Case Report

ANNALS OF MEDICINE & SURGERY

OPEN

Dumble cystic lymphangioma as an underlying cause of vague abdominal complaints in a 2-year-old girl: case report

Khashayar Atqiaee, MD[a,*], Ali Samady Khanghah, MD[b], Leily Mohajerzadeh, MD[f], Ali Mardi, MD[d], Farhad Khayat Zahiri, MD[e], Sajjad Barin, MD[c]

Introduction: Lymphatic malformations (LM) refer to very rare hamartomatous benign lymphatic ectasias with an incidence of 1:250 000. They almost involve children more than adults, and among them, infants under one age are mostly affected. Macrocytic LM is found to be more than 2 cm in diameter or 2×2 cm^2 in volume. The proper treatment for mesenteric LM is complete surgical excision unless there is vital structure involvement.

Case presentation: The authors report a mesenteric macrocytic LM in a 2-year-old girl complaining of vague abdominal discomfort and persistent vomiting in which ultrasonography revealed a cystic masse with seromucous components. She then underwent exploratory laparotomy. The operation and the follow-up duration were uneventful.

Discussion: LMs are rare benign lesions of vascular origin with lymphatic differentiation, according to the latest International Society for the Study of Vascular Anomalies (ISSVA 2018). Under light microscopy, these malformations are characterized by their thin-walled endothelium and lymphatic tissue. These mobile lesions are incidentally found or appear with intestinal obstruction or acute abdomen scenarios.

Conclusion: Although benign, the LMs have the potential for invasion and recurrence. Thus, the examiner physician must keep such intra-abdominal lesions in mind.

Keywords: abdominal cystic lymphangioma, lymphatic malformation, mesenteric cyst, surgical exision

Introduction

Lymphatic malformations (LM), new terms for lymphangiomas, are very rare. They are hamartomatous benign lymphatic ectasias to true neoplasms, which have an incidence of 1:250 000[1]. They are commonly found in children, of which 60% appear at birth and 40% by the first year of infancy. Mesenteric MLs represent 5–6% of paediatric benign masses[2]. The abdominal cavity is a rare source of origin for these lesions. In almost all cases, the surgeon schedules the operation with the differential diagnosis list of bowel ischaemia or obstruction and congenital mesenteric or ovarian cysts in female patients. However, in some cases, mesenteric LMs tightly attached to the intestinal wall may necessitate intestinal resection and then anastomosis[3]. In this case, we report a 2-year-old female suffering from vague gastrointestinal complaints that underwent exploratory laparotomy with the final histopathological diagnosis of macrocytic LM. This work has been reported in line with the SCARE criteria[4].

HIGHLIGHTS

- Lymphangioma is the former term used for the lymphatic malformation.
- Lymphatic malformations (LM) refer to very rare hamartomatous lymphatic ectasias.
- The abdominal cavity is a rare source of origin for LMs.
- Macrocytic LM is found to be more than 2 cm in diameter or 2×2 cm2 in volume.
- The LMs must be completely excised in the cases of bowel obstruction or ischaemia.
- The resection may be challenging in the neurovasculature or vital organs invasion.

Case presentation

History

A 2-year-old girl presented to the paediatric emergency department complaining of restlessness, poor feeding, persistent

[a]Department of Pediatric Surgery, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, [b]Department of Surgery, [c]Department of Pathology, Fatemi hospital, [d]Department of pediatrics, Buo-Ali.Hospital, [e]Department of Anesthesiology, Imam Reza Hospital, School of Medicine, Ardabil University of Medical Sciences, Ardabil and [f]Pediatric Surgery Research Center, Research Institute for Children Health, Shahid Beheshti University of Medical Sciences, Tehran, Iran

Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

*Corresponding author. Address: Department of Pediatric Surgery, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran. Tel.: +985 138 713 801; fax: +985 138 709 201. E-mail: khashayaratqiaee@gmail.com (K. Atqiaee).

Annals of Medicine & Surgery (2023) 85:4041–4044

Received 27 December 2022; Accepted 25 February 2023

Published online 28 June 2023

http://dx.doi.org/10.1097/MS9.0000000000000395

vomiting, and a single episode of diarrhoea. There was no remarkable perinatal or past medical history.

