

Superinfected Cystic omental Lymphangioma, a rare lymphatic malformation presenting as an Acute Abdomen in a 9 year old male child. Case report

Alazar Gebrehana¹, Bethlehem Asfaw¹, Andualem Alemahu¹, Wondwosen Dereje¹, Nestanet Kasawudeg¹, Mesfin Wassie¹, and Belete Chaklu¹

¹University of Gondar College of Medicine and Health Sciences

November 23, 2024

Introduction

Omental lymphangiomas are rare vascular anomalies of benign nature. The etiology remains a subject of controversy, but they are considered to arise due to anomalies that occur during the normal embryogenic developmental process of lymphatic vessels. They are thin-walled cystic formations and only 5% of them originate from sites within the abdomen(1). This lymphatic malformations mostly arise in the head and neck as well as axillary region of neonates and adults (2).

This case report aims to highlight the uncommon site and age of clinical presentation of cystic lymphangioma and discuss the diagnostic process, and management strategies. The case report also shows the need for a high index of suspicion and early diagnostic imaging for pediatric patients who present with recurrent abdominal pain.

Case History/ examination

A 9-year-and-5-month-old male schooler came to Gondar University Specialized Hospital with acute abdomen like symptom of sudden onset of severe, sharp lower abdominal pain lasting 4 days and associated with intermittent fever. Initially, the pain started in the left lower abdomen and progressively involved the whole abdomen within one day, making it difficult for him to walk. He also developed vomiting of ingested matter which started a day prior to presentation. The patient had previous history of recurrent bouts of acute abdominal pain which started around the age of seven For which he visited nearby health centers 6 times where he was investigated with complete blood count and stool exams and recieved treatment for intestinal parasites with out any significant improvement. He had no abdominal imaging done during those times because the scan was deemed unnecessary by the treating physicians.

Upon Physical Examination he was Acutely sick-looking and in pain, otherwise conscious, not in respiratory distress, well-nourished. His blood pressure was: 90/50 mmHg, pulse rate was 112 beats per minute, respiratory rate 25 breath per minute and a fever of 38.3 degree celcius. Upon Abdominal Examination bowel sounds were normal and on Superficial Palpation, there was involuntary muscle rigidity and direct tenderness over the left lower quadrant (LLQ) and rebound tenderness but no superficial palpable mass. Upon Deep Palpation there was Smooth, round, tense, tender, ill-defined 4 centimeter by 4 centimeter palpable mass over LLQ extending to the left flank area. Mass was not bimanually palpable. There was no organomegaly. Digital rectal examination was normal.

Methods(differential diagnosis, investigations and treatment)

After admission to our pediatric ward he was investigated with Complete blood count and results showed Leukocytosis of $19,000/\text{mm}^3$ with neutrophil percentage of 83%. Hemoglobin and platelet counts were normal. Liver and renal function tests were within reference ranges. Blood cultures were taken and later showed no organisms. Stool examination and urinalysis were non-revealing. Abdomino- pelvic sonographic scanning revealed 8.6x7.4x4cm left intraperitoneal multiloculated cystic lesions which pushes the bowel wall posteriorly and has multiple septa having color flow & echodebris Figure 1.

For further description of the cystic lesions abdominopelvic CT scan was done and it showed 12x7.7x5.3cm left mesenteric multiloculated cystic masses with thin wall and septa and no solid component is seen in the cystic mass Figure 2.

Ultrasound guided fine needle aspiration was taken from the cystic lesions and it showed Lymphocytes in cluster and dispersed form admixed with scattered mesothelial cells on proteinaceous background. No malignant cells were seen Figure 3. After consultation with the surgical team it was decided to do an exploratory laparotomy and excision of this benign looking mass as an elective after treating the possible infection.

