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Spontaneously Ruptured Choledochal Cyst: Not a very rare happening- A case series

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Abstract

Introduction: Choledochal Cysts are a rare congenital abnormality of the Biliary tract which mostly present in childhood. Spontaneous rupture is a rare complication of this rare anomaly which occurs as case reports.

Methods: All patients presenting at our institute with ruptured choledochal cyst during a period of 6 years from January 2015- January 2020 were included in the study. All patients were resuscitated followed by exploration. All were managed with external drainage and definite surgery was deferred for later. MRCP was done during the 2nd post-operative week. Complete excision of choledochal cyst with Roux en Y hepatico-jejunostomy was done at 6 weeks. After All children were called for 3 monthly follow up to note any abdominal symptoms or icterus.

Results: There were five patients, who presented with ruptured choledochal cyst. Four of our patients were discharged after initial laparotomy and returned after 6 weeks for excision of Choledochal cyst and establishment of Bilio-enteric continuity. In one of our patients, the drain was spontaneously dislodged requiring reexploration. Because of poor general condition, this child had a long post-operative stay and was discharged only after definite repair. There has been no significant episode of abdominal pain, icterus requiring re admission and all children are thriving well.

Conclusion: Spontaneous rupture of Choledochal cyst is an important differential of acute abdomen with biliary peritonitis. Initial Laparotomy should be done with the purpose of peritoneal lavage and external drainage. Deferring the definite surgery for 6 weeks is associated with good outcome.

Keywords: Rupture choledochal cyst; pediatric choledochal cyst, management

INTRODUCTION

Choledochal cyst (CDC) is a rare congenital anomaly where there is dilatation of the Biliary tract. It is predominant in Eastern world and more common in females [1,2]. Complete triad of pain, jaundice and abdominal mass is only rarely present in 10-25% of cases [3].

Spontaneous rupture is one of the rare complications of this rare anomaly which occurs as case reports. Neverthless, all Pediatric surgeons atleast once if not more during their residency come across patients with spontaneous billiary peritonitis in their emergency room. Spontaneous Rupture of Choledochal cyst is one of the causes of billiary Peritonitis.

Here we report five such patients we encountered over a span of 6 years from January 2015- January 2020 who presented to emergency department with acute abdomen and shock and confirmed to be cases of

spontaneous Choledochal Cyst Rupture who were treated at our institution.

MATERIAL AND METHODS

This study is retrospective in design. The study was carried out in the department of Pediatric Surgery at a tertiary care hospital located in New Delhi.

All Pediatric patients under 12 years of age presenting to Emergency Department with acute abdomen and confirmed of having biliary peritonitis with choledochal cyst at emergency Laparotomy during a period of 6 years from January 2015- January 2020 were included in our study.

All patients were resuscitated in the emergency room, routine Lab investigations were done followed by urgent abdominal ultrasonography and Xray Abdomen in view of suspected perforation. After Primary Resuscitation, and adequate blood arrangement, in view of acute abdomen, children were taken up for Exploratory Laparotomy.

The primary laparotomy was performed with an objective to achieve thorough peritoneal Lavage and external drainage. A sub hepatic or a drain in the cyst was placed for external drainage. An additional pelvic drain was left in situ. Post-operative management was guided by patient's general condition, electrolyte abnormalities, oral intake, and drain output. Definite surgery was deferred for 6 Weeks.

MRCP was done during the 2nd post-operative week to identify dilation of intrahepatic billiary radicals and billiary pancreatic maljunction.

Complete excision of choledochal cyst with Roux en Y hepatico-jejunostomy was done at 6 weeks.

After the first post op visit at 14 days, children were called for 3 monthly follow up to note any abdominal symptoms or icterus.

RESULTS

There were five patients, who presented with acute abdomen and shock and were confirmed of biliary peritonitis with choledochal cyst at first exploratory Laparotomy. Salient points in their history with ultrasound findings are enlisted in Table 1.

Two of our patients were 8-Year-old Female presented to Emergency department with main complaint of sudden onset of abdominal pain and vomiting and dehydration for 24 hours. They had history of epigastric pain on and off for past 1 month. One of our patients was a 5-year-old male and one was a 3-year-old female who similarly presented with sudden onset abdominal pain, distension. Both these patients did not have any abdominal or other symptoms previously. None of them had any previous history of jaundice. None of them were known to have Choledochal cyst before this presentation. Another 3-year-old female, was a known case of Todani Type 1 choledochal cyst [4] diagnosed at neighbouring hospital and awaited surgery. She presented with abdominal pain, vomiting and distension. There was no history of jaundice in this patient, but was investigated for Lump abdomen by ultrasonography confirming Choledochal cyst. She presented to our emergency room with sudden abdominal distension, tachycardia, sweating and decreased oral intake for 2 days.

