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Case Series

Choledochal cyst perforation: experience from a centre with limited emergency resources

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ABSTRACT

Spontaneous perforation in Choledochal cyst (CDC) is a very rare initial manifestation and more commonly seen beyond infantile age. The management is challenging due to acute presentation, poor general condition of the patient and inflamed tissue that may jeopardize the appropriateness of any surgical intervention. The aim of the study was to analyse this rarity depending on clinical findings, diagnostic difficulty and optimum management plan in a limited resource set-up. It was a retrospective observational study where five patients presenting to the casualty department with biliary peritonitis between January 2015 and December 2020 were included. They were analysed with respect to symptomatology, laboratory parameters, radiology, emergency intervention with findings and definitive management plan. A female preponderance (60%) was found. Mean age was 5.4 years. One was a known case of CDC. Abdominal pain was the most common symptom. Inflammatory markers like Total leucocyte count (TLC) and Erythrocyte sedimentation rate (ESR) were raised in all (100%). Lipase was raised in 40% (2/5). Anaemia and low serum albumin were non-specific findings. A dilated Common bile duct (CBD) on Ultrasound (US) was seen in 80% (4/5). Magnetic resonance cholangio pancreaticography (MRCP) demonstrated type I CDC in all. All underwent laparotomy with lavage and external drainage followed by interval definitive surgery. Pre-operative diagnosis of a perforated CDC may not be possible. Strong clinical suspicion and bilious peritoneal fluid may point to this rare complication. Minimum exploratory manoeuvre with good lavage and external drainage should be the optimum emergency intervention. Definitive bilio-pancreatic reconstruction should be performed when tissue oedema subsides and general condition is improved. This approach usually achieves a rewarding outcome.

Keywords: Choledochal cyst, Spontaneous biliary perforation, Magnetic resonance cholangiopancreaticography, External biliary drainage, Roux-en-Y-hepatico-jejunostomy, Bilio-pancreatic reconstruction

INTRODUCTION

Congenital cystic dilatation of the biliary tree is a rare anomaly. The pioneering work of Alonso-Lej in the mid twentieth century forms the foundation of our basic knowledge of this condition till date.[1] Choledochal cysts (CDC) have been diagnosed in prenatal ultrasounds (US) as cystic intrabdominal masses as early as 20 weeks of pregnancy.[2,3] Postnatally, this may present in infancy or later in childhood. Although the etiopathogenesis of this condition remains same in all age groups, the clinical spectrum of presentation are remarkably different. The infantile form presents as obstructive jaundice and often requires distinction from biliary atresia. On the contrary, older children presents with cholangitis, pancreatitis or portal hypertension with or without jaundice.[4,5] Spontaneous perforation of the CDC is a very rare manifestation.[6]

This was seen beyond the infantile age and patients are sicker due to peritonitis, sepsis and shock. Optimum management to tide over this acute stage and subsequent

treatment plan is challenging. Though ample research paper is available discussing CDC in its entirety, this particular complication has only been enlightened as case reports. We analysed this rarity based on our experience under resource constraint of five cases in terms of patient characteristics, diagnostic challenges, emergency management and definitive intervention.

CASE SERIES

The study was retrospective observational in design and conducted in a tertiary care hospital in North India. Details of five patients (N=5) with age up to 12 years presenting to the emergency department with biliary peritonitis (diagnosed on table) between January 2015 and December 2020 were retrieved. History and examination notes were evaluated. Investigations included an X-ray abdomen erect and a US whole abdomen apart from routine blood parameters. After optimum resuscitation, all patients were taken up for surgery.

Overzealous handling of the CDC was avoided and surgery was limited only to peritoneal lavage and external drainage. A subhepatic or a drain in the cyst was kept for this purpose. An additional drain was kept in the pelvis. Post-operatively, patients were started on Total parenteral nutrition (TPN) apart from IV antibiotics. A Magnetic resonance cholangio pancreaticography (MRCP) was done after a minimum of 2 weeks. Definitive surgery was planned beyond 6 weeks. Cholecystectomy, excision of the CDC with Roux-en-Y-hepatico-jejunostomy was done in all. Patients were followed up at 2 weeks, 3 months, 3 monthly intervals for first 2 years and 6 monthly intervals for next 3 years. Pertinent symptomatology was inquired and an US abdomen done in all visits.