The patient was then started on intravenous antibiotics empirically with ceftriaxone 75mg/kg IV daily and metronidazole 30mg/kg in three divided doses. After 48 hours of antibiotic therapy patient's fever subsided and abdominal pain improved and after a week of therapy the pain and tenderness completely resolved but the palpable mass remained. He was then discharged and appointed for elective surgery after 2 weeks. Finally Exploratory laparotomy was done and a 4x8cm cystic mass arising from the omentum and adhered to distal transverse colon was resected and sample sent to histopathologic evaluation which later revealed adipose tissue containing differently sized cystically dilated lymphatic vessels lined by single flat endothelial cells containing proteinaceous secretory material which confirmed cystic omental lymphangioma Figure 4 .

Conclusions and result (Outcome and follow up)

After surgical resection of the mass and a hospital stay of 5 days, the patient was discharged with significant improvement and no post operative complications. On follow up, the patient had no longer pain and no palpable mass imaging after 1 month showed no recurrence.

This case of a super-infected cystic omental lymphangioma in a nine year old male who had long-standing recurrent symptoms underscores the need for increased awareness of the atypical presentations. It also highlights the need for early diagnostic imaging studies for pediatric patients with recurrent abdominal pain. Continued research into the etiology, age and types of presentations, optimal management of omental lymphangiomas will enhance patient outcomes and inform clinical practice. With comprehensive follow-up and tailored management strategies, patients can achieve favorable results.

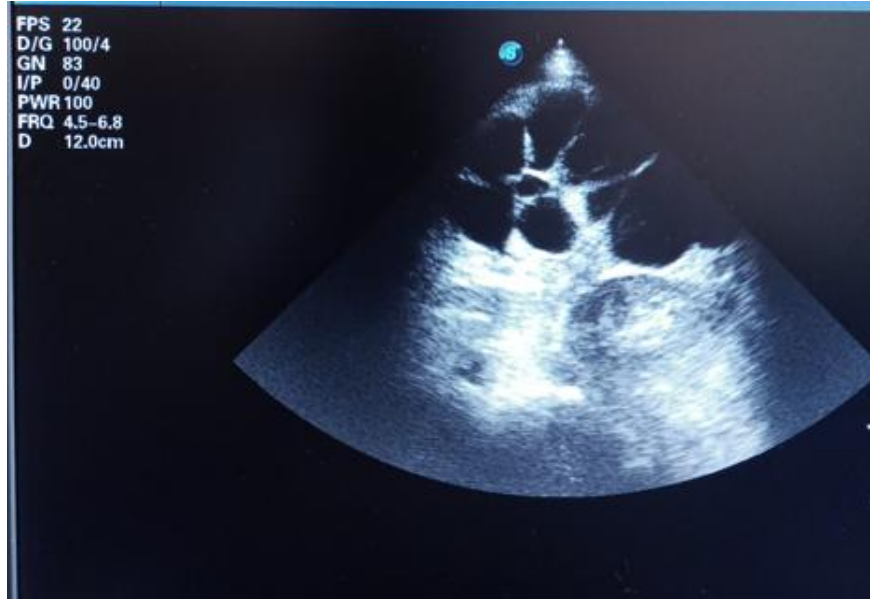


Figure 1- Abdominal Ultrasound showing a 8.6x7.4x4cm left intraperitoneal multiloculated cystic lesions which pushes the bowel wall posteriorly and has multiple septa having color flow & echodebris



Figure 2- Non contrast Abdominal CT scan showing a 12x7.7x5.3cm left mesenteric multiloculated cystic masses with thin wall and septa and no solid component

