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Case Report

Surgery Section

Inflammation-induced Giant Retroperitoneal Lymphangioma in an Adult: An Unusual Case Report

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ABSTRACT

Cystic lymphangiomas belong to the group of tumours arising from the lymphatic system. In adults, they are less commonly seen, with the abdomen being the least common site. Here, the present case study included a 32-year-old female who had a history of tube drainage of an abdominal cystic mass six years ago at the same hospital. Following that procedure, she developed tachycardia clinically, along with fever and rigors. The patient now presented with abdominal pain and distension for five months. She was evaluated with an ultrasound and a Computed Tomography (CT) scan of the abdomen and pelvis, which revealed a similar retroperitoneal cystic lesion. The patient underwent complete excision of the cystic lesion, and her postoperative course was uneventful. We are reporting this case of a lymphangioma that became iatrogenically infected and resulted in a giant lymphangioma within six years. According to the literature, only three cases of retroperitoneal lymphangioma in adults with a size greater than 20 cm have been reported. Compared to these, this cystic lymphangioma represents the fourth largest reported case.

Keywords: Cystic, Histopathology, Infection, Surgery

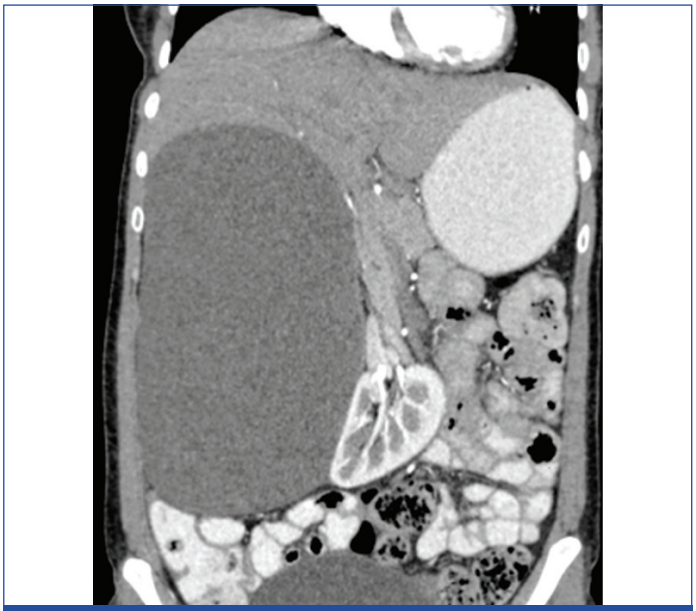
CASE REPORT

A 32-year-old female presented with abdominal distension associated with pain for five months. The pain was insidious in onset, diffuse in nature, and involved all abdominal quadrants. She had previously visited several local practitioners but was treated only symptomatically.

When she visited the hospital, she reported a similar episode six years earlier. At that time, an Ultrasound (USG) of the abdomen revealed a cystic mass. A USG-guided pigtail insertion was performed during the same admission, which drained the entire cystic mass. Within 48 hours of the procedure, the patient developed fever with rigors and clinically significant tachycardia. She was treated symptomatically with intravenous antibiotics until her fever subsided, over approximately one week, followed by an additional week of observation. Cytology of the drained fluid was negative for malignant cells. Her recovery was delayed, with intermittent fever episodes, and she was discharged after two weeks. The patient was subsequently lost to follow-up, and further details from that period were not available.

On current examination, there was significant abdominal fullness extending from the right hypochondrium to the right iliac region. Ultrasound revealed a large retroperitoneal cystic lesion with internal echoes, measuring 22×11×12 cm on the right side of the abdomen. A contrast-enhanced CT scan of the abdomen and pelvis showed a well-defined, thin-walled cystic lesion in the retroperitoneum involving the right hypochondriac, right lumbar, and right iliac fossa regions, measuring 22×11×13 cm, with no solid component [Table/Fig-1,2]. There was no evidence of free fluid or enlarged retroperitoneal lymph nodes. The cystic lesion was compressing and displacing the liver superiorly, the ascending colon and hepatic flexure inferiorly, and the right kidney inferomedially.

