Malrotation with Midgut Volvulus with Mesenteric Cyst - A Rare Association

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Abstract

Case Report

Incomplete intestinal fixation or malrotation of gut with midgut volvulus is one of the important causes of bilious vomiting in neonates. The incidence of malrotation of gut in population is 4% [1]. Mesenteric and omental cysts are rare; the incidence is approximately 1 per 105,000 admissions to general hospitals and 1 per 20,000 admissions to pediatric hospitals. These cysts are reportedly more common in females than in males and in white persons than in nonwhite persons. The present case refers to a 2month old male child who presented with bilious vomiting and abdominal distention. After doing ultrasonography abdomen, the patient underwent exploratory laparotomy with Ladd's procedure and excision of Mesenteric cyst.

Keywords: Mesenteric cyst, malrotation, volvulus.

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INTRODUCTION

Intestinal malrotation is a congenital anomaly of intestinal rotation and fixation. The "classical" clinical manifestation of malrotation is that of acute midgut volvulus in a previously normal new born presenting acutely with bilious vomiting with or without abdominal distension. Mesenteric cysts have been rarely observed in association with malrotation, but whether this is a primary anomaly or results from lymphatic obstruction due to chronic midgut volvulus is not clear. This is a case report of an infant who presented with abdominal distention and bilious with mesenteric cyst; a rare association.

CASE REPORT

A 2 month old male child presented with abdominal distention and bilious vomiting. Patient was a full term baby born by normal vaginal delivery with history of NICU stay or any history of delayed passage of meconium. On clinical examination patient had abdominal distention and guarding, no rigidity.

RT was inserted and all routine investigations were done and X-ray and ultrasonography abdomen was done. X-ray was showing multiple air fluid levels and USG abdomen was done. Ultrasonography of abdomen was suggestive of mesenteric cyst measuring 2.2 cm x 1.8 cm with mildly dilated bowel loops with reduced peristalsis and features suggestive of acute small bowel obstruction.

Exploratory laparotomy was done. Intra operatic findings showed malrotation with midgut volvulus with Mesenteric cyst around 4 feet from IC junction (Fig-1 and Fig-2).

Type 1 mesenteric cyst was seen which could be excised with out bowel resection. Ladd's procedure was done. Procedure was uneventful and patient was extubated. POD 4 RT clamping was started. POD 5 feeds started and patients shifted on full breast feed. Patient tolerated feeds and passed stools. POD 6 patient was discharged.



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DISCUSSION

Intestinal malrotation is a congenital anomaly of intestinal rotation and fixation approximately 1 in 500 births [2, 3] however only 1 in 6000 cases are symptomatic [4]. 25% cases are asymptomatic and 75% are symptomatic; most of them presenting in first year of life [3, 5, 6]. Classically malrotation presents as acute midgut volvulus with bilious vomiting with or without abdominal distention [5, 7]. Acute midgut volvulus if left untreated may lead to small bowel necrosis with small bowel loss leading to short bowel syndrome and dependance on parenteral nutrition and other complications [5, 7].

Normal intestinal rotation and fixation occurs between the 4th and 10th weeks of gestation. During this time, the rapid growth and lengthening of the midgut in comparison to the abdominal cavity forces the bowel to herniate through the yolk sac.

As the bowel later reduces back into the abdominal cavity, it creates two distinct 270° counterclockwise turns in the proximal and distal midgut with relation to the superior mesenteric vessels [8]. Rotation may be described according to how it affects the two ends of the intestinal tract (i.e., the proximal duodenojejunal loop and the distal cecocolic loop) and the simultaneous rotation of these two components [2]. The physiologic fixation is a normal occurrence and keeps the ascending and descending colons anchored in the right and left abdominal gutters. In malrotation, the right colon can be abnormally positioned, resulting in aberrant attempts at fixation.

These attempts cause the colon to develop attachments to the right side of the retroperitoneum that have become known as Ladd's bands [9]. The midgut is divided into the pre-arterial (duodenojejunal) and postarterial (cecocolic) loops by its position proximal or distal to the SMA, respectively. At the completion of the process of rotation, the DJJ lies posterior and to the left of the SMA and the cecum lies in the right lower quadrant. At the conclusion of this rotation, normal fixation occurs during the 12th week of gestation and results in a broad mesentery extending from the DJJ to the cecum.

The terms "prearterial" and "postarterial malrotation", while absent from the radiology literature, can be found in the surgical literature and refer to the rotational anomaly that results from abnormal rotation and fixation of the duodenojejunal loop or cecocolic loop, respectively [9]. Classic malrotation is defined as abnormal rotation of both loops resulting in a malpositioned DJJ and a high, medially positioned cecum. Universal procedure for malrotation being Ladd's procedure.

Cysts of the mesentery, retroperitoneum, and omentum are uncommon; omental cysts are the least common of the three [10]. The incidence of abdominal cysts has been reported to be approximately 1 per 100,000 hospital admissions [11].

The most commonly accepted theory, proposed by Gross, is benign proliferation of ectopic lymphatics in the mesentery that lack communication with the remainder of the lymphatic system. These cysts are thought to arise from lymphatic spaces associated with the embryonic retroperitoneal lymph sac, analogous to cystic hygromas of the neck arising in association with the jugular lymph sac.

Beahrs and colleagues classified cystic disease of the mesentery into four categories on the basis of possible etiology: embryonic and developmental cysts, traumatic cysts, neoplastic cysts, and infective cysts.

A "mesenteric cyst" is defined as any cyst located in the mesentery; it may or may not extend into the retroperitoneum. It has a recognizable lining of endothelial or mesothelial cells. Mesenteric cysts can occur in the mesentery anywhere along the gastrointestinal tract from the duodenum to the rectum. They may extend from the base of the mesentery into the retroperitoneum. Omental cysts are located in the lesser or greater omentum. The most common location is in the ileal mesentery. In the colonic mesentery, cysts occur most commonly in the sigmoid mesocolon.

Mesenteric cysts are most commonly single and multiloculated; the fluid is generally serous when the cyst involves the distal small bowel or colonic mesentery and chylous when it is located in the proximal small bowel mesentery.

Complications associated with mesenteric and omental cysts include intestinal obstruction (most common), volvulus, hemorrhage into the cyst, infection, rupture, torsion of the cyst, obstruction of the urinary or biliary tract, and malignancy. The reported incidence of malignant conditions (sarcoma, lymphangio endothelioma, or, rarely, adenocarcinoma) is 3%, although in a recent adult series of 16 patients (ages 12 to 68 years), there were three malignant cysts (18.8%). No malignant mesenteric or omental cysts have been reported in children.

On physical examination, a majority of children with mesenteric and omental cysts have abdominal distention with or without a palpable mass. A definite mass may be difficult to palpate because of its large size, soft and fluid consistency, and great mobility. The mass may be huge, filling the abdominal cavity and simulating ascites. It is dull to percussion. If a definite mass is palpable, mesenteric cysts are generally movable in the transverse plane. The goal of surgery is complete excision of the mass. If enucleation or resection is not possible, the third option is partial excision with marsupialization of the remaining cyst into the abdominal cavity. Approximately 10% of patients require this form of treatment. If this procedure is done, the cyst lining should be sclerosed with 10% glucose solution, electrocautery, or tincture of iodine in an attempt to minimize recurrence.

A simple and universal pathologic classification system that considers the different varieties of mesenteric cysts has been proposed by Losanoff and colleagues [12]. Types 1 and 2 are easily cured with resection or enucleation, with or without concomitant bowel resection. Types 3 and 4, extending into the retroperitoneum, require complex surgical procedures and often sclerotherapy as well. Recurrence in types 3 and 4 is more common than in types 1 and 2.

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