Physical examination

In her first physical examination in the emergency department, the heart and respiratory rates were detected at 125 and 20 per min, accordingly. She was afebrile. The abdominal examination revealed asymmetry, especially in the right hypochondrium and hypogastric region. Bowel sounds were reduced, and palpation revealed a periumbilical and hypogastric tenderness besides non-voluntary abdominal rigidity. The haemoglobin concentration was 12.1 mg/dl without leukocytosis, and the electrolytes and liver transaminases were within normal limits.

Laboratory data and imaging study

Ultrasonography demonstrated a cystic lesion of 10×5 cm in diameter in the right inferior abdominal quadrant with extrinsic compression of the jejunoileal loops, probably mesenteric or arising from the right ovary cystic lesions.

Treatment plan

After hemodynamic resuscitation, the patient underwent exploratory laparotomy to resect the lesion. After supraumbilical transverse laparotomy, the surgical team initially encountered the cystic lesion. However, the mass was made of twin cystic components tightly attaching to the small intestinal mesentery with fine septations containing fluid [Figure 1]. In the middle part of the jejunum, in the antimesenteric region, a segment of the bowel was trapped between the two cysts. The twisted segment was affected in transmural ischaemia (arrows) [Figure 2] that was derotated and then resected with a margin of 2 cm accompanied by twin cysts [Figure 1]. Finally, an end-to-end anastomosis was performed. Other intra-abdominal organs appeared normal grossly. After all, the specimens were sent to pathology. One of the cysts was whitish-grey, containing milky proteinoid fluid, and another light yellow filled by a serosal connected the twins by the two fenestrae into the intestinal wall. The immunohistochemistry staining represented positive for D2-40 and factor VIII and negative for Calretinin and HMB45. The specimens were cut into multiple slices while epithelial cells with scattered lymphoid aggregation in the walls. There was no evidence of atypia or mitosis [Figure 3]. The diagnosis was compatible with macrocytic lymphatic malformation. The patient's weekly follow-ups until a month and then monthly were unremarkable.

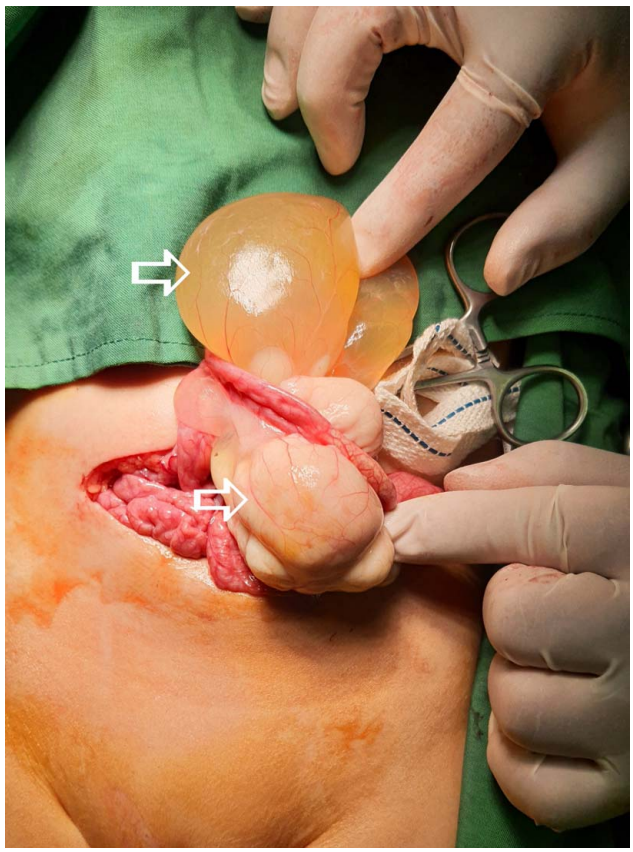

Figure 1. After laparotomy, the twin cystic lesions (hollow arrows) connecting to the intestinal wall appeared just 100 cm away from the ileocecal valve.

