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Abdominal Whispers of the Lymphatics: An Unusual Case of Paediatric Ileal Cystic Lymphangioma

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Abstract

Background: Ileal cystic lymphangiomas are uncommon benign lymphatic malformations, constituting a rare entity in the pediatric population. While typically located in the head and neck region, intra-abdominal presentations, particularly involving the ileal mesentery, are exceedingly rare, accounting for less than 1% of all lymphangiomas. Their clinical manifestation varies from incidental findings to acute abdominal emergencies and can mimic other intra-abdominal pathologies, often posing diagnostic challenges preoperatively.

Case Details: We report a case of a 1-year-old male child presenting with intermittent severe abdominal pain. Imaging suggested a multiloculated cystic lesion with a clinical differential diagnosis of Meckel's diverticulum or enteric duplication cyst. Intraoperative findings revealed an 8 × 4 cm nodular, multiloculated ileal mesenteric cyst associated with the ileum. Resection of the cyst along with ileal segment was performed followed by histopathological examination, which allowed us to make a definitive diagnosis.

Conclusion: The rarity and uncommon presentation of mesenteric cystic lymphangiomas pose a diagnostic challenge. It can often be mistaken for other common causes of acute abdomen in paediatric patients, prompting unnecessary investigations and thus, should be considered in the differential diagnosis. Surgical intervention with complete resection is essential for timely management of patient.

Keywords: Cystic lymphangioma, Mesenteric, Ileum, Paediatric, Lymphatics.

Introduction

Lymphangiomas are uncommon benign lesions characterized by abnormal proliferation of lymphatic vessels, typically arising due to developmental

malformations rather than true neoplasia. They represent a spectrum of lymphatic anomalies that can manifest in various anatomical locations, most frequently in the cervicofacial region and axilla during childhood. However, intra-abdominal lymphangiomas, particularly those arising from the mesentery, are exceedingly rare, accounting for less than 1% of all lymphangiomas reported in the paediatric population [1]. The incidence of ileal lymphangiomas in the general population is estimated at approximately 1 in 250,000 hospital admissions, making them a rare clinical entity [2,3,4]. Their etiology is attributed to congenital malformation of lymphatic vessels, although acquired factors like trauma, inflammation, or lymphatic obstruction have also been implicated. In children, ileal lymphangiomas may present asymptomatically or with acute abdominal symptoms like progressive abdominal distension, pain, nausea, vomiting, or symptoms of intestinal obstruction often mimicking other pathologies like Meckel's diverticulum, enteric duplication cyst or appendicitis, thus complicating preoperative diagnosis [2,3,5].

Radiological investigations like USG and CT scan may help to identify cystic lesions and their anatomy, but definitive diagnosis of lymphangioma requires histopathological confirmation. The cornerstone of management is complete surgical excision of the lesion, which serves both diagnostic and therapeutic purposes [2]. Complete resection significantly reduces the risk of recurrence, particularly when the lesion is extensive or involves critical vascular or intestinal structures [3].

Here, we describe a rare case report of ileal mesenteric cystic lymphangioma in a one-year-old male child with an acute abdominal presentation.

Case Description

A 1-year-old male child presented to the paediatric outpatient department with chief complaints of intermittent severe abdominal pain. There was no history of vomiting, altered bowel habits, or prior surgeries. Ultrasonography revealed a multiloculated cystic lesion prompting a clinical suspicion of an enteric duplication cyst or Meckel's diverticulum. The surgical team performed a complete excision of the cyst along with the attached segment of ileum. Intraoperatively, an 8 × 4 cm chylous, multiloculated mesenteric cyst with a nodular component was identified, closely associated with a segment of the ileum. The resected specimen was submitted to the department of pathology for histopathological examination.

Grossly, a mesenteric cyst with an attached ileal segment were received in 10% neutral buffered formalin measuring 11cm in length, with additional dimensions of 5.5 × 4 cm. Multiple cysts enveloped the ileum, with the largest measuring 4 × 2 x 1.5 cm and the smallest measuring 2 × 1.5 × 1 cm. On sectioning the cysts, milky white chylous fluid and whitish mucoid material oozed out. A solid nodule measuring 3.5 × 3 × 2 cm was also present, with a bossellated external surface. The nodule also showed multiple cystic spaces on sectioning, ranging in size from 2 × 1 cm to 0.5 cm across. A few calcified areas were also noted. Multiple representative areas were processed following the standard protocol and haematoxylin and eosin-stained sections were examined.