On examination, all these children were toxic, had tachycardia, and were dehydrated. Diffuse abdominal guarding was present. Laboratory markers were essentially within normal limits [Table 2] Hemoglobin ranged from 9.7 to 10.7g/dL (Mean 10.38), Total Leukocyte count were mostly raised 12.5 to18.4X10^9/L (Mean 14.3X10^9/L), Alkaline Phosphatase 180 to 256 U/L (Mean 210 U/L), Total bilirubin was normal to mildly raised 0.3 to 1.8 g/dL (Mean 1.02 g/dl).

Operative findings of all patients are enlisted in Table 3. At primary Laparotomy, all these patients had billiary peritonitis with pus flakes covering edematous and friable small bowel. Omentum was adhered to the site of rupture except in one patient where site of perforation could not be found. Perforation was seen on the anterior wall at the junction of common hepatic duct to the cystic duct in two of our patients [Figure1]. In other two patients, perforation was seen on the posterior lateral wall of common hepatic duct. Sub hepatic drains were placed in all patients except one where site of rupture was not seen. Drain was placed directly into the cyst cavity in this patient and complete external diversion of bile was achieved.

During post-operative stay, Colour of stools was noted and drain output progressively decreased in all patients. Nil drain output was noted from day7 to day 10 in all patients except the one with intra cystic placement of drain. There was accidental dislodgement of drain in this patient and deterioration of clinical condition. She was taken for re-exploration.

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At Second Laparotomy, there was around 700ml gross contamination of peritoneal cavity with bile, Large Choledochal cyst, and friable bowel densely adhered with the cyst and parietal wall. Previous site draining the cyst could not be freed from bowel and hence new site for drainage of cyst was chosen.

MRCP was done in the 2nd post-operative week for all patients which confirmed type1 choledochal cyst and no significant dilation if intrahepatic bile ducts in all patients. Drains were removed in 4 of our patients with uneventful post op and they were discharged to return for definitive surgery after 6 weeks. One patient with post op dislodgement of drain underwent MRCP on 10^{th} post op day after 2^{nd} Laparotomy. This child could not be discharged after 2^{nd} Laparotomy due to poor general condition and was managed with combined enteral and parenteral nutrition along with fluid electrolyte monitoring during the post-operative stay. Definite surgery was performed after 6 weeks when condition stabilized.

CDC Excision and Roux-en-Y Hepatico Jejunostomy was done at 6 weeks in all our patients. Post op period after definite surgery was uneventful in all 5 patients.

All patients have been under Follow up 3 monthly. One patient has been reviewed once in follow up after 3 months, one has been reviewed for 1 year, two of our patients have been reviewed for 2 years, and one has been reviewed for 5 years. There has been no significant episode of abdominal pain, icterus requiring re-admission and all children are thriving well.

DISCUSSION

Choledochal Cyst are a rare congenital abnormality of the Billiary Tract [5]. It mostly presents in childhood. Its varied presentations are known to us in different age group: infantile presentation with neonatal cholestasis; Lump or incidental finding in toddlers; recurrent cholangitis in adolescence. Except above common clinical presentations, rare spontaneous rupture of Choledochal cyst with billiary peritonitis occur as case reports. In a large case series, the incidence of perforated choledochal cyst being the initial manifestation has been reported as 1-2%[6].

The age of spontaneous rupture has been varied too. Infantile rupture of choledochal cyst as early as 23 weeks has been reported [7]. Most commonly the cases of spontaneous rupture of Choledochal cysts reported belong to late childhood or early adolescence. Out of five cases that we report in this article two were 8 years old, one was 5 years old and two were 3 years old. Although it should be noted that we are working in the department of Pediatric Surgery and we deal only with patients under the age of 12 years. Ruptured Choledochal Cyst has been reported in adulthood at the age of 28 years as well [8].

The cause of rupture has been obscured. Role of anomalous Pancreatico Billiary junction in reflux of pancreatic secretions to Cyst causing inflammation and weakening the cyst wall has been hypothesized but with no definitive evidence to support the same [5]. Some have suggested association with trauma [9]. There has been reports of post-partum rupture of choledochal cyst as well [10].

The cause of rupture remains unclear in our patients. The relation of size of choledochal cyst and wall thickness with rupture is also not well studied as most patients present in acute setting with no previous imaging done. Moreover, ongoing inflammation causes edematous wall due to which intraoperative assessment of cyst wall thickness is not possible.

Possible relationship of chances of rupture to type of choledochal cyst also remains unknown. All the patients reported in our series were case of Todani type 1(Sacular or Fusiform) choledochal cyst [4], which is also the most common type.

Most of the reported cases have been managed with external drainage and definite surgery deferred for later after improvement of patient's general condition and when bowel is less edematous and suitable for anastomosis [11,12,13]. We used the same approach and deferred definitive surgery at initial presentation. We managed to discharge four of our patients after initial laparotomy who returned after 6 weeks for Excision of Choledochal cyst and establishment of Billio-enteric continuity. One of our patients had an eventful post-operative period with spontaneous dislodgement of drain requiring reexploration, and also because of poor general condition of the child, she could not be discharged. She had a long post-operative stay discharged only after definite repair thus adding to bed occupancy and High-cost recovery.