Discussion

Lymphatic malformations are new terms for a spectrum of rare benign lesions of vascular origin with lymphatic differentiation, according to the latest International Society for the Study of Vascular Anomalies (ISSVA 2018). Formerly, they were named lymphangiomas. Rokitansky, in 1842 for the first time, described abdominal cystic lymphangiomas as benign cystic developmental lesions of connective tissue origin[5]. These tumours involve less than 1% in the mesentery, omentum, and retroperitoneum[6]. Under light microscopy, these malformations are characterized by their thin-walled endothelium beside marginalized smooth muscle and collagen bundles, adipose cells, and lymphatic tissue. Immunohistopathologically, the endothelial cells are positive for CD31[7]. For the nature of being lymphoid, lymphocytic aggregation and the cysts filling milky, protein-rich fluid is not uncommon. Breaking down to arrange a normal lymphatic drainage net induces the dilatation of abnormal channels. This is supposed to be the pathophysiology of forming a cystic mass[8]. In comparison, some literature suggests that inflammation, abdominal trauma, surgeries in the abdomen, radiation, or lymphatic obstruction may be accused in the malformation genesis[9].

It is said that benign hamartomatous malformations are the best describing term instead of true neoplasms. The recurrence risk depends on the status of the margins[10]. In this case, the surgeon worked cautiously, whereas the margins were free of invasion.

The majority of cases detected are infants and children under 5 years old. Almost always, prenatal evaluations are incapable of diagnosis, as in our case. In a retrospective case study on 14 intra-abdominal cystic lymphatic malformations treated from 1996 to 2007 in the Pediatric Surgery Department at CHU Lapeyronie, Montpellier, France, the most common complaint was abdominal pain with or without a palpable mass. In only four out of fourteen cases, perinatal ultrasonogram was suspected of a visceral cystic lesion[11].

It could be an incidental discovery, and in addition, the essence of aqueous consistency, highly mobile, and slippery of the malformations makes them difficult to palpate. Unlike

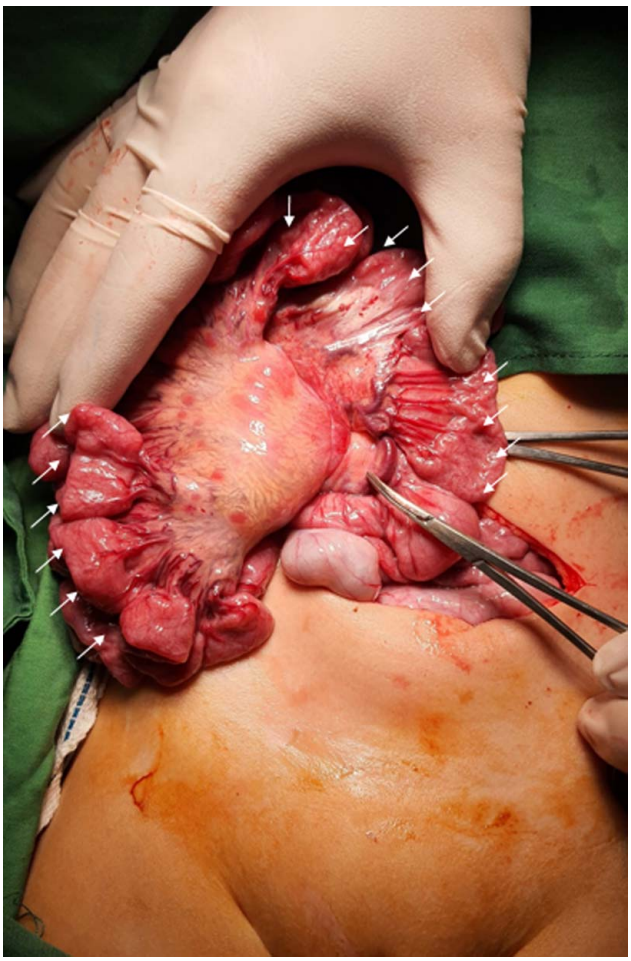

Figure 2. The mass effect causing gangrene (solid arrows) of about 20 cm of ileum.

retroperitoneal lymphatic malformations, which are strongly sessile, ones located in mesentery and omentum are mobile. Ultrasonography usually shows a multicystic septated lesion with well-defined thin walls. However, computed tomography reveals the origin of the cystic lesion, anatomical relationships, and mass size[12].