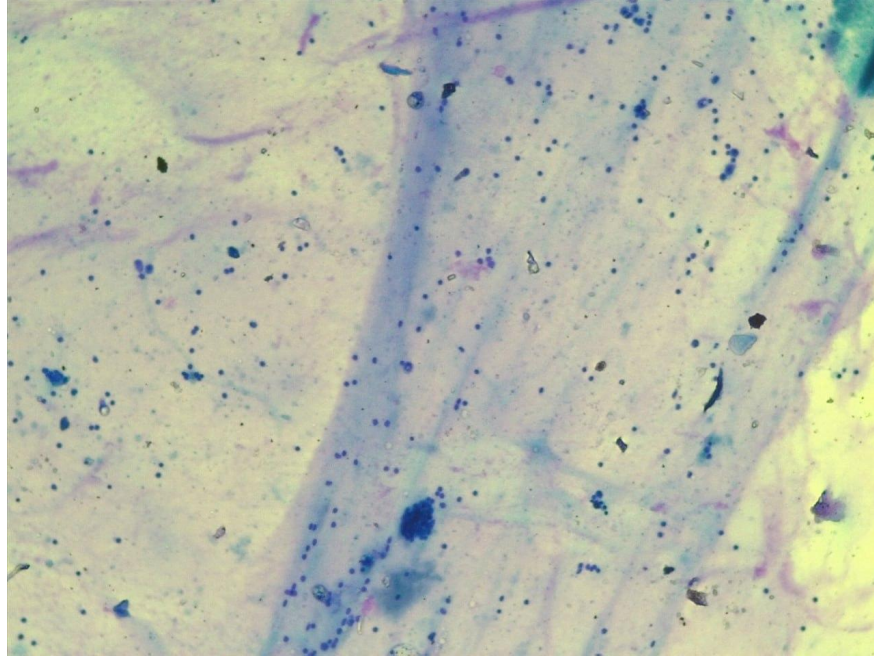


Figure 3- FNAC showing Lymphocytes in cluster and dispersed form admixed with scattered mesothelial cells on protienacious background and no malignant cells

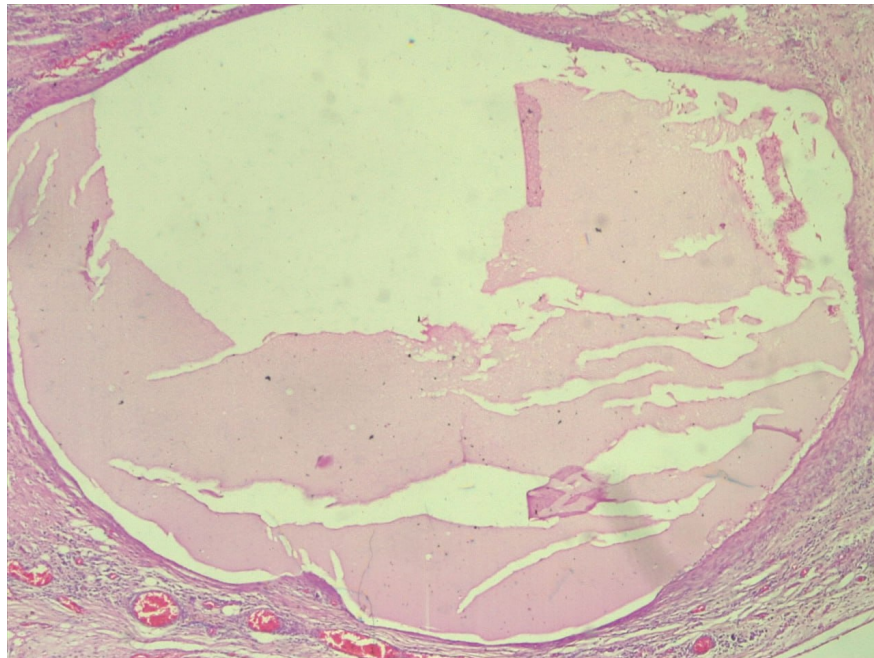


Figure 4- Biopsy showing adipose tissue containing differently sized cystically dilated lymphatic vessels lined by single flat endothelial cells containing protienacious secretory material.

