Case Report تقرير حالة طبية

GIANT CYSTIC LYMPHANGIOMA OF THE ILEAL MESENTERY IN A FEMALE CHILD: CASE REPORT

ورم وعائي لمفاوي كيسي عرطل في مساريقا الدقاق عند طفلة: تقرير حالة طبية

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ملخص الحالة

الورم الوعائي اللمفاوي هو ورم سليم ينجم عن شذوذات تحدث خلال تطور الجهاز اللمفاوي، تشاهد هذه الأورام عادةً عند الأطفال والبالغين الشباب وهي نادرة الحدوث في البطن. تم في هذا التقرير وصف حالة طفلة عمرها 11 سنة تشكو من كتلة بطنية كبيرة مرئية، أظهرت الدراسة بالأمواج فوق الصوتية والتصوير الطبقي المحوسب CT قبل العمل الجراحي وجود كتلة كبيرة كيسية متعددة الأجواف تشغل الحيز داخل البريتوان. بفتح البطن تبين وجود كتلة كبيرة كيسية على حساب مساريقا الأمعاء الدقيقة، تم استئصالها كاملاً مع جزء من الدقاق المجاور، أظهر الفحص النسيجي التشريحي المرضي للكتلة المستأصلة كونها ورم وعائي لمفاوي.

ABSTRACT

Lymphangiomas are rare benign tumors originating due to abnormalities that occur during the development of lymphatic system. It is usually seen in children and young adults. The incidence of intra-abdominal lymphangiomas is very low. We herein describe an 11-year-old female patient presented with a huge visible lump in the abdomen. Preoperative studies, including abdominal ultrasonography and CT imaging, showed a very large multilocular cystic mass occupying the peritoneal cavity. Surgical exploration revealed a large cystic tumor of the small bowel mesentery. Total resection of the mass and the adjacent ileum was done. Histopathologically, the tumor proved to be a cystic lymphangioma.

INTRODUCTION

Lymphangiomas are rare benign tumors of lymphatic origin. They are preferentially located in the head, neck and axilla in children. However; lymphangiomas in the peritoneal cavity are extremely rare.

In the abdomen, they are frequently of cystic type and most commonly located in the small bowel mesentery. Next in frequency come omentum, mesocolon, and retro-peritoneum.

The clinical presentation of intra-abdominal cystic lymphangiomas (ICLs) is variable and nonspecific, and usually unhelpful in establishing the exact diagnosis. Abdominal ultrasonography (US) and

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computed tomography (CT) very useful in delineating its cystic nature and providing important information regarding its size, location, and the involvement of adjacent organs. The definite diagnosis of lymphangiomas is made by pathologic study, whereas radical excision is the optimal treatment. This report describes a child with a giant lymphangioma of the ileal mesentery.

CASE PRESENTATION

An 11-year-old female patient presented with progressive asymmetric abdominal distension and mild abdominal pain. She was otherwise symptom free. Her past medical history was uneventful, and there was no prior abdominal surgery or trauma. She had no family history of malignancy or congenital abnormalities.

