepatic and pelvic drain	HIDA scan showed normal passage of in bile duct without any retention and MRCP showed normal intra and extra hepatic biliary anatomy
2	11 m/M	Abdominal distension, clinical jaundice and acholic stools	8 days	#Perforation at cystic duct and bile duct junction, very minute perforation and CBD not dilated	Laparotomy, lavage and subhepatic and pelvic drain	HIDA scan showed normal passage of in bile duct without any retention and MRCP showed normal intra and extra hepatic biliary anatomy surgery required
3*	3 years/F	Abdominal distension, vomiting, fever and abdominal pain	-	Perforation on posteriolateral aspect of dilated bile duct (OT notes)	PCD followed by definitive repair after 3 m Type IV Choledochal cyst	Roux-en-Y hepaticojejunostomy done. Doing well, no complication in follow up of 15 m
4**++	4 years/M	Abdominal distension, vomiting, clinical jaundice, acholic stool,	5 days	On posterior-lateral wall of bile duct. Grossly dilated bile duct	Laparotomy, lavage and subhepatic and pelvic drain with T-tube insertion and definitive repair after 6 weeks Type – 1 CDC	Roux-en-Y hepaticojejunostomy done.
5 ++	8 years/F	Abdominal distension, pain, clinical jaundice, Fever, acholic stool,+	8 days	On posterior-lateral wall of bile duct. Grossly dilated bile duct.	Laparotomy, lavage, subhepatic and pelvic drain with T-tube insertion and definitive repair after 6 weeks Type- 1CDC	Roux-en-Y hepaticojejunostomy done.
6	2 month/M	Abdominal distension, pain, vomiting, clinical jaundice, acholic stool,	5 days	Bile duct not dilated site of perforation not seen	Laparotomy, lavage and sub-hepatic and pelvic drain with T-tube insertion and definitive repair after 6 weeks. Type – 1 CDC	Under follow up 2 months

*patient presented to us with attempted cystojenunostomy for perforated bile duct with leaked cystojejunostomy; # very minute perforation through which Tube was not negotiable ; + patients diagnosed as choledochal cyst elsewhere but developed abdominal distension before surgery and hence referred to our centre; ++ patients included in another study; ** operated in another institute by author'

Table 2: Biochemical parameters

Patient	Serum bilirubin mg/dl	Conjugated serum bilirubin mg/dl	Fluid Bilirubin mg/dl	ALT/AST U/L	GGT U/L	Serum amylase U/L	Fluid amylase U/L
1	6.7 (0.10-1.30)	3.8 (0.0-0.40)	23	54/ (5-40)27	480 (13-86)	56 (16-110)	110
2	4.2	2.0	16	34/30	398	28	80
3*	9	4.4	Not done	48/36	270	12	Not done
4	5.8	3.2	not avialble	38/16	272	38	54
5	8	4.2	28	26/19	347	46	51
6	4	2.1	14	32/21	286	97	260

Values in bracket are normal reference value of our lab; ALT/AST – liver transaminases; GGT- Gamma Glutaryl Transpeptidase; * Patient was operated elsewhere before presentation to our hospital hence fluid bilirubin and amylase was not done

of frank peritonitis is the usual mode of presentation as seen in all cases of present series. Most of the published reports have documented site of perforation at the junction of cystic and common bile duct with a hypothesis of developmental weakness of bile duct at this site. In present series site of perforation was seen in 4 out of 6 cases, in 3 cases perforation was seen on the posterior-lateral aspect of the bile duct whereas in one case perforation was on the anterior surface of the bile duct proximal to the junction of cystic duct [Table 1].

The exact cause of SPBD is unknown and aetio-pathogenesis has not been established which may be related to a single or multiple factors.[6-9] None of our patients had sludge or stone in gall bladder or bile duct on initial imaging which is contrary to previously reported series where distal obstruction was the significant finding and was considered as an important factor for spontaneous perforation [10,11] Three out of 6 cases found to have dilated CBD hence only 3 cases were the true cases of SPBD. Hyperamylasemia was observed in 30% cases whereas it was the constant finding in few

series where anomalous union of the pancreaticobiliary duct was considered as an important etiological factor. In present series we observed that none of our patient had evidence of distal CBD obstruction and only one patient had elevated amylase level so we consider that multiple factors are responsible for spontaneous perforation of bile duct.

