



## A Rare Case of Retroperitoneal Lymphatic Cyst in Adult

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### Abstract

*Retroperitoneal lymphangioma is a rare, benign mesodermal tumor arising from the retroperitoneal lymphatics which usually presents in infancy and it is worthy to report a case when it has presented in an adult. Apart from being a rare entity it also presents as a diagnostic dilemma and final diagnosis is often made on surgical exploration. We report a case of retroperitoneal lymphangioma in a 58-year-old female who presented with abdominal distension and dull aching abdominal pain with loss of appetite. Radiological imaging revealed large globular cystic lesion of size 175mm x 160mm x 115mm having well defined margin in the right side of abdominal cavity extending from subhepatic area to the right iliac region. It appears separate from the right kidney, liver and major vessels. Surgical removal of the cyst was accomplished without any incident. A benign cystic retroperitoneal lymphangioma was diagnosed on histology and confirmed with immunohistochemical stains.*

**Keywords:** Retroperitoneum, Cystic, Lymphangioma.

### Introduction

Lymphangiomas are rare cystic tumors of the lymphatic system. These are benign, slow-growing lesions characterized by proliferating lymphatic vessels <sup>[1]</sup>. Most (95 %) occur in the neck and axillary regions, the remaining 5% are located in the mesentery, retroperitoneum, abdominal viscera, lung, and mediastinum <sup>[2]</sup>. Retroperitoneal cystic lymphangioma is a rare benign tumor of the retroperitoneal lymphatics that usually manifests in infancy. It is worth to report an unexpected presentation specially in adult. Symptoms may vary depending on the exact location and size of the cyst. Retroperitoneal lymphangiomas are often asymptomatic and are

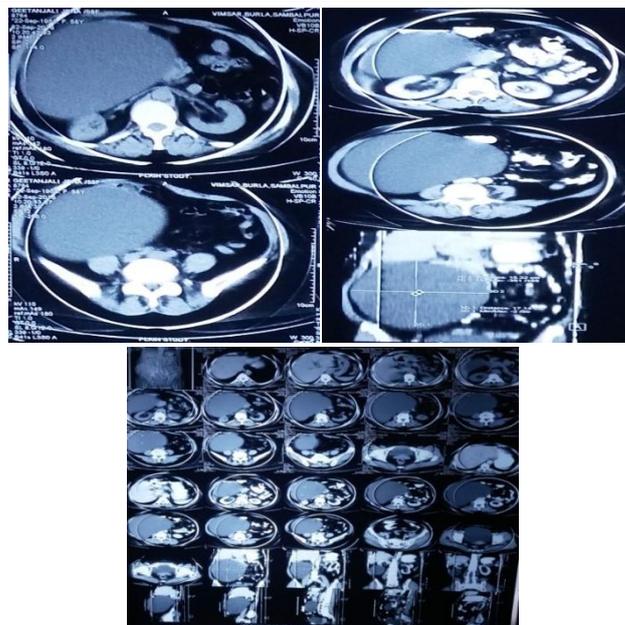
usually detected intraoperatively or during imaging for some other conditions<sup>4</sup>. Less commonly when the cyst is large, patient may present with abdominal distension, pain, fatigue, and weight loss. A large cyst which undergoes torsion, hemorrhage, and rupture may present as acute abdomen. When presenting as a palpable abdominal mass they are easily confused with other cystic tumors including those arising from the liver, kidney, and pancreas. Imaging studies are often inconclusive in differentiating cystic lymphangiomas from other cystic lesions and surgery or diagnostic laparoscopy is most frequently required for definitive diagnosis and management<sup>5</sup>. Because of the rarity and the

diagnostic dilemma that it poses, we are reporting a case of retroperitoneal cystic lymphangioma in a 65-year-old male.

**Case Report**

We report a case of a 58 years old Hindu lady, presented with a mass in the right side of the abdomen for 4 months, associated with abdominal distension, loss of appetite and discomfort. Not associated with fever, weight loss. She was a known case of hypertension, on antihypertensive medication. Her bladder and bowel habits were normal. She had undergone bilateral tubal ligation and open cholecystectomy 30 years back. She is multiparous and attained menopause 10 years back.

On Examination, a soft cystic mass of size 15cm X 15cm, non tender, fluctuant, mobile with well defined margin, extending from iliac to lumbar and hypochondrium on right side of abdomen. On carnette test it was found as intra abdominal. There is no pulsation or bruit herd. Ultrasonogram finding shows large globular cystic lesion of size 175mm x 160mm x115mm having well defined margin in the right side of abdominal cavity extending from subhepatic area to the right iliac region. It appears separate from the right kidney, liver and major vessels. Bowel loops appears to be displaced by the lesion.