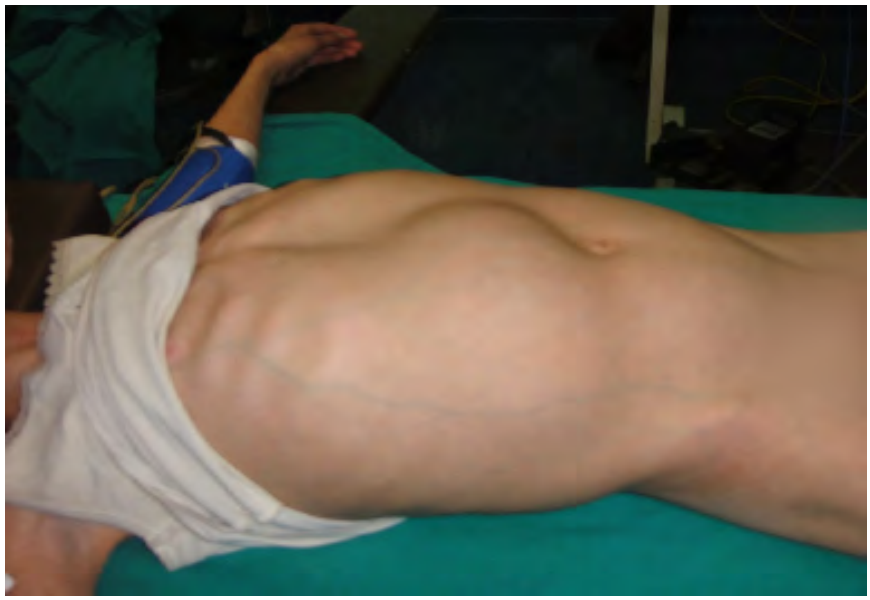

Figure 1. Visible abdominal lump.

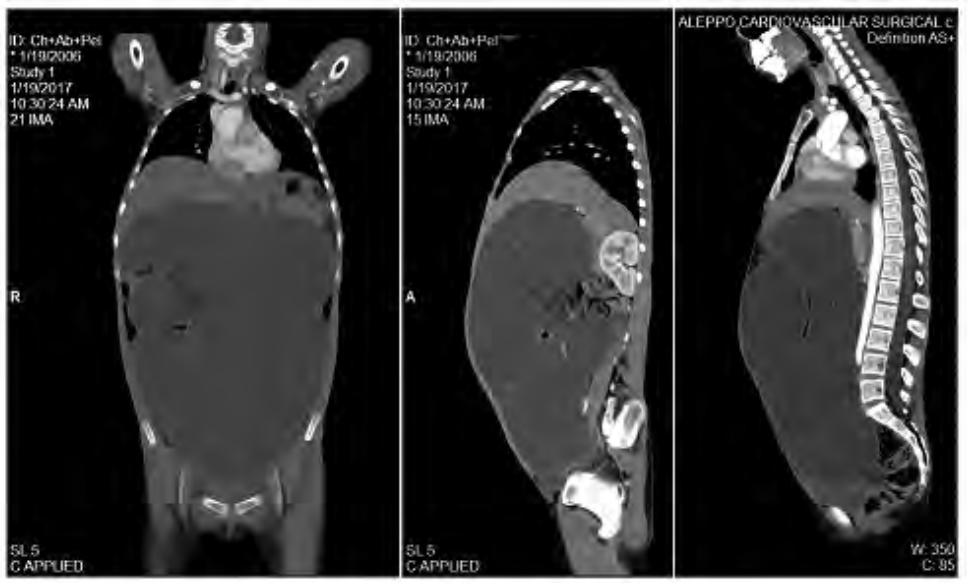

Figure 2. Abdominal CT scan showing large cystic mass.

Her physical examination revealed a very large soft abdominal mass, Figure 1.

Ultrasonography (US) and computerized tomography (CT) revealed a cystic mass filling the whole abdominal cavity. The mass was multilocular containing homogeneous fluid and showing displacement of intra-abdominal viscera. The largest cyst was 25 cm in diameter, Figure 2.

Exploratory laparotomy by muscle-cutting transverse upper abdominal incision was done, and

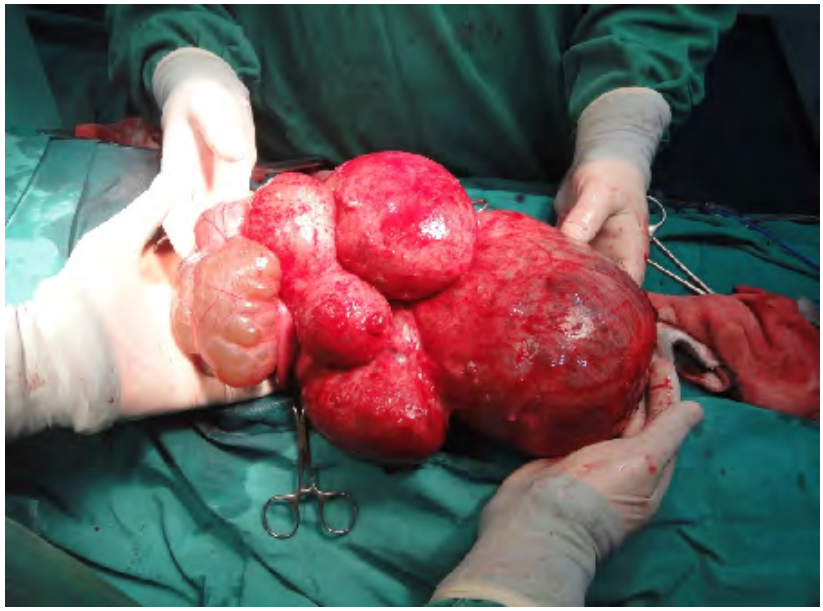

Figure 3. Intra-operative appearence of a huge cystic mass.

