Abdominal Cystic Disease of The Neonate - A Systematic Review

Dr.S.Vijayabaskaran¹, Dr.M.Raghul², Dr.G.Rajamani³
Department of Paediatric Surgery, Coimbatore Medical College Hospital¹²³
Corresponding Author: Dr.S.Vijayabaskaran

Abstract: Intestinal obstruction in the newborn might be due to atresia, stenosis, annular pancreas, malrotations, duplication cysts, meconium ileus, meconium plug syndromes, neonatal small left colon syndrome, Hirschspring’s disease, neoplasias, trauma, and some other rare causes. Mesenteric cysts are rare intra-abdominal benign tumors without any characteristic clinical findings with an incidence of 1 per 100,000 up to 1 per 250,000 hospital admission. They have an identical pathogenesis, but may have different histopathological derivation and structures. Treatment of mesenteric cyst is indicated if it becomes symptomatic as a result of the enlargement of the cyst or complications. We outline the presentation, management and histological findings of 10 neonates who presented to this hospital from 2016–2017. Out of the 10, 3 were diagnosed antenatally and 7 postnatally. Abdominal distension and intestinal obstruction were the presenting features. All underwent surgical resection and the postoperative period was uneventful. Out of these 4 were duplication cysts, 3 were mesenteric cysts, 2 ovarian cysts and one chylolymphatic cyst.

A successful management of a newborn suffering from bowel obstruction depends largely on making a prompt diagnosis and treatment. The diagnosis usually could be based on the history, physical examination and simple radiographic studies. Lack of specific symptoms makes mesenteric cysts a diagnostic and therapeutic challenge. Undoubtedly awareness of the circumstance would direct to earlier diagnosis with proper treatment.

Keywords: chylolymphatic cyst, cystic disease of newborn, intestinal obstruction, mesenteric cyst

Date of Submission: 20-06-2018
Date Of Acceptance: 04-07-2018

I. Introduction:
Intestinal obstructions are the most common surgical emergencies in the neonatal period. Early and accurate diagnosis of intestinal obstruction is paramount for proper patient management. For evaluation and diagnosis, intestinal obstruction in neonates can be divided into either high or low obstruction on the basis of the number of dilated bowel loops present on the initial abdominal radiographs. Although three or fewer dilated bowel loops are typically seen with high intestinal obstruction, more than three are generally seen with low intestinal obstruction in neonates. In the pediatric field, the most of the mesenteric cyst cases are symptomatic intestinal obstruction is a frequent presentation, usually due to compression of the adjacent intestine by the cyst. Approximately two thirds of them are presented with acute abdomen.

Mesenteric cysts could be classified according to their histopathological features including lymphatic, mesothelial, enteric, urogenital, dermoid cystic and pseudo-cystic. Although neonates with classic radiographic findings of high intestinal obstruction, such as duodenal atresia, may directly undergo surgery without any additional imaging, an upper gastrointestinal series is typically performed for further evaluation. Similarly, an enema examination is used for further investigation of low intestinal obstruction in neonates.

A successful management of a newborn suffering from bowel obstruction depends largely on making a prompt diagnosis and treatment. The diagnosis usually could be based on the history, physical examination and simple radiographic studies.

II. Methodology:
Symptomatic neonates diagnosed with cystic lesions of the abdomen antenatally and postnatally presenting to Coimbatore Medical College Hospital from 2016–2017 and underwent surgical treatment were taken up for the study. Their presentation, management and histological findings of these neonates were studied and analysed. Their post operative period was monitored.

Asymptomatic abdominal cystic lesions diagnosed antenatally or postnatally were excluded from the study.
III. Results:
Ten neonates were included in the study. Out of these 3 were diagnosed antenatally and 7 postnatally. (Table 1) Their presenting symptom is listed below.

<table>
<thead>
<tr>
<th>Antenatal (3)</th>
<th>Postnatal (7)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Asymptomatic</td>
<td>Abdominal distension (3)</td>
</tr>
<tr>
<td>Abdominal distension</td>
<td>Intestinal obstruction (3)</td>
</tr>
<tr>
<td>Intestinal obstruction</td>
<td>Obstruction with volvulus (1)</td>
</tr>
</tbody>
</table>

Their preoperative radiographic images are shown below (Fig 1, 2, 3, 4)

**Fig: 1** - Right sided space occupying lesion displacing bowel loops to the left

**Fig: 2** - Features of intestinal obstruction
Five neonates with chyolymphatic cyst (1) and enteric duplication cyst (4) underwent surgical resection of the cyst along with the corresponding bowel segment. The rest with mesenteric cysts underwent cyst excision and two children with ovarian cyst had cystectomy but had no underlying healthy ovaries as they had undergone antenatal torsion. Their intra operative and post pictures are shown below.(Fig 5,6,7,8)
Table 2 summarises the patient characteristics with their post operative outcomes.