Discussion

Cystic lymphangiomas are an uncommon benign maldevelopment of lymphatic vessels that mostly arise in the head and neck as well as axillary region of neonates and adults (2). Omental lymphangioma is a rare lymphatic malformation characterized by the presence of dilated lymphatic vessels within the omentum, the fold of peritoneum extending from the stomach and covering the intestines. These malformations typically arise from developmental anomalies in the lymphatic system and are most commonly diagnosed in children, although they can occur at any age (3). Most intra-abdominal lymphangiomas are asymptomatic and are often found incidentally on imaging (4). The clinical presentation of the few symptomatic intra-abdominal cystic lymphangiomas is usually nonspecific, often mimicking other abdominal conditions. Patients may exhibit symptoms such as abdominal pain, distension, or signs of intestinal obstruction. Due to its rarity, omental lymphangioma may be overlooked in the differential diagnosis of abdominal masses, which can lead to delays in diagnosis and management (3).

Imaging studies are crucial for the evaluation of omental lymphangiomas. Ultrasound often shows well-defined cystic structures, while CT and MRI can reveal the extent of the lesion and its relationship with surrounding structures (3, 5). The use of contrast-enhanced imaging techniques has improved diagnostic accuracy, allowing for better differentiation from other abdominal masses, such as cysts or tumors (7, 8).

Fine-needle aspiration cytology (FNAC) of an omental lymphangioma usually shows moderate cellularity with a proteinaceous background and the absence of malignant cells. Definitive diagnosis is established through surgical excision and histopathological examination, which reveals dilated lymphatic vessels lined by endothelial cells (6).

Histopathologically, omental lymphangiomas are characterized by dilated lymphatic vessels lined by endothelial cells, sometimes associated with a chronic inflammatory component (9, 10). These features are essential for distinguishing lymphangiomas from other similar-appearing lesions, such as lymphadenopathy or cystic tumors (11, 12). The management of omental lymphangiomas typically involves surgical intervention, especially in symptomatic cases. Complete excision is considered the gold standard treatment and is associated with a favorable prognosis (13, 14). However, the potential for local recurrence, though rare, underscores the need for long-term follow-up (15, 16). A review of the literature reveals that the recurrence rate for lymphangiomas can be as low as 5% when complete excision is achieved (17, 18).

Management of omental lymphangioma generally involves surgical excision, particularly in symptomatic cases. Laparoscopic techniques are increasingly preferred due to their minimally invasive nature, resulting in quicker recovery times and reduced postoperative complications. Laparotomy is another surgical approach. The prognosis following complete surgical resection is generally favorable, with low recurrence rates (7).

Comparing our case with those documented in existing literature, it is evident that variations in clinical presentation and management strategies exist, emphasizing the necessity for individualized treatment plans (19, 20). Future research should focus on establishing standardized guidelines for the diagnosis and treatment of omental lymphangiomas, particularly given their rarity (21, 22). Recent studies have also begun exploring the genetic and molecular basis of lymphangiomas, which could lead to novel therapeutic approaches (23, 24). The integration of multidisciplinary care, including surgical, radiological, and pathological expertise, is vital in managing these complex cases (25, 26).

Abbreviations

CBC complete cell count

WBC White blood cell

FNAC. Fine needles aspiration cytopathology

LLQ. Left lower quadrant

CT computed tomography

MRI Magnetic resonance imaging

Declarations

Acknowledgments

The authors gratefully acknowledge that all team members dedicate their best efforts to care for our patient. We would also like to thank the parents of our patient and the patient himself for being cooperative all the way through.

Funding

This research received no funding or grant support.

Availability of data and materials

All data generated and analyzed during the study are included in this published article

Clinical Trial Number: Not applicable

not-yet-known not-yet-known

not-yet-known

unknown

Ethics approval and consent to participate This study was conducted in accordance with the fundamental principles of the Declaration of Helsinki.

Competing interests

The authors declare that they have no competing interests.