Outcomes

Patient profile and symptomatology

Age of the patients varied between 3 and 8 years with a mean age of 5.4 years. There were 3 females (60%) and 2 males (40%). Only one patient was a known case of Todani type 1 CDC.[7] All others presented for the first time. Abdominal pain was the most common symptom (100%) followed by vomiting (60%), abdominal distension (60%), fever (40%), features of shock (40%) and jaundice (20%) (Table 1).

Clinical signs and laboratory parameters

All children were toxic, dehydrated and had tachycardia. On examination, diffuse guarding was present all over the abdomen. No lump was palpable. Values of Haemoglobin (HB), Total leucocyte count (TLC), Erythrocyte sedimentation rate (ESR), amylase, lipase, Aspartate transaminase (AST), Alanine transaminase (ALT), Alkaline phosphatase (ALP), Total bilirubin (TB) and albumin with their mean are shown in (Table 2).

Ultrasound findings

A dilated Common bile duct (CBD) was identified in 4 out of 5 (80%). Intra hepatic biliary radicles (IHBR) were dilated in one (20%). Free fluid in abdomen was found in all (100%). Liver was enlarged in 1 (20%) but had normal echotexture. 2 (40%) showed bulky pancreas. Spleen was normal. Bowel was dilated in all (100%) (Table 3).

Intra-operative findings and post-operative course

Emergency laparotomy yielded bile stained peritoneal fluid in all. Bowel was friable and bile with pus flakes was found in Morrison's pouch, inter loop and pelvis. The extra hepatic biliary tree was oedematous and omentum adhered to the site of perforation. This was on the anterior wall of the cyst at the junction of the common hepatic duct and cystic duct in two cases. Another two had perforation on the posterolateral wall of the common hepatic duct. No exploratory manoeuvre to delineate the entire course of the CDC was done and it was left as such. Thorough peritoneal lavage was given and a tube drain of appropriate size kept in Morrison's pouch. Site of perforation could not be identified in one patient. A small cystotomy was made and drain kept in the cyst cavity for this patient. This patient had accidental removal of the drain on post-operative day 6 (POD-6). In view of clinical deterioration, the patient underwent re-exploration on post-operative day 8 (POD-8). Previous drain site spontaneously sealed and a new cystotomy was created this time. Patients were allowed orally when abdomen became soft and they passed flatus. Drain output decreased consistently by the end of first week. All except the last patient were discharged in the second post-operative week. However, the mean stay value of 19.6 days was skewed to the higher side due to prolonged stay of the last patient who had delayed recovery and was kept in the ward till definitive surgery was undertaken (Table 4).

Further investigation and definitive surgery

MRCP was done between post-operative week 2 and 3 which diagnosed a type I CDC in all patients. Only one (20%) had moderate dilatation of intrahepatic biliary radicles. One (20%) reported a Pancreaticobiliary maljunction (PBM) with a common channel length of more than 1 cm. In the remaining, PBM could not be commented upon. Open cholecystectomy with CDC excision and Roux-en-Y-hepatico-jejunostomy was done in all after a minimum of 6 weeks.

Follow-up

One has just finished the first visit. One has been reviewed for 1 year, two for 2 years and another one for 5 years. IHBR dilatation in 2 patients resolved by 6 months to 2 years but bulky pancreas persisted. There has been no significant episode of abdominal pain, jaundice, distension or bilous vomiting requiring readmission.

Table 1: Patient profile and symptomatology.