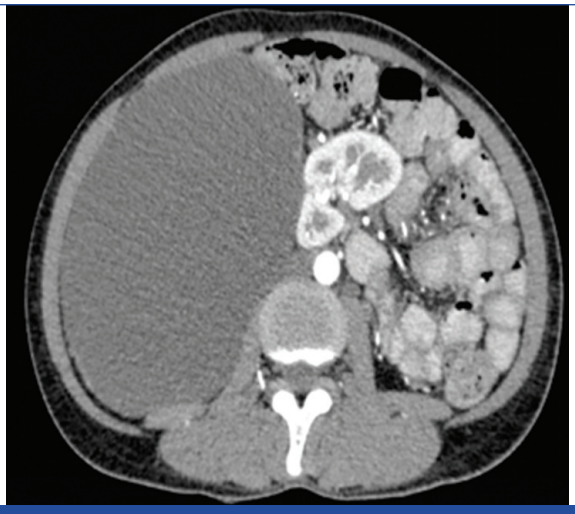
Following evaluation, the patient was scheduled for laparoscopic retroperitoneal excision of the cystic mass. The surface of the abdomen was marked preoperatively to delineate the extent of the mass [Table/Fig-3]. Due to extensive adhesions, the procedure was converted to open surgery. Intraoperatively, the cystic lesion was noted to extend cranially up to the right hemidiaphragm and caudally to the right psoas muscle, involving the right hypochondriac, right lumbar, and right iliac fossa regions [Table/Fig-4]. The lesion was adherent to the pelvic peritoneum, the ascending and transverse colon, and the right kidney. The right kidney was completely displaced medially, with evidence of compression on the right renal artery and vein.



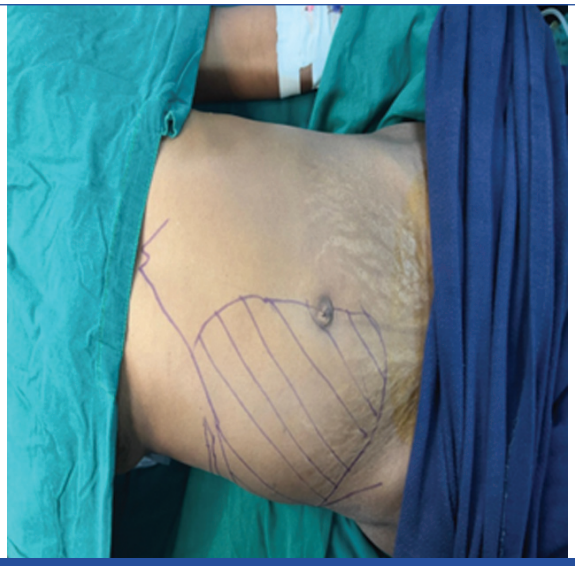
[Table/Fig-1]: Contrast enhanced CT scan coronal view showing the large cystic lesion displacing the kidney medially and liver superiorly.

Mobilisation of the cyst was started from caudal to cranial to relieve the compression on the right kidney. Careful dissection was performed near the renal hilum to preserve renal blood supply. After the cyst was completely freed from the renal vasculature, the right kidney was repositioned to its original location using a derotational maneuver. Following release of adhesions from the right lateral abdominal wall and transverse colon, the final portion of the cyst was delivered from the subdiaphragmatic region. During the last stage of excision, the liver was adequately retracted, and the tail end of the cyst was excised after releasing adhesions from the right hemidiaphragm.

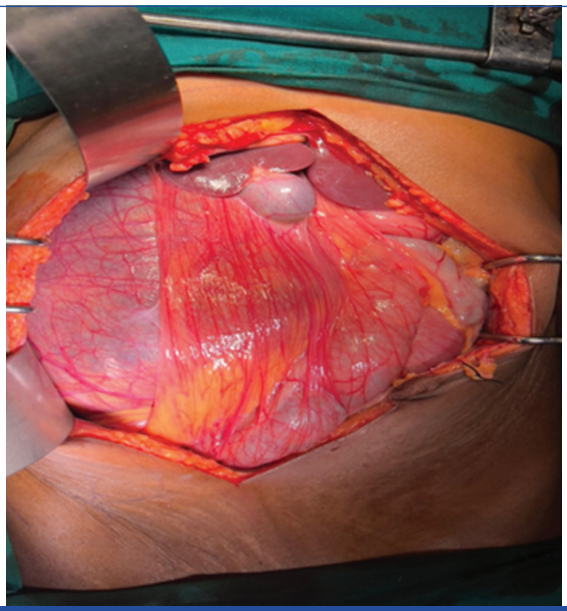
The patient had an uneventful postoperative course and was discharged on postoperative day seven.