Microscopy revealed variably sized, thin-walled dilated lymphatic channels lined by flattened endothelium. The lumina contained amorphous proteinaceous material along with scattered inflammatory cell infiltrates, comprising lymphoid aggregates with reactive germinal

centres in some of them. The lesion extended into the submucosa and muscularis propria of the ileal wall. There was no evidence of ectopic tissue or malignant features in the sections studied.

Discussion

Lymphangiomas represent a spectrum of benign vascular malformations arising from the lymphatic system, typically due to congenital developmental defects rather than true neoplastic processes. Ileal mesenteric cystic lymphangiomas (MCL) are classified into four types based on morphology and surgical considerations. Pedunculated MCL (type 1) are prone to volvulus and torsion, and can typically be excised without compromising blood supply. Type 2 MCL are sessile, fixed along the ileal border and often require bowel resection to preserve vascular integrity. MCL with retroperitoneal extension involving vital structures like the aorta and vena cava and limiting complete resection comprise type 3 MCL. Multicentric MCL (type 4) present with widespread cysts in the abdomen and retroperitoneum and have more guarded prognosis in view of surgical challenges [2,7]. These are further categorized histologically into three distinct subtypes: capillary, cavernous, and cystic lymphangiomas [1]. Capillary lymphangiomas comprise small, uniform, thin-walled lymphatic spaces usually confined to the dermis. Cavernous lymphangiomas consist of larger, dilated lymphatic channels, often associated with intervening lymphoid tissue and communicating with the adjacent normal lymphatic system. In contrast, cystic lymphangiomas, or cystic hygromas, are composed of large, dilated lymphatic spaces that lack direct communication with the surrounding normal lymphatics and are often embedded within a connective tissue stroma containing collagen and smooth muscle fibres.

Intra-abdominal lymphangiomas are exceedingly rare, constituting approximately 5% of all lymphangioma cases, with the majority presenting in the neck and axilla[6,7]. Among intra-abdominal sites, the mesentery is the most frequent location, with a marked predilection for the ileal mesentery over the jejunum or duodenum. Mesenteric cystic lymphangiomas, particularly those involving the ileum, are reported in less than 1% of all lymphangiomas, rendering them a distinct clinical rarity. The precise etiology remains uncertain; however, congenital lymphatic sequestration during embryogenesis is the most widely accepted hypothesis [8,9,10]. Literature indicates a male predominance in intra-abdominal lymphangiomas, with reported male-to-female ratios ranging from 1.6:1 to 3.2:1.[11] which is in concordance with this case report.

The clinical manifestation of mesenteric cystic lymphangiomas is notably heterogeneous. While many cases remain asymptomatic and are discovered incidentally during imaging or surgical exploration, others may present with nonspecific gastrointestinal symptoms such as intermittent abdominal pain, distension, nausea, vomiting, and changes in bowel habits [7]. In children, these lesions may present acutely due to complications including volvulus, haemorrhage within the cyst, rupture, or intestinal obstruction; conditions that can mimic more common paediatric surgical emergencies like Meckel's diverticulum or appendicitis. Imaging modalities such as ultrasound and computed tomography play pivotal roles in preoperative assessment. However, diagnostic challenges arise when cystic lesions are radiologically misinterpreted as fluid-filled bowel loops or duplication cysts, as occurred in our case.

Lymphangiomas typically present macroscopically as smooth, spheroidal or ovoid masses ranging from a few millimetres to over 25 cm. The external surface is usually whitish and glistening, with occasional septations within the cavity. The cystic content is commonly clear and watery but may vary in colour and consistency depending on lipid or protein content, sometimes appearing yellowish, chylous, or haemorrhagic [10].

Microscopically, the cyst wall comprises three layers: an inner, often discontinuous endothelial lining; a middle layer of loose, vascularized connective tissue with occasional smooth muscle fibres and lymphoid aggregates forming follicles; and an outer layer of dense, poorly vascularized connective tissue connected to the peritoneum. Lymphatic spaces within the wall may contain leukocytes and macrophage-like giant cells. Although lymphangiomas exhibit slow infiltrative growth with a risk of recurrence, malignant transformation has not been documented [10]. Our case also revealed a histologically similar multiloculated cystic lesion. Histopathological evaluation is indispensable for definitive diagnosis.

The definitive management of ileal mesenteric lymphangiomas is complete surgical excision and minimizes the risk of recurrence, which remains a concern when resection is incomplete or when lesions are diffusely infiltrative, involving vital vascular or intestinal structures. The recurrence rate varies widely in literature, ranging from 10% to 100% in incompletely resected cases. However, the feasibility of complete resection may be limited by the lesion's proximity to vital organs or blood vessels, entailing careful surgical planning. In our patient, the lesion was completely excised along with a segment of the ileum, ensuring clearance of the affected tissue while preserving bowel function. Serial ultrasonography remains the preferred follow-up tool in paediatric patients. Our patient is under regular monitoring and remains asymptomatic, with no recurrence observed on follow-up imaging.

Conclusion

Mesenteric cystic lymphangioma, despite its rarity, should be considered in the differential diagnosis of paediatric patients presenting with nonspecific or acute abdominal symptoms. A high index of suspicion, supported by thorough histopathological examination complemented by imaging modalities is crucial for definitive diagnosis while differentiating it from mimics. Complete surgical resection offers the best prognosis and prevents recurrence and associated complications.

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Legend Figures

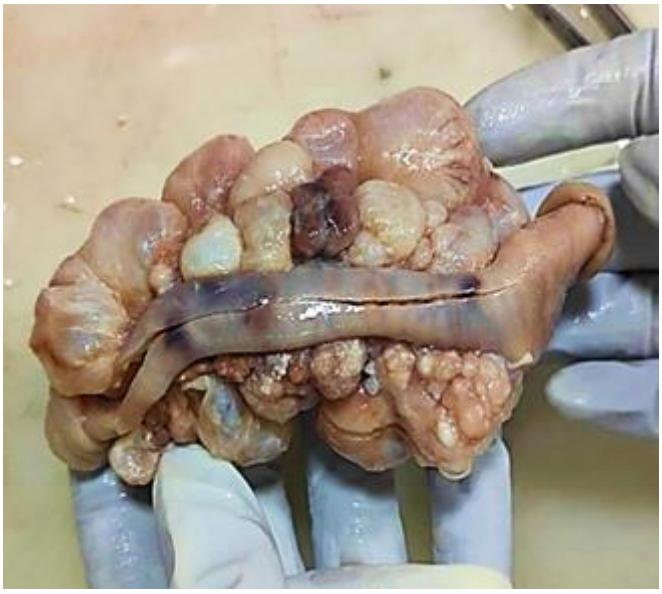

Figure 1a: Gross specimen showing multiple cysts surrounding an 11.2 cm ileal segment and inset shows cut section of the cyst.

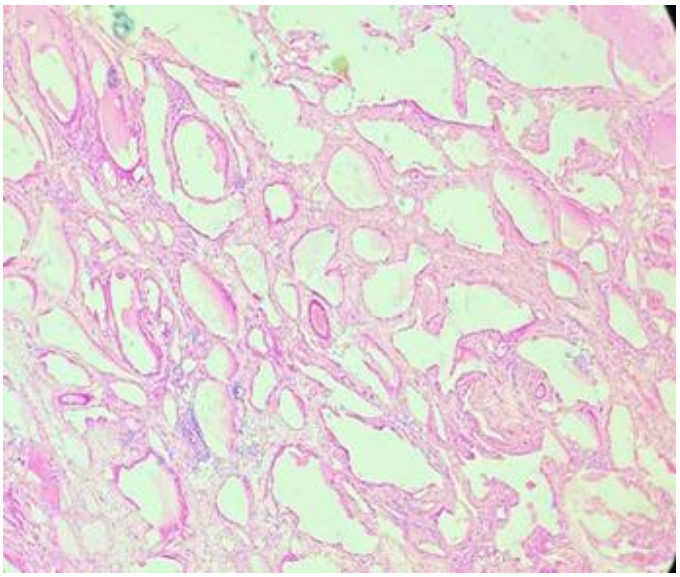

Figure 1b: Photomicrograph showing variably sized, thin walled dilated lymphatic channels lined by flattened endothelium and inset shows dilated lymphatic channels lumina filled with amorphous proteinaceous material