In the case of radiation exposure, the therapeutic team should conform to as low as reasonably achievable protocol. For this reason and for that, ultrasonography can demonstrate the malformation and stands as the first imaging modality[13]. They may present with acute abdomen or various digestive complications such as intestinal gangrene due to volvulus, cystic intracavitary haemorrhage, infection, traumatic rupture, and torsion of the cyst[14] as in this case that there was no way in front of the surgeon except resection of ischaemic trapped bowel segment of about 20 cm. There has already no malignant transformation been reported in the paediatric population. All serous or fluid-filled benign abdominal lesions such as intestinal duplication, ovarian, splenic, or choledochal cysts, cystic teratomas, hydronephrosis, hydatid cyst, and ascites are included in the differential diagnosis of paediatric abdominal macrocytic LM. Paediatric abdominal cystic masses may be surgically treated or radiologically followed up. Complete resection of the tumour remains the gold standard of the treatment that is achieved as simply as laparoscopic approaches are coming to work[15]. However, sometimes this could not be possible since proximity to vital vascular structures.

Conclusion

However, potentially invasive to vital organs, the lymphatic malformations are benign. Typically, the best therapeutic approach is complete excision, and the physician should think over the visceral mass pathologies besides common gastrointestinal diagnoses of childhood.

Ethical approval

This issue has been raised and approved by the ethics committee of Ardabil University of Medical Sciences, Iran with the code of IR.ARUMS.REC.1400.247

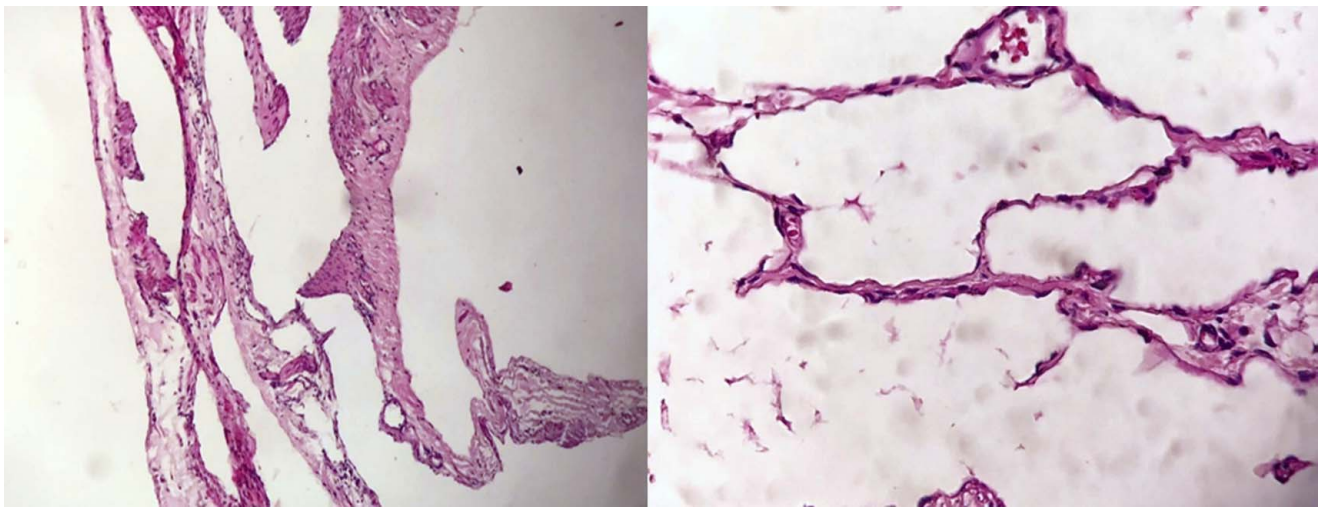

Figure 3. Hematoxylin and eosin-stained histopathologic specimen with ×100 on the left and ×1000 magnifying on the right showing vacant chambers of lymphatic fluid enclosed in epithelial tissue.

Consent

The consent in which the patient has allowed to use medical records and therapeutic information is attached to the medical document. The authors testify the patient privacy maintenance. Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request. The authors ensure that all the images/figures/photos are suitably anonymised with no patient information or means of identifying the patient.