[Table/Fig-2]: Contrast-enhanced CT scan axial view showing well-defined thin-walled cystic lesion in the retroperitoneum, compressing the bowel loops inferiorly and laterally.



[Table/Fig-3]: Surface marking of the cystic tumour.



[Table/Fig-4]: Intraoperative image of the cystic tumour.

Histological examination of the cyst revealed a cyst wall lined partly by cells with attenuated nuclei and partly by histiocytes, lymphocytes, and macrophages. There was also evidence of degenerative changes and fibrosis, consistent with lymphangioma. This diagnosis was later confirmed by immunohistochemistry, which showed positivity for CD31.

The patient has been on regular follow-up in the outpatient department, with the last follow-up six months post-surgery, showing no evidence of recurrence on recent scans.

DISCUSSION

Among cystic lymphangiomas occurring in adults, only about 7% arise in the abdominal region [1]. The most commonly involved abdominal sites are the mesentery, spleen, liver, and gastrointestinal tract, while the retroperitoneal region accounts for only about 1% of cases. These tumours are usually asymptomatic at presentation and only produce symptoms once they become large enough to cause compression of surrounding structures [2].

Several theories have been proposed regarding the formation of cystic lymphangiomas, including inflammation, trauma, and fibrosis, which can lead to congenital malformations characterised by a lack of communication between the main lymphatic channel and surrounding channels [3]. Infection can recruit M2-polarised macrophages, which secrete pro-angiogenic factors and accelerate the progression of lymphangioma [4].

In a recent study of 64 patients with adult primary retroperitoneal tumours, Mansour et al., reported that only 17% of patients had unilocular cystic lesions, with just 4% located on the right side of the abdomen, and an average tumour size of 9.3 cm [5]. According to the literature, only three case reports describe retroperitoneal lymphangiomas in adults exceeding 20 cm in size: Shayesteh et al., (29 cm), Hubli et al., (40 cm), and Su et al., (27.8 cm) [6-8]. Compared to these, the present cystic lymphangioma (22×11×13 cm) represents the fourth largest reported case.

Lymphangiomas are classified based on the size of ductal dilation into cystic, cavernous, and simple types. Retroperitoneal lymphangiomas are hypothesised to occur due to atypical connections between the venous and lymphatic channels in the retroperitoneum [9].

Zhang et al., demonstrated that M2-polarised macrophages are recruited to infected lymphangiomas, which are rich in these cells [4]. These macrophages secrete pro-angiogenic factors that promote lymphangioma growth. Following the acute phase of infection, antibiotic penetration is poor in lymphangiomas, leading to chronic inflammation. This results in the formation of tertiary lymphoid organs, which recruit M2-polarised macrophages. Excessive production of pro-angiogenic factors accelerates the proliferation of lymphatic malformations. Lymphotoxins and lymphotoxin-related inducible ligands within these tertiary lymphoid organs contribute to the rapid growth of lymphatic malformations [10].

Maghrebi et al., reported a series of 32 cases of abdominal retroperitoneal lymphangiomas, with a recurrence rate of 9.4% [11].

CONCLUSION(S)

This article highlights that chronic inflammation can contribute to the development of giant lymphangiomas due to a sustained inflammatory response in a controlled environment, even without external signs. Percutaneous drainage of lymphangiomas should be avoided; if performed, it should be done by a trained interventional radiologist under strict aseptic conditions. Infection in lymphangiomas can lead to the formation of tertiary lymphoid organs, accelerating the growth of the lesion.

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