Table 2:

<table>
<thead>
<tr>
<th></th>
<th>Age / Sex</th>
<th>Clinical presentation</th>
<th>Intra op findings</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>28 d / M</td>
<td>Intestinal obstruction</td>
<td>Chylolymphatic cyst</td>
<td>Uneventful</td>
</tr>
<tr>
<td>2</td>
<td>4 d / F</td>
<td>Asymptomatic</td>
<td>Left ovarian cyst</td>
<td>Uneventful</td>
</tr>
<tr>
<td>3</td>
<td>8 d / F</td>
<td>Abdominal distension</td>
<td>Ileal duplicaton cyst</td>
<td>Uneventful</td>
</tr>
<tr>
<td>4</td>
<td>15 d / M</td>
<td>Abdominal distension</td>
<td>Sequesterated ileal</td>
<td>Uneventful</td>
</tr>
</tbody>
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<tbody>
<tr>
<td>5</td>
<td>10 d / F</td>
<td>Abdominal distension</td>
<td>Ileal duplication cyst</td>
<td>Uneventful</td>
</tr>
<tr>
<td>6</td>
<td>8 d / F</td>
<td>Intestinal obstruction</td>
<td>Mesenteric cyst</td>
<td>Uneventful</td>
</tr>
<tr>
<td>7</td>
<td>12 d / M</td>
<td>Intestinal obstruction</td>
<td>Mesenteric cyst</td>
<td>Uneventful</td>
</tr>
<tr>
<td>8</td>
<td>3 d / M</td>
<td>Intestinal obstruction with volvulus</td>
<td>Mesenteric cyst</td>
<td>Post operative ileus</td>
</tr>
<tr>
<td>9</td>
<td>3 d / M</td>
<td>Intestinal obstruction</td>
<td>Ileal duplication cyst</td>
<td>Uneventful</td>
</tr>
<tr>
<td>10</td>
<td>2 d / F</td>
<td>Abdominal distension</td>
<td>Ileal duplication cyst</td>
<td>Uneventful</td>
</tr>
</tbody>
</table>

IV. Discussion

Mesenteric cysts arise from benign multiplication of ectopic lymphatic channels lacking communication with the remaining normal lymphatic system [1,2]. Their etiology has not been clearly discovered yet [3]. The mean incidence is estimated at 1 in 100,000 in-patients [4,5]. These benign tumors are unusual causes of intra-abdominal masses in childhood [6,7]. The diagnosis is made before the age of 10 years in 25% [7,8]. They may occur anywhere in the mesentery of the GI tract from the duodenum to the rectum, but they are most commonly localized in the mesentery of the ileum followed by the mesentery of the large intestine and retroperitoneum.

A chylolymphatic cyst is a rare variant of a mesenteric cyst. These cysts present within the mesentery, lined with a thin endothelium or mesothelium and filled with chylous and lymphatic fluid. Although mesenteric cysts in general have been reported in the literature fairly frequently, chylolymphatic cysts in the pediatric age group are extremely rare in the modern medical literature, therefore very little information is available regarding their presentation and complications.

The clinical presentation of the lesion depends primarily on the location as well as the size of the cyst. In fact, they can present either with nonspecific abdominal complaints or with acute abdominal pain [7,8]. The clinical presentation depends also on its associated complications. However, many of these cases are asymptomatic and diagnosed incidentally[2].

Abdominal pain is documented as the commonest symptom and an acute abdominal presentation revealing a bowel obstruction is often reported in infants. Rarely an abdominal distension or a mass are found on physical examination [2,6].

Our experience revealed that five neonates presented with an intestinal obstruction. Complications include torsion, infarction, volvulus formation the latter observed in one of our cases. Volvulus due to the mesenteric cyst in newborn is rare life-threatening condition. Late diagnosis of volvulus contributes to a high rate of morbidity and mortality. It has variable degrees of presentation and survival.

Other complications were also reported like perforation, infection, anemia from intracystic bleeding, and rupture which is a rare condition usually occurring following an abdominal trauma. Cystic abdominal masses are easily evaluated radiologically by ultrasonography, CT scan and MRI. Ultrasound is a very sensitive and specific radiological imaging modality used not only for the diagnosis but also for the follow-up of these cysts, even in the prenatal period [9]. Ultrasonography is a sensitive and specific radiological assessment that provide several radiological features of the lesion. Once an abdominal mass is suspected, ultrasonography should be performed for an initial radiological evaluation. It is feasible and reveals fluid filled cystic lesion.

Computed Tomography and Magnetic Resonance Imaging could be helpful and essential in order to obtain a better description of the mass [10]. Additional information including lesion origin, its relationships and its adhesion to visceral organs, this can be useful if a laparoscopic surgery is considered. Abdominal ultrasonography was performed in all of our reported cases and showed a cystic mass in all cases. Malignant mesenteric cysts have not been reported in children.

Different treatment modalities that have been attempted include simple drainage, enucleation, marsupialization and surgical excision. The preferred mode of treatment is an operative resection. A mesenteric cyst can be removed completely by enucleation. Laparoscopic management of mesenteric cysts is also being reported [11].

The treatment of choice is a total excision of the cyst whenever feasible because of the risk of recurrences. In fact, the sole aspiration of the cyst is not indicated and a complete removal of the cyst is essential and is reported as a procedure of choice to avoid the cyst recurrence. Segmental bowel resection could be necessary if the intestinal blood supply can’t be preserved especially in huge cysts with excision difficulties [2].
In our report a complete excision of the cyst required a small bowel resection in five cases. Intraoperatively, the cysts were huge and closely approximating the bowel wall, therefore a complete excision without intestinal resection was impossible. On histological examination, lesions have an endothelial cell layer, lack muscular lining and are minimally vascular. Once completely removed, cysts have an excellent prognosis with a low risk of recurrence. No recurrence was noted in our patients during the follow-up period.

V. Conclusion:
A successful management of a newborn suffering from bowel obstruction depends largely on making a prompt diagnosis and treatment. The diagnosis usually could be based on the history, physical examination and simple radiographic studies.

Lack of specific symptoms and rare incidence makes mesenteric cysts as a diagnostic and therapeutic challenge, undoubtedly awareness of the circumstance would direct to earlier diagnosis with proper treatment.

References: