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Mini Review

Chylolymphatic Cyst with Midgut Volvulus and Malrotation in Paediatric Population: A Rare Triad and Comprehensive Scoping Review

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Abstract

Introduction: Occurrence of Chylolymphatic cyst with malrotation and midgut volvulus is a rare clinical entity and occurs as a result of lymphatic proliferation due to lymphatic obstruction caused by volvulus.

Methods: Since its first inception very few studies have been published on this topic. We did a scoping review to identify the existence of the said condition in literature. We conducted this analysis using Mesh search terms "midgut volvulus", "mesenteric cyst", "Chylolymphatic cyst" and "malrotation" in PubMed, from the inception till current date.

Results: Scoping review revealed that only 13 cases are reported in literature so far. The cyst predominantly occurred in the jejunum (61%) and, to a lesser extent, in the ileum (30%) among the cases investigated. Most postoperative recoveries proceeded without complications, barring two cases identified during postmortem examinations. Intraoperative findings included the presence of a Ladd's band in 5 out of the 13 cases; however, the referenced studies did not note significant additional anomalies, except for congenital atresia of the transverse colon.

Conclusion: As this is a rare combination of two congenital anomalies, preoperative clinical diagnosis is difficult and requires careful evaluation and surgical management.

Keywords: Chylolymphatic cyst; Midgut volvulus; Malrotation

Introduction

Malrotation of the midgut is a congenital anomaly originating during embryonic development, characterized by improper rotational alignment around the axis of the superior mesenteric artery. This condition leads to bilious emesis and acute intestinal obstruction [1]. Midgut volvulus represents a medical emergency with the potential to progress to bowel ischemia, necrosis, and, in severe cases, bowel perforation. It necessitates prompt surgical intervention, as no effective nonsurgical treatments currently exist [2]. Mesenteric Chylolymphatic cysts, rare congenital lesions occurring in approximately 1 in 20,000 cases in the pediatric population, are thought to arise due to the benign proliferation of ectopic lymphatic tissue within the mesentery due to lymphatic obstruction resulting from the twist of the volvulus [3]. Mesenteric cysts are rare intra-abdominal masses in the pediatric population, with the Chylolymphatic variant comprising only 7.3% of all abdominal cysts [3]. The co-occurrence of these anomalies is even rarer and is sparsely documented in existing literature. We conducted a rapid scoping review of previous studies to confirm its rarity.

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Discussion and Scoping Review

Midgut volvulus, a congenital rotational anomaly, arises due to non-rotation, reverse rotation, partial rotation, or adherence to the mesentery following the physiological herniation of bowel loops around the Superior Mesenteric Artery (SMA) axis during the embryological period [1]. The Ladd's procedure is performed by open or laparoscopic techniques to reduce volvulus and remove Ladd bands if present [4]. The derotation of volvulus is relatively easy by open technique and involves anticlockwise derotation which can be performed using "steering wheel technique" in laparoscopy [5]. Post-surgical recurrence risk is reduced by widening the mesentery, with recurrence rates ranging from 2% to 8% [6]. Bowel loops are examined for necrosis, a potential cause of short bowel syndrome in cases of acute midgut volvulus [7]. In the present case it was complicated by a Chylolymphatic cyst which needed resection.

As this anomaly is sparsely reported, we conducted a scoping review and identified 8 case reports and 1 case series available as of October 2023, all dating back to the inception of the literature search (Table 1) [3,8-14], with Inclusion criteria as pediatric patients less than 18 years having mesenteric cyst, mid gut malrotation, without any other congenital anomalies or iatrogenic/traumatic mesenteric cysts. Overall, 13 cases fulfilling the inclusion criteria were taken for review for which the PRISMA diagram (Figure 1).

We conducted this analysis using Mesh search terms "midgut volvulus", "mesenteric cyst", "Chylolymphatic cyst" and "malrotation"

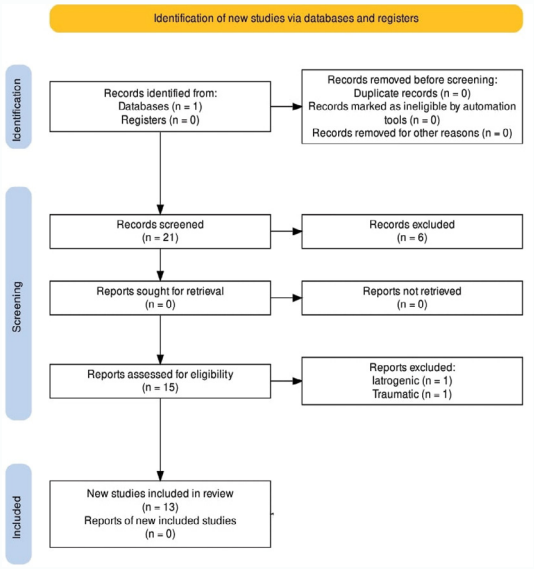

Figure 1: PRISMA diagram.

in PubMed, from the inception till current date. Notably, we observed an almost equal incidence between males and females in these cases. Commonly reported symptoms across most presentations included abdominal pain, abdominal distension, and a previous episode. However, abdominal examination findings indicated mesenteric cysts in only 3 out of the 13 cases (23%). Imaging techniques, including ultrasound and computed tomography, identified mid-gut malrotation with volvulus in only 5 out of the 13 cases (38% sensitivity), with 3 cases showing cysts alone. Further review of literature reveals, that the cyst was most frequently located in the jejunum (61%), followed by the ileum (30%) in the cases examined. Postoperative recoveries were generally uneventful, except for 2 cases that were diagnosed after conducting postmortem. A Ladd's band was observed intraoperatively in 5 out of the 13 cases, however, the aforementioned studies did not report significant additional anomalies, with the exception of congenital atresia of the transverse colon found in a study by Fukuta et al. [15].

Mesenteric cysts are fluid-filled structures located within the mesentery, and they may or may not extend into the retroperitoneum, often featuring a mesothelial cell lining [16]. These cysts vary in size, ranging from small sub-centimetric cysts to larger ones that resemble complex ascites [6]. In the present case, intraoperative findings revealed a dumbbell-shaped fluctuant cystic lesion partially enveloping the jejunal loop, lacking major feeding vessels. Small mesenteric cysts are challenging to diagnose clinically, and provisional diagnosis is typically reliant on imaging, which can miss such cyst. Recurrence and malignancy are rare in this condition, although secondary complications may include fluid leakage leading to infection, bowel loop herniations, and obstructions [17]. A confirmed diagnosis is typically established through laparotomy and histopathological assessment. However, it could not be confirmed if the mesenteric cyst was causing the volvulus or if both entities are a mere coexistence.

Conclusion

In conclusion, midgut volvulus, a congenital rotational anomaly, presents a complex scenario in pediatric surgery. The presence of Chylolymphatic cyst associated in these cases, while uncommon, adds an intriguing dimension to the understanding of midgut volvulus. Their diagnostic challenges and rarity emphasize the clinical importance of identification and management of said condition. A scoping review in the manuscript adds to the information on this particular entity which confirms the rarity of the condition and the present case is 14th case to be reported in literature.

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Table 1: Table showing observations of rapid scoping review of literature on mesenteric cyst associated with midgutmalrotation.

S No	Author	Year	Age (yrs)	Gender	Clinical identification	Identification on Imaging	Location	Surgery Done
1	Kron and satinsky [8]	1954	6	Male	No	No	Jejunum	Excision
2	Bentley and O'Donnell [9]	1959	6	Female	Yes	No	Jejunum	Intestinal Resection
3	Bentley and O'Donnell [9]	1959	4	Female	No	No	Ileum	Intestinal Resection
4	Bentley and O'Donnell [9]	1959	6	Female	No	No	Jejunum	IS (death)
5	Bentley and O'Donnell [9]	1959	2	Male	No	0	Ileum	IS (death)
6	Namasivayam et al. [10]	1992	10	Female	Yes	Yes	Jejunum	Excision
7	Weeda et al. [11]	2008	6	Male	No	Yes	Jejunum	Intestinal Resection
8	Weeda et al. [11]	2008	0	Male	No	Yes	Jejunum	Intestinal Resection
9	Botchway et al. [12]	2012	2	Male	No	Yes	Present	Excision
10	Botchway et al. [12]	2012	9	Male	No	Yes	Ileum	Intestinal Resection
11	Singh et al. [13]	2013	22	Male	No	Yes	Ileum	Excision
12	Alfadhel et al. [14]	2019	2	Male	No	Yes	Ileum	Resection
13	Pai et al. [3]	2021	16	Female	Yes	Yes	Jejunum	Excision

IS: In-situ, couldn't be operated

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