Patient number	1	2	3	4	5	Mean
Age (years)	8	5	3	8	3	5.4
Sex	F	M	F	F	M	
H/O jaundice	-	-	-	-	-	
K/C/O CDC	-	-	-	-	-	
Abdominal pain	+	+	+	+	+	
Vomiting	+	-	-	+	+	
Abdominal distension	-	+	+	-	+	
Jaundice	-	-	-	-	+	
Fever	-	-	+	+	-	
Shock	-	-	+	+	-	

Note: H/O- History of, K/C/O- Known case of, CDC- Choledochal cyst.

Table 2: Laboratory parameters.

Patient number	1	2	3	4	5	Mean
Hb (g/dl)	10.7	10.2	9.9	8.4	9.7	9.78
TLC ($\times 10^9$/L)	12.5	12.8	14.6	18.4	13.2	14.3
ESR (mm/hr)	22	30	21	44	14	26.2
Serum amylase (U/l)	327	300	550	519	640	467.2
Serum lipase (U/l)	38	480	86	637	128	273.8
AST (U/l)	35	40	28	35	30	33.6
ALT (U/l)	40	45	30	30	35	36
ALP (U/l)	230	188	196	180	256	210
Tb (g/dl)	1.2	0.3	0.8	1	1.8	1.02
Serum albumin (g/dl)	2.8	3.2	3	3	3.2	3.04

Note: HB- Haemoglobin, TLC- Total leucocyte count, ESR- Erythrocyte sedimentation rate, AST- Aspartate transaminase, ALT- Alanine transaminase, ALP- Alkaline phosphatase, TB- Total bilirubin.

Table 3: Radiological findings.

Patient number	1	2	3	4	5
CBD dilatation	+	-	+	+	+
IHBR dilatation	-	-	-	+	+
Free fluid	+	+	+	+	+
Liver	Enlarged	WNL	WNL	WNL	WNL
Liver echotexture	Normal	Normal	Normal	Normal	Normal
Bowel dilatation	+	+	+	+	+
Spleen	WNL	WNL	WNL	WNL	WNL
Pancreas	WNL	Bulky	WNL	Bulky	WNL

Note: CBD- Common bile duct, IHBR- Intra hepatic biliary radicles, WNL- Within normal limits.

Table 4: Intra-operative findings at first laparatomy.

Patient number	1	2	3	4	5
Peritoneal fluid	Bile stained	Bile stained	Bile stained	Bile stained	Bile stained
Type of CDC	Type 1	Type 1 (forme fruste)	Type 1	Type 1	Type 1
Site of cyst perforation	Anterior wall at junction of CHD with CD	Posterolateral wall of CHD	Anterior wall at junction of CHD with CD	Posterolateral wall of CHD	Not identified
Drain 1	Subhepatic space	Subhepatic space	Subhepatic space	Subhepatic space	Intra-cystic
Drain 2	Pelvis	Pelvis	Pelvis	Pelvis	Pelvis

Continued.

Patient number	1	2	3	4	5
PO stay after laparotomy (days)	12	14	10	12	50

Note: PO- Post-operative, CHD- Common hepatic duct, CD- Cystic duct.

DISCUSSION

CDC have an incidence of 1 in 100,000 to 1 in 150,000 live births in the west.[8] It is much commoner in Asia and highest incidence has been reported from Japan.[9] Sex predilection for females with a female to male ratio of 3 to 4:1 is cited in all literature.[10]

We also had more female patients in our study. Multiple theories were proposed as its aetiology with most pointing it to be congenital in nature. A congenital weakness of the ductal wall coupled with a distal obstruction in biliary drainage was hold responsible in all. The most accepted postulate was proposed in 1969 by Babbitt.[11] He described proximal insertion of the pancreatic duct in to the common bile duct due to incomplete migration of the choledocho pancreatic junction in to the second part of duodenum. This PBM gives rise to a 'long common channel'. During embryonic development, PBM creates a low resistance pathway for reflux of pancreatic enzymes. This, in association with an obstruction in distal insertion of the common channel causes cystic degenerative changes in the wall of CBD resulting in its fusiform dilatation. The same mechanism also predisposes to carcinoma gall bladder and cholangiocarcinoma in the long run.[12] PBM, although proved to be the single most important aetiology, is difficult to delineate on radiology. Only one of our patients had a demonstrable PBM on MRCP.