References

1. Al Laham O, Khalek GM, Almaydaani M, Abazid E, Abazeed O, Alshalabi AM. A rare occurrence of an incidental primary intra-abdominal cystic lymphangioma in a Middle Eastern adult female: A case report. *Ann Med Surg.* 2023;85
2. Levine C. Primary disorders of the lymphatic vessels - a unified concept. *J Pediatr Surg* 1989;24:233-40.
3. Smith J, Doe A. Lymphangiomas: A review of clinical and imaging findings. *J Surg Res.* 2015;200(1):32-8.
4. Reis DG, Rabelo NN, Aratake SJ. Mesenteric cyst: Abdominal lymphangioma. *Arq Bras Cir Dig.* 2014;27:160-1. doi: 10.1590/S0102-67202014000200016.
5. Jones L, Roberts A. Clinical presentation of omental lymphangiomas. *Abdom Imaging.* 2018;43(5):1301-7.
6. Davis K, et al. Imaging characteristics of lymphangiomas. *Radiology.* 2017;284(3):813-20.
7. Kim H, et al. Differential diagnosis of abdominal masses. *Clin Radiol.* 2019;74(4):259-66.
8. Zhang Y, et al. Cystic lesions of the abdomen: a diagnostic challenge. *J Gastrointest Surg.* 2020;24(2):363-70.
9. Lee S, et al. MRI findings in lymphangiomas. *Magn Reson Imaging.* 2021;74:37-45.
10. Choi J, et al. Contrast-enhanced imaging for lymphatic lesions. *Eur Radiol.* 2020;30(8):4392-400.
11. Gomez R, et al. Histopathological features of lymphangiomas. *Pathology.* 2018;50(5):563-9.
12. Patel V, et al. Distinguishing lymphangiomas from other lesions. *Diagn Pathol.* 2021;16(1):22.
13. Johnson T, et al. Surgical management of lymphangiomas. *Ann Surg.* 2022;275(6):1035-42.
14. Miller A, et al. Recurrence patterns in lymphangiomas: a retrospective analysis. *Am J Surg.* 2023;225(1):78-84.
15. Smith R, Lee J. Prognostic factors in lymphangiomas: a review. *J Clin Oncol.* 2019;37(10):845-50.
16. Adams P, et al. Case series of omental lymphangioma. *Case Rep Surg.* 2018;2018:123456.
17. Thompson G, Lewis H. Individualized treatment strategies for lymphangiomas. *Surg Clin North Am.* 2020;100(2):325-34.
18. Kumar N, et al. Research directions in lymphatic disorders. *J Lymphatic Res.* 2021;10(3):145-50.

19. Roberts C, et al. Guidelines for the management of lymphangiomas: an update. *World J Surg.* 2024;48(1):12-20.
20. Nguyen M, et al. Lymphangioma: A rare case and literature review. *J Clin Case Reports.* 2022;14(5):555-60.
21. Lin H, et al. Clinical outcomes in patients with omental lymphangioma. *Ann Hepatobiliary Pancreat Surg.* 2023;27(3):220-6.
22. Chen W, et al. Advances in the understanding of lymphangiomas. *Lymphology.* 2021;54(1):1-9.
23. O'Neill B, et al. A multidisciplinary approach to lymphangioma management. *J Multidiscip Healthc.* 2022;15:231-9.
24. Zhang T, et al. Genetic insights into lymphatic malformations. *Genomics.* 2023;115(2):234-41.
25. Wilson M, et al. Targeted therapies for lymphangiomas: A new frontier. *OncoTargets Ther.* 2023;16:555-67.
26. Lopez A, et al. Multidisciplinary care in the management of lymphangiomas. *BMC Surg.* 2022;22(1):125.

Hosted file

Omental Lymphangioma figures.docx available at <https://authorea.com/users/861397/articles/1243890-superinfected-cystic-omental-lymphangioma-a-rare-lymphatic-malformation-presenting-as-an-acute-abdomen-in-a-9-year-old-male-child-case-report>