The goal of first Laparotomy is crucial. We should limit ourselves to prevention of further contamination of Peritoneal Cavity. This was achieved by careful

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placement of sub hepatic drains. Attempts to formal closure of site of rupture are not very useful. The wall of the cyst is friable and increased intraluminal Pressure which was probably the cause rupture in the first place will lead to give way of the wall repair. In one of our patients where there was billiary peritonitis and a large choledochal cyst but the site of rupture could not be seen, we decided to place the drain inside the cyst which leads to complete external diversion of bile. A very strict post-operative monitoring and replacement of electrolytes and Nutritional support was required in this patient and her post-operative stay was prolonged.

The results of our treatment approach which has long been followed by others have been good. All the children are thriving well in Follow up and no significant complications occurred which would require readmission. Hence this traditional approach of deferring definite repair till edema subsides and general condition improves should not be experimented with.

Alternate means of image guided placement of drain in subhepatic collection or in the dilated bile duct may save the child from the morbidity of the initial Laparotomy. This can be tried whenever the expertise is available in selected cases with localized collection, where the patient is less toxic. Pediatric surgery backup with informed operating room is necessary before any image guided intervention in case complication occurs. Problem which limits above approach is that interventional Radiologists are not always available in Emergency setting not only in Developing countries like ours, but also in developed Nations. This makes the Laparotomic placement of drains and lavage the only option.

CONCLUSION

Spontaneous rupture of Choledochal cyst is an important cause of Billiary peritonitis and should be a differential in patients presenting with acute abdomen. Initial Laparotomy should be done with the purpose of peritoneal lavage and external drainage. Traditional approach of deferring definite repair till improvement of general condition and bowel condition provides good outcome and should be continued.

‘What this study adds’

Spontaneous rupture of Choledochal cyst is not as rare as it is thought to be.

One should keep a high index of suspicion to detect this problem when the patient presents to emergency with Peritonitis.

‘What is already known’

Initial management should aim at preventing sepsis and definitive repair should be deferred to later stage

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Figure Legends

Figure 1- Choledochal Cyst Perforation in Patient No 3

Table 1- Salient points in History and imaging of all 5 patients of interest

Patient No.	**1**	**2**	**3**	**4**	**5**
Age (Years)	8	5	3	8	3
Sex	F	M	F	F	F
History of jaundice	-	-	-	-	-
Known case of Choledochal cyst	-	-	-	-	+
Abdominal pain	+	+	+	+	+
Vomiting	+	-	-	+	+
Abdominal Distension	-	+	+	-	+
Fever	+	-	-	-	-
Shock	-	-	+	+	+
Abdominal Ultrasonography in Emergency					
CBD dilatation	+	+	+	+	+
IHBR dilatation	-	-	-	-	+

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Free fluid in abdominal cavity	+	+	+	+	+
Hepatomegaly	+	-	-	-	+
Liver echotexture	Increased Echogenecity	Normal	Normal	Normal	Normal
Bowel dilatation	+	+	+	+	+
splenomegaly	-	-	-	-	-
Others		Bulky Pancreas		Bulky Pancreas	

Table 2- Laboratory markers at presentation of all 5 patients of interest

Patient No	**1**	**2**	**3**	**4**	**5**	**Mean**
Hemoglobin (g/dL)	10.7	10.2	9.9	11.4	9.7	10.38
Total Leuocyte count ($x10^9$/L)	12.5	12.8	14.6	18.4	13.2	14.3
Alkaline Phosphatase (U/L)	230	188	196	180	256	210
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	36.0
Total Bilirubin (g/dL)	1.2	0.3	0.8	1.0	1.8	1.02
Serum Albumin (g/dL)	2.8	3.2	3.0	3.0	3.2	3.04
ESR	12	10	12	14	14	12.4

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Table3- Operative details of all 5 patients of interest

Patient No.	**1**	**2**	**3**	**4**	**5**
Peritoneal fluid	Bile stained	Bile stained	Bile stained	Bile stained	Bile stained
Type	Type 1	Type 1	Type 1	Type 1	Type 1
Site of Cyst perforation	Anterior wall at junction of cyst with cystic duct	Postero lateral wall of common hepatic duct	Anterior wall at junction of cyst with cystic duct	Posteriolateral wall of common hepatic duct	Not identified
Drain 1	Subhepatic space	Subhepatic space	Subhepatic space	Subhepatic space	Inside the choledochal cyst
Drain 1	Pelvis	Pelvis	Pelvis	Pelvis	Pelvis
Post operative stay after initial Laparotomy	12days	15 days	10 days	12 days	50 days

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