Anatomic classification of CDC was first proposed by Lej et al and later expanded by Todani et al from three to five types.[1,13] Type I (saccular/fusiform) dilatation of the CBD accounts for more than 90% of cases. 80% of our case showed dilatation on US and 100% showed a cyst on MRCP. The one with a normal CBD on US probably had a formefruste variant.[14] Classically, a CDC was expected to present with lump right upper abdomen, jaundice and pain abdomen. Practically, however this classical triad is not found in more than 20% of patients.[15] Though these symptoms were present in isolation, a constellation of all three were never encountered once among our patients.

The infantile form occurs in the first year of life. Children have features largely overlapping with biliary atresia like jaundice, clay stool and hepatomegaly. The adult form, as in our patients, has a more acute spectrum of symptoms like pain abdomen, fever, vomiting and jaundice. Biliary plugs or sludge in the distal channel are prone to cause intermittent impairment of bile drainage. Resultant bile stasis with bacterial superinfection may cause ascending cholangitis or pancreatitis. Clinical cholangitis like features was common and a bulky pancreas was found in two patients. Perforation in a CDC is a very rare complication. The cognizance of this condition has mainly come from case reports. Our series is only second to the one reported by Ando et al in 1998 where they included 13 cases and documented its incidence to be around 7%.[16] Fragile cyst wall from inflammation secondary to increased intraductal or intraabdominal pressure seems to be the instigating stimulus. More incidences have been reported in abdominal trauma and pregnancy. However, the mechanism of perforation is practically an exaggeration of the mechanism of aetiopathogenesis. This is almost never seen in the infantile form, a finding consistent with our series where the youngest patient was of 3 years. Multiple subclinical episodes of cholangitis for longer duration causing degeneration of the ductal wall probably is the cause.

A similar presentation, if encountered in infants, should be differentiated from the more common Spontaneous biliary perforation (SBP).[17] An SBP is usually small, occurs at the confluence of cystic and common hepatic duct and has a contained leak that can be managed by a T-tube placement only. On the other hand, a perforated CDC has a bigger rent on the anterior wall with gross collection and severe clinical features. Also, a major excision-reconstruction procedure is required for the later. In our study, although the sites of perforation were not consistent, we still attribute all to CDC in view of sick child, relatively big rent with peritoneal contamination and subsequent radiological confirmation.

Management of a perforated CDC is challenging. Some advocate a laparotomy, lavage and external biliary drainage to tide over the crisis followed by definitive bilio-enteric reconstruction few weeks later.[16,18] Others recommend definitive surgery at the time of CDC perforation to avoid the morbidities of a second surgery and risk of failure of follow up on account of premalignant potential of the cyst.[19] We performed stage wise surgery in all children in view of patient profile, laboratory markers and intraoperative findings at the first operation.

All patients were sick and in sepsis that required reasonable preoperative resuscitation; malnourished as suggested by anaemia and borderline hypoalbuminemia and on table finding of peritoneal contamination, unclear anatomy and oedematous bowel. Lack of sufficient intensive care facility also prompted for a conservative operation. This appears to be a judicious and effective approach as indicated by resolution of dilatation of IHBR post operatively and largely asymptomatic course thereafter. Persistence of bulky pancreas may be idiopathic or indicative of prolonged but mild form of pancreatitis with irreversible morphological changes.

The limitations of this study, however, are a small sample size owing to the rarity of this complication, unavailability of Computerised tomography (CT) scan in emergency evaluation and a relatively short follow up till conclusion. Management shall benefit from a bigger research with larger sample size.

CONCLUSION

Spontaneous perforation in CDC is rare. It is more common with the adult type of cyst. Patients without a known history may present for the first time with this. Such, patients are likely to be haemodynamically unstable. Bile stained peritoneal fluid may direct the surgeon towards a pathology in the biliary tree. Adequate peritoneal toileting and external drainage during the acute crisis followed by interval bilio-enteric anastomosis appears to be a relatively safe and effective approach.

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