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Case Report

Giant Cystic Lymphangioma as a Rare Cause of Acute Abdomen in Infancy: a Case Report and Literature Review

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Abstract

Lymphangioma, a benign mass of lymphatic origin, primarily affects the head, neck, and oral cavity, with the abdomen being an uncommon location. Clinical presentation is defined by site of origin and size. We present the case of a 10-month-old male infant with no previous medical history, admitted to our department with diffuse abdominal pain and distension. Physical examination revealed a huge, palpable mass arising from the left abdomen, abdominal tenderness, guarding, and rigidity. Abdominal ultrasound revealed a multilocular, non-measurable mass with both cystic and solid components that were suggestive of a giant cystic lymphangioma. Multidetector computed tomography and magnetic resonance imaging were performed to estimate the size and extension of the tumor for preoperative planning. Moreover, an elevated alpha-fetoprotein level further supported the neoplastic origin of the tumor. Exploratory laparotomy was performed, and a mass measuring approximately 13.9×16.7×15.4 cm was identified and completely excised in negative margins. Histopathological findings confirmed the initial diagnosis. Our patient had an uncomplicated postoperative course and recovery, with no signs of recurrence on a 6-month follow-up. Giant abdominal lymphangiomas in infants are extremely rare and close attention for complications is vital. Surgical intervention with complete resection is considered the optimal management.

Keywords

abdominal distension, abdominal mass, cystic lymphangioma, infant, giant lymphangioma

INTRODUCTION

Lymphangiomas are rare benign malformations of the lymphatic system, commonly diagnosed during the first few years of life. They are usually found in the cervical region, although they may originate from any region of the body, including the axilla, retroperitoneal space, thorax, trunk, pelvis, or extremities.[1,2] The incidence of abdominal lymphangiomas is even smaller and accounts for less than 9% of the total cases.[2,3] Clinical presentation and complications depend on the location and the size of the mass.[3,4] The diagnosis is based on ultrasonography and computed

tomography (CT).[5,6] Magnetic resonance imaging (MRI) may assist in preoperative planning of surgical resection as the definite treatment.[6,7] Complete resection, when possible, is necessary to avoid any further complications.[4,8,9] The following case report is presented in accordance with Surgical CAse REport (SCARE) guidelines.[10] Parents were fully informed, and written informed consent for publication of the current case report and accompanying images was obtained. We also review the relevant literature regarding the rarity, clinical presentation, and management of giant abdominal lymphangiomas on infants.

CASE REPORT

We present the case of a 10-month-old male patient with no significant previous medical or surgical history, who visited the Emergency Department of a tertiary care unit with worsening diffuse abdominal pain. The infant was born at full term by uncomplicated vaginal delivery and followed normal developmental milestones. Parents reported that the pain started five days ago, progressively worsened, and was also accompanied by abdominal distention with anorexia. The boy was afebrile and no other signs of gastrointestinal discomfort, such as vomiting, change in bowel habits or melaena, were reported.

During physical examination of the abdomen, the patient presented diffuse abdominal tenderness, guarding, and rigidity. Intestinal sounds were present and increased in right abdomen but were hypoactive in left abdomen. Rectal examination along with respiratory clinical examination was normal.

An abdominal ultrasound was performed, revealing a sizeable, non-measurable mass with both cystic and solid components and with septa located in the left abdominal cavity, while ascitic fluid was also present. Index laboratory investigation revealed leukocytosis (WBCs: 18.000/μL) with neutrophilia. Additionally, among tumor markers examined, alpha-fetoprotein was found exceptionally high (19.46 ng/dL, with normal values <9 ng/ml).

Multidetector computed tomography (MDCT) of the abdomen and retroperitoneal space showed a multilocular, cystic mass (13.9×16.7×15.4 cm) that occupied the greatest part of the peritoneal cavity, displacing the adjacent structures of the colon and small intestine without infiltrating them, the superior and inferior mesenteric arteries, and the superior aspect of the bladder, as well as the presence of ascitic fluid in the left paracolic space ***(Fig. 1)****. Additionally, MRI confirmed tumor dimensions and also revealed the retroperitoneal extension of the lesion, which is necessary for preoperative planning of adequate surgical resection* ***(Fig. 2)****.*

The patient proceeded to the operating theatre and exploratory laparotomy under general anesthesia was performed. After a transverse supraumbilical incision, a sizeable multilobular, fluid-filled cystic mass was identified in the left abdomen. The cystic mass was composed of four large cysts and multiple smaller ones, containing yellowish serous fluid. The two largest cysts were located in the paracolic space while the rest presented in association with the duodenojejunal junction and the pancreas ***(Fig. 3)****. Mesentery and omental anatomy were disturbed all over their course. Meticulous dissection permitted the complete excision of the tumor without capsular rupture* ***(Fig. 4)****. In addition, a 10-cm-long part of*

Figure 1. Computed tomography of a vast multilocular cystic mass involving the majority of the abdominal cavity.