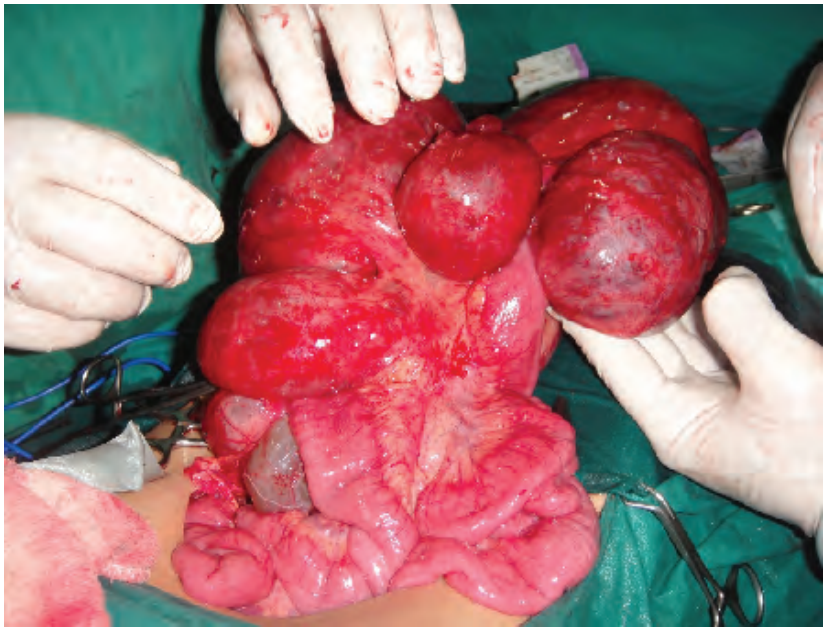

Figure 4. Mesenteric cystic mass infiltrating the ileal wall.

it showed that the huge multicystic mass involved the mesentery of ileum and infiltrated to the ileal wall, Figures 3 and 4. Surgical excision of the mass together with 25 cm of the ileum was done, and the continuity of the ileum was established by end to end anastomosis.

The child's postoperative course was uneventful, and she was discharged home on the 5th postoperative day. Histology of the mass showed mesenteric cystic lymphangioma with extension to the bowel walland a free surgical border (complete excision).

DISCUSSION

Cystic lymphangioma (CL) is a rare malformation first described by Koch in 1913.[1] CLs are usually found in the neck (75%, also called cystic hygromas) and axilla (20%).[2] Intra-abdominal cystic lymphangiomas ICLs are rare and comprise less than 5% of all cystic lymphangiomas, corresponding to an incidence between 1:27,000 and 1:250,000.[3] Cystic lymphangiomas represents 7% of abdominal cystic lesions in adults.[4] This type of lymphangiomas is generally detected in the first decade of life.

The etiology of lymphangiomas is probably a congenital abnormality of the lymphatic system, causing sequestrations of lymphatic tissue during embryologic development.[5,6] This theory would explain why lymphangiomas occur primarily in children. However; it is suggested that abdominal trauma, lymphatic obstruction, inflammatory process, surgery, or radiation therapy may lead to the secondary formation of such a tumor.[7-9]

Traditionally, lymphangiomas are classified as capillary, cavernous, or cystic. The capillary lymphangiomas are usually situated superficially in the skin and composed of small thin-walled lymphatic vessels. The cavernous lymphangiomas, however, are composed of dilated lymphatic vessels and lymphoid stroma, and have a connection with spaces of various normal adjacent lymphatics. Lastly, the cystic lymphangiomas consist of lymphatic spaces of various sizes that contain fascicles of smooth muscle and collagen bundles, but have no connection with adjacent normal lymphatics. It is, however, not always easy to differentiate between cystic and cavernous lymphangiomas because cystic lymphangiomas may also contain cavernous areas.[5,10]

Clinical presentation of abdominal lymphangiomas is variable with no pathognomonic signs and symptoms. Generally they are asymptomatic with the first symptoms being abdominal distention, mild abdominal pain, and abdominal asymmetry due to an enlarging mass.[11]

In some cases, they are revealed incidentally during abdominal examination or radiology for another abdominal condition. However, there are several reported cases of lymphangiomas leading to surgical emergencies, such as hemorrhage, bowel obstruction, ureteric obstruction, and sepsis.[12-18] Our patient presented with a huge and visible abdominal lump causing a mild abdominal discomfort only.

Preoperative diagnosis of intra-abdominal lymphangiomas is usually difficult, with Ultrasonography, CT, and MRI, being crucial modalities in delineating ICLs:

Ultrasound examination is useful initially and typically shows a cystic lesion with multiple thin septa, containing hypoechoic fluid. Sometimes the cyst may be complicated by an intracystic hemorrhage, causing a partially echogenic content.[19,20]

On CT imaging, they appear as a uni- or multilocular mass with enhancement of the wall and septum by contrast medium. The fluid component is typically homogeneous with low attenuation values, but the density is lower in case of chylous content and denser if lesion is complicated by hemorrhage.[21] These studies (US and CT) help to determine, if the tumor is cystic as well as its size and location, but they are insufficient to establish an accurate diagnosis preoperatively.

On the other hand magnetic resonance imaging is the most useful preoperative radiological tool for diagnosis and in surgical planning.[22,23]

The definitive treatment for ICL is radical excision,

even when asymptomatic. But, with increasing tumor size, radical resection becomes more difficult and local recurrence more probable. In our patient, resection of 25 cm of the adjacent bowel was necessary.

The ICLs can be excised using laparoscopic techniques.[24,25] The laparoscopic approach to resection of mesenteric cysts was first described by Mackenzie in 1993.[26] Partial drainage of the cyst may be necessary to confirm the site of origin of the cyst and allow its removal through the umbilical port.

For those cases requiring a concomitant bowel resection, either an intra-corporeal technique or a laparoscopic-assisted extracorporeal technique can be used.

If resection is not possible, the other option is partial excision with marsupialization of the remaining cyst into the abdominal cavity. Approximately 10% of patients require this form of treatment. If this procedure is done, the cyst lining should be sclerosed with 10% glucose solution,[4] electrocautery, or tincture of iodine in an attempt to minimize recurrence.[27]

Intra-cystic administration of sclerosing agents, such as bleomycin, OK432, and doxycycline, is regarded as an adjuvant therapy in unresectable lesions.

Percutaneous injection of the lyophilized incubation mixture of group A Streptococcus pyogenes OK432 has been successful in the treatment of large, nonresectable lymphangiomas in children.[26] The mechanism of action of OK432 seems to be related to activation of the white cells (an increased number of natural killer cells and T cells), and an increase in cytokine-mediated endothelial permeability (increased activity of tumor necrosis factor and interleukin-6), resulting in shrinkage of the cystic spaces.[26] This agent may be useful to complement the surgical treatment of mesenteric cysts extending into the retroperitoneum.[27]

The characteristic histological criteria of cystic lymphangioma are presence of lymphatic spaces lined by flat endothelial cells, lymphoid aggregates and foam cells containing lipoid material in stroma and smooth muscle fibers in their walls.

The recurrence rate following complete resection ranged from 0% to 14% and malignant degeneration is very rare.[28]

CONCLUSIONS

The ICL is a benign tumor of the lymphatic system mostly affecting young children. The diagnosis is often suggested by ultrasound or CT scan. Mesenteric lymphangiomas including giant ones are almost usually amenable to radical excision. Nevertheless, it is sometimes necessary to sacrifice the part of bowel infiltrated with the tumor if any.

Unless access to the distal colon and rectum is required, the muscle-cutting transverse upper abdominal incision is suitable for most surgical interventions in infants and children including ICL resection.

Complete excision of the tumor (ICLs) with/without intestinal resection is mandatory to prevent recurrence and long-term prognosis is generally excellent.

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