Nontraumatic perforation of the bile duct is difficult to diagnose because of its non-specific presentation, usually diagnosed intra-operatively,[7,12,13] In most cases of SBDP surgical intervention is performed for diffuse peritonitis or unstable vital signs but none of patients in our series had presented with features of overt peritonitis (wide table 1) because biliary peritonitis is usually a sterile chemical inflammation and hence may not create signs similar to bacterial peritonitis secondary to bowel perforation.[14] Main presenting features in our series were progressive abdominal distension, clinically evident jaundice with failure to thrive and recent change in color of stool which is contrary to the recently published series where most common presenting features were pain[10] and vomiting.[14]

Although early diagnosis of SPBD is essential for immediate surgical management and better prognosis, it is a challenging process because perforation presents with non-specific symptoms hence imaging is important to differentiate this condition from other causes of obstructive jaundice. Biochemical studies usually reveal conjugated hyperbilirubenemia with near normal transaminase with elevated serum gamma-glutamyl transpeptidase and alkaline phosphates indicating obstructive jaundice. Elevated serum amylase as well as extremely high bile amylase is important finding in these cases in many series but in present series only on patients had hyperamylesemia. Tani *et al*.[15] suggest spontaneous perforation of a choledochal cyst and biliary peritonitis should be suspected if abdominal US shows extrahepatic bile duct dilatation in association with ascites, but most common ultrasonographic finding in cases of bile duct perforation is loculated fluid collection or pseudocyst formation in and around the portal hepatis.[16]

Hepatobiliary scintigraphy can provide useful information about liver function, biliary patency, and site of perforation based on localized accumulation of radiotracer, and any biliary leakage into the peritoneum [11] and presence of peritoneal soiling can confirm that the intra-peritoneal fluid is bile without the need for paracentesis.[17] MRCP is a useful diagnostic tool for this entity in children, including infants as well as helps in delineating ductal abnormalities in such patients.[18] Delineation of Hepato-pancreatico-biliary anatomy is must in all cases if possible by intra-operative or post-operative cholangiogram or by MRCP to detect ductal abnormalities and rule out choledochal cyst to label it as a SPBD. We believe that exploration of the porta hepatis may be hazardous at time of emergency laparotomy because of inflammation and therefore recommend simple peritoneal drainage with T-tube drainage even if there is a distal obstruction or dilated CBD is evident on exploration especially if patient is presenting late as in our series. This entails less morbidity and has a good chance of curing the condition or at least stabilizing the patient for second-look definitive surgery if required depending on the hepatobiliary anatomy. If further cholangiography (either cholangiogram by T – tube or by MRCP) is suggestive of choledochal cyst biliary intestinal anastomosis is necessary to prevent biliary cirrhosis, portal hypertension, recurrent pancreatitis and ultimately biliary carcinoma. However, this can be done at a second laparotomy when inflammation has settled, though recently few authors had recommended single stage repair for this entity[14] with promising result but it should be attempted if patients present very early after perforation and proper expertise for this type of surgery is available. In our series we did not attempt for definitive surgery because all patients presented after 4 days of suspected perforation. Repair of the perforation at the time of initial surgery may be hazardous; there is also the risk of post-operative stricture. In our series we have no mortality and morbidity with this line of management and recommend this line of management for such patients especially in developing countries where patients usually present late and availability of expertise hepatobiliary surgery is limited.

CONCLUSION

Spontaneous perforation of bile duct is rare disease and high index of suspicion is required for diagnosis. Simple bed side test can help in diagnosis. However it is important to rule out choledochal cyst before labeling it as SPBD. Staged surgery is safer especially in developing countries.

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