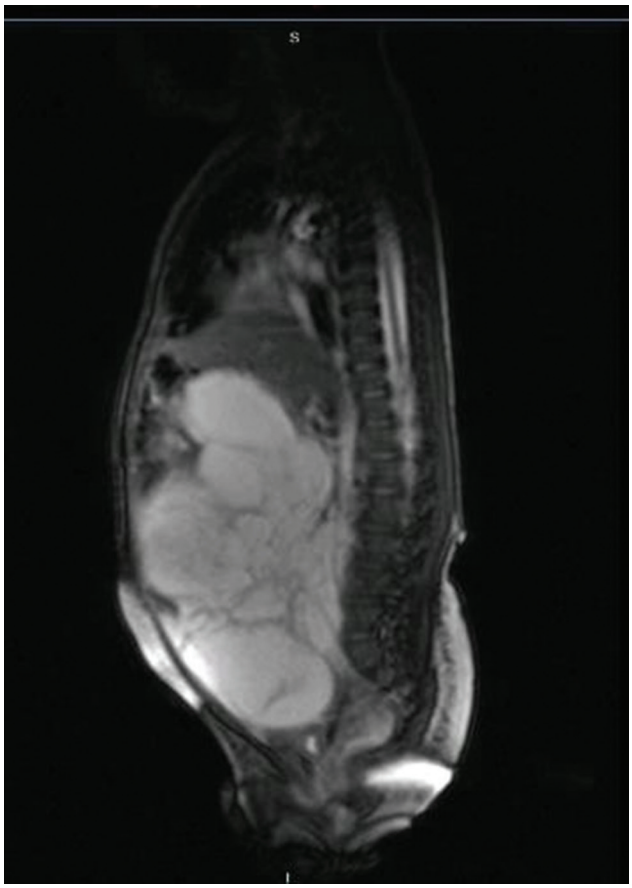

Figure 2. Magnetic resonance imaging (sagittal plane) of the abdominal mass where the multilocular character of the tumor and its extent are evident.

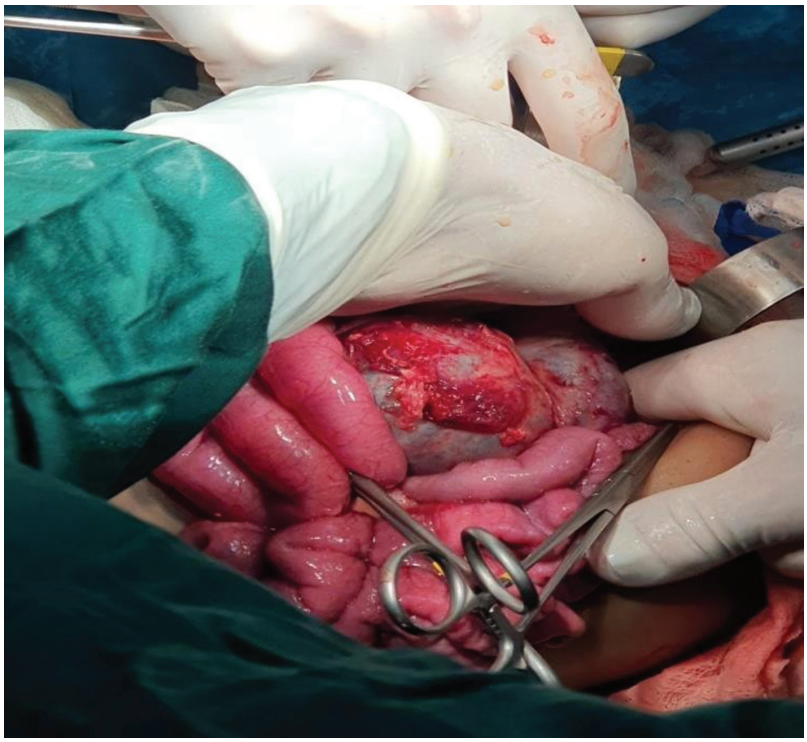

Figure 3. Image of the two paracolic large cystic masses identified through the transverse supraumbilical incision.

the sigmoid colon, which presented with disrupted vasculature of normal viability and absent mesosigmoid, was transfixed on the left lateral abdominal wall in order to minimize the risk of postoperative hernia or volvulus. Histopathological examination confirmed initial diagnosis of cystic lymphangioma. The postoperative course was uncomplicated, and the boy was discharged seven days after the procedure. At six-month follow-up, no signs of recurrence were detected.