Source of funding

This research received no specific grant from the public, commercial, or not-for-profit funding agencies.

Author contribution

K.A. operated on the patient and proposed the writing of the manuscript and the role of supervision. A.S.K. wrote the preliminary version of the manuscript, prepared the images, and regulated the necessary files. L.M. was in the role of supervision and scientific edition. A.M. initially visited the patient, made the preoperative diagnosis, and contributed to the manuscript preparation. F.K.Z. was involved in the preoperative preparation, during the surgery, and the postoperative stages as the anaesthesiologist and intensive care provider. S.B. was the clinical and anatomical pathologist who made the final diagnosis.

Conflicts of interest disclosure

The authors declare there are no conflicts of interest.

Research registration unique identifying number (UIN)

Name of the registry: -
Unique identifying number or registration ID: -
Hyperlink to the specific registration: -

Guarantor

The corresponding author accepts full responsibility for the work and approves the whole process from designing the study to publication.

Provenance and peer review

Not commissioned, externally peer-reviewed.

References

[1] Maranna H, Bains L, Lal P, *et al*. Cystic lymphangioma of the greater omentum: a case of partial spontaneous regression and review of the literature. Case Rep Surg 2020;2020:8932017.
[2] Mesić M, Cigit I, Zganjer M, *et al*. Cystic lymphangioma of jejunal mesentery mimicking acute appendicitis: case report. Acta Clin Croatica 2013;52:129–31.
[3] Chaudry G, Burrows PE, Padua HM, *et al*. Sclerotherapy of abdominal lymphatic malformations with doxycycline. J Vasc Intervent Radiol 2011;22:1431–5.
[4] Agha RA, Franchi T, Sohrabi C, *et al*. The SCARE 2020 guideline: updating consensus surgical CAse REport (SCARE) guidelines. Int J Surg 2020;84:226–30.
[5] Rokitansky K. Manual of pathological anatomy. Manual of special pathological anatomy. Braumiller & Seidel [Handbuch der pathologischen Anatomie: Bd Handbuch der speciellen pathologischen Anatomie. Braumiller & Seidel]; 1844;2:1842–1844.
[6] Takiff H, Calabria R, Yin L, *et al*. Mesenteric cysts and intra-abdominal cystic lymphangiomas. Arch Surg 1985;120:1266–9.
[7] de Perrot M, Rostan O, Morel P, *et al*. Abdominal lymphangioma in adults and children. J Br Surg 1998;85:395–7.
[8] Goh BKP, Tan YM, Ong HS, *et al*. Intra-abdominal and retroperitoneal lymphangiomas in pediatric and adult patients. World J Surg 2005;29:837–40.
[9] Daniel S, Lazarevic B, Attia A. Lymphangioma of the mesentery of the jejunum: report of a case and a brief review of the literature. Am J Gastroenterol 1983;78:726–9.
[10] Surlin V, Georgescu E, Dumitrescu C, *et al*. Retropancreatic cystic lymphangioma–considerations upon a case. Rom J Morphol Embryol 2011;52(1 suppl):493–6.
[11] Granata C, Lonati L, Scarsi PL, *et al*. Abdominal cystic lymphangiomas and mesenteric cysts: the clinical considerations. La Pediatr Med Chirurgica Med Surg Pediatr 1994;16:277–9.
[12] Chiappinelli A, Forgues D, Galifer RB. Congenital abdominal cystic lymphangiomas: what is the correct management? J Maternal-Fetal Neonatal Med 2012;25:915–9.
[13] Hebra A, Brown MF, McGeehin KM, *et al*. Mesenteric, omental, and retroperitoneal cysts in children: a clinical study of 22 cases. Southern Med J 1993;86:173–6.
[14] Onur MR, Bakal U, Kocakoc E, *et al*. Cystic abdominal masses in children: a pictorial essay. Clin Imaging 2013;37:18–27.
[15] Weeda VB, Booij KAC, Aronson DC. Mesenteric cystic lymphangioma: a congenital and an acquired anomaly? Two cases and a review of the literature. J Pediatr Surg 2008;43:1206–8.