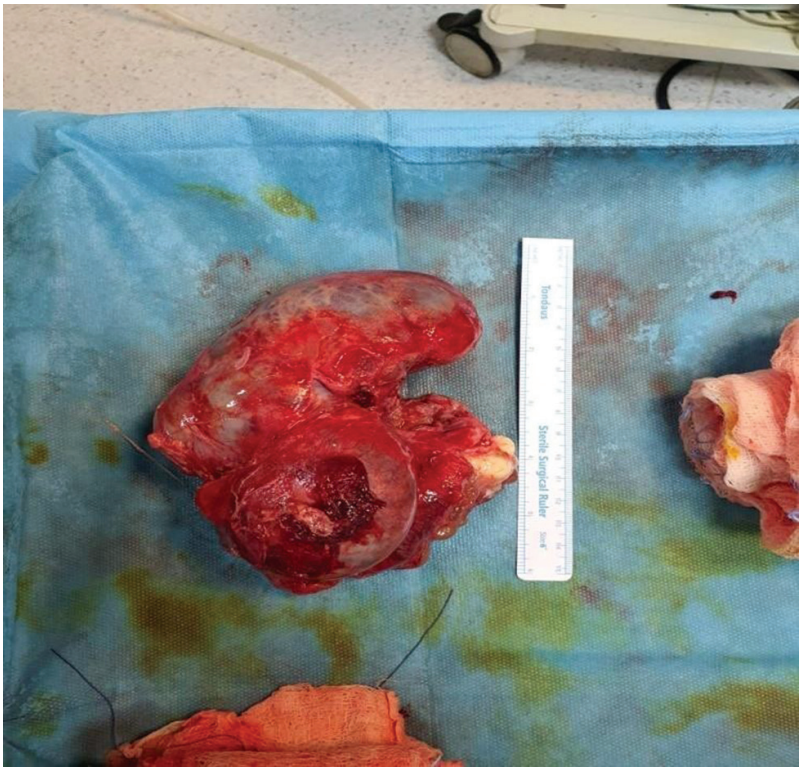

Figure 4. The resected surgical specimen without evidence of rupture.

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DISCUSSION

Lymphangiomas are rare congenital lesions of the lymphatic system generally classified into simple, cavernous, and cystic.[11] Categorization includes type I (pedicle), type II (sessile), type III (retroperitoneal), and type IV (multi-centric).[12] As for the etiologic mechanism, controversy still exists with regards to failure of lymphatic system development in the early fetal intrauterine life[13] or inflammation of lymphatic channels that may cause obstruction[14]. One theory suggests that cystic lymphangiomas are caused by the absence of communication of lymphatic channels in the small bowel, mesentery, or retroperitoneum with the main vessels, resulting in sequestration and cyst formation.[15]

The most common sites for cystic lymphangiomas are the cervical and axillary regions, which account for 95 percent of such lesions. In these cases, tumors are often referred to as "cystic hygromas" and are associated with genetic abnormalities, most commonly Turner syndrome.[15] Intra-abdominal cystic lymphangiomas are particularly rare encountered in 2%–8% of cases. Usual intra-abdominal locations are the small bowel (80%), the omentum, the mesocolon, and rarely, in less than 1% of cases, the retroperitoneal space.[16,17]

The clinical manifestation of patients with lymphangiomas located in the abdomen varies from absence of symptoms, almost in one-half of the cases, to chronic abdominal pain with or without distension and presence of a palpable abdominal mass. The presentation as acute abdomen, including acute abdominal pain, distension, vomiting, fever, and peritonitis, is less common and is associated with compression phenomena, intracystic hemorrhage, perforation, torsion, or rupture towards the adjacent structures.[18] In the presence of large cysts, preoperative imaging may give a false impression of ascitic fluid collection.[19]

According to Li et al, diagnostic accuracy of up to 90% is achievable with both ultrasonography and abdominal CT scan.[20] Abdominal ultrasound is used as a first-line diagnostic procedure, revealing a well-defined cystic or multicystic mass with internal septations.[21] However, abdominal CT scan is the diagnostic tool most commonly used, especially in cases of acute symptomatology, for investigation of tumor origin, anatomical correlations, complications and the precise size of the mass.[7,19] Nevertheless, recent studies suggest that MRI is also the preferable diagnostic method to determine how the mass is associated with adjacent structures, providing benefit for planning of the surgical resection, although unable to distinguish between a lymphocele, cystic teratoma, cystic lymphangioma, or a dermoid cyst.[3] As a result, preoperative diagnosis combining both CT and MRI scan accomplish better results.[6]

Despite its benign character, an abdominal cystic lymphangioma can become locally invasive; thus, complete surgical resection is the treatment of choice, especially if there are signs of mass enlargement or acute symptoms. Percutaneous or intraoperative aspiration should be avoided due to high rates of recurrence, while in case of bowel involvement, limited intestinal resection is suggested.[22] Furthermore, successful laparoscopic resection of large abdominal cystic lymphangiomas has also been reported in recent years in selected patients.[23,24] However, open surgical management provides the ability of thorough examination and removal of the lesion, especially in cases in which the size and extent of the mass is substantially large, as in our patient.

Two case series referring to pediatric population found that the location of the lesion had no effect on the outcome but increased the surgical procedure in patients whose jejunal mesentery was affected by lymphangiomas.[25,26] Since partial resection may result in recurrence, radical excision is the preferred course of treatment. Particularly, in cases of incomplete resection, long-term follow-up is necessary due to the possibility of recurrence of approximately 10%.[27] Postoperative prognosis after complete resection is excellent. In addition, gradual malignant transformation to lymphangiosarcoma has been described.[28]

CONCLUSION

The differential diagnosis of an infant presenting with acute abdomen may involve a variety of conditions. Medical history accompanied by thorough clinical evaluation and additional information provided by the laboratory (tumor markers) and imaging studies (abdominal ultrasound, CT, MRI) may provide a prompt and accurate diagnosis of cystic lymphangioma. Immediate and meticulous surgical resection of the entire cystic mass is usually safe with no complications. Infants with large cystic lymphangiomas, completely resected, as was our case, can demonstrate an uncomplicated postoperative course, with excellent prognosis. This case report and literature review highlights aspects of the clinical presentation that might increase clinical suspicion for preoperative diagnosis.

Author contributions

C.M., E.B., and A.P. designed the research; C.M., V.M., M.P., I.G., and E.A. drafted the manuscript; E.C., I.C., and G.P. supervised the paper. All authors read and approved the final manuscript.

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Conflicts of Interest

All authors declare that they have no conflict of interest.

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Гигантская кистозная лимфангиома как редкая причина острого живота у младенцев: описание случая и обзор литературы

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Резюме

Лимфангиома, доброкачественная масса лимфатического происхождения, в первую очередь поражает голову, шею и ротовую полость, а живот является редкой локализацией. Клиническая картина определяется местом происхождения и размером. Мы представляем случай 10-месячного младенца мужского пола без предшествующей истории болезни, поступившего в наше отделение с диффузной болью в животе и вздутием. Физическое обследование выявило огромную, пальпируемую массу, исходящую из левой части живота, болезненность живота, защитное напряжение и ригидность. УЗИ брюшной полости выявило многокамерную, неизмеримую массу с кистозными и солидными компонентами, которые предполагали гигантскую кистозную лимфангиому. Для оценки размера и распространения опухоли для предоперационного планирования были выполнены мультидетекторная компьютерная томография и магнитно-резонансная томография. Более того, повышенный уровень альфа-фетопротеина дополнительно подтвердил неопластическое происхождение опухоли. Была проведена диагностическая лапаротомия, и была обнаружена масса размером приблизительно 13.9×16.7×15.4 cm, которая была полностью удалена в отрицательных краях резекции. Гистопатологические данные подтвердили первоначальный диагноз. У нашего пациента послеоперационный период протекал без осложнений и выздоровление, без признаков рецидива при последующем наблюдении в течение 6 месяцев. Гигантские абдоминальные лимфангиомы у младенцев встречаются крайне редко, и пристальное внимание к осложнениям имеет жизненно важное значение. Хирургическое вмешательство с полной резекцией считается оптимальным лечением.

Ключевые слова

вздутие живота, брюшная масса, кистозная лимфангиома, младенец, гигантская лимфангиома