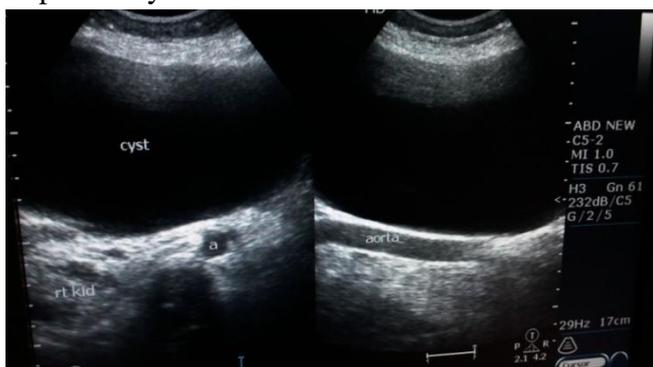


**FIG 2a, 2b, 2c:** Showing retroperitoneal lymphatic cyst in CT scan.

Then we planned for exploratory laparotomy and excision of cyst. Intraoperatively it was found as large cystic lesion containing clear fluid occupying the right suprarenal and renal area. The fluid was aspirated and the cyst was excised.



**FIG 3:** Showing the cyst intraoperatively.

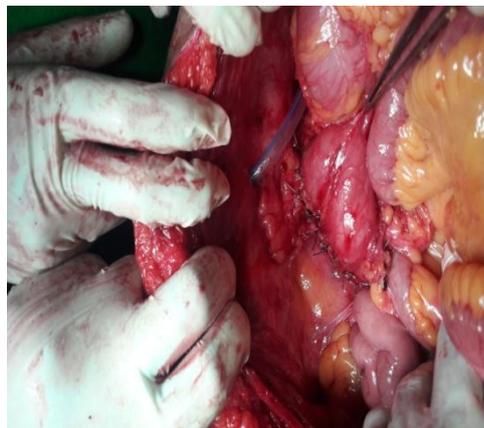


**FIG:1** Showing ultrasound appearance of cyst.

CT scan showed a well defined simple cystic lesion of size 17x15cm noted in the right side of abdominal cavity. It appeared separate from all other visceral organs. with this clinical and investigative findings it was diagnosed as retroperitoneal lymphatic cyst.



**Fig 4a**



**FIG 4 b**

**FIG 4 a , 4 b :** Showing the complete excision of cyst from retroperitoneum and other organs not affected.

Postoperatively the patient recovered well and was discharged.

### Discussion

Diseases of the abdominal lymphatic system can be categorised as those that are primarily lymphatic in origin and those secondary to other diseases. The primary disorders of the lymphatic system include congenital or developmental conditions, such as lymphangiomas, lymphangiomatosis, lymphatic agenesis, or dysplasia. Lymphangiectasia secondary to infections and lymphocoeles due to trauma to the cisterna chyli of any nature constitute the secondary causes. Chylous ascites has both primary and secondary causes. A lymphangioma is a benign proliferation of lymphatic tissue believed to originate from the early sequestration of lymphatic vessels that fail to establish connections with normal draining lymphatics at about 14–20 weeks of intrauterine life<sup>[5]</sup>. Lymphangiomas are therefore considered a congenital rather than an acquired tumor. After birth, they can become markedly dilated as a result of both the collection of fluid and the budding of preexisting spaces<sup>[6]</sup>. The other explanations for the origin of lymphangioma include obstruction of lymph channels secondary to fibrosis, inflammation, trauma, node degeneration; or failure of endothelial secretory function<sup>[7]</sup>. Lymphangiomas at retroperitoneal location are rare. Commonly accepted hypothesis regarding

their origin is the development of abnormal connections between the iliac and retroperitoneal lymphatic sacs, and the venous system, leading to lymphatic fluid stasis in the sacs. In 1877, Wegner histologically divided lymphangiomas into three categories: (1) lymphangioma simplex (capillary lymphangioma) with small, thin-walled lymphatic channels and not commonly found intraabdominally; (2) cavernous lymphangioma with larger thin-walled channels, more common than lymphangioma simplex, but still rare intraabdominally, and may undergo malignant transformation; (3) cystic lymphangioma (always benign) composed of large cystic spaces lined with flat endothelium. Retroperitoneal lymphangiomas are usually of cavernous or cystic types, of which most reported cases have been of a cystic type, as was in our case<sup>[3]</sup>. In our case the patient is 58 years old which is very rare presentation and the patient has history of previous surgeries which suggests the cause to be secondary may be due to trauma caused by the surgery. Slow growing in size and no history of weight loss suggests benign nature of the mass. Due to its large size it presented as abdominal distension. The retroperitoneum is the second most common site in the abdomen next to the mesentery. Lymphangiomas in the retroperitoneum are usually large in size and are insinuating in nature. When they grow to larger sizes, determination of the origin may become difficult. Adrenal lymphangiomas are included in this group and confirming the organ of origin may be possible only per operatively or by histology. Differentials include diaphragmatic mesothelial cyst, retroperitoneal neoplasms, such as cystic schwannoma and other adrenal cysts, cystic mesothelioma, teratoma, undifferentiated sarcomas like liposarcoma and leiomyosarcoma, metastatic lymphadenopathy, cystic metastases (especially from ovarian or gastric primaries), benign tumors such as lymphangioma, adenoma, and other tumors such as retroperitoneal hematoma, abscesses, duplication cysts, ovarian cysts and pancreatic pseudocysts. Preoperative

diagnosis of retroperitoneal lymphangioma is difficult. Ultrasound (US), contrast enhanced CT and MRI scans appear to be complementary to each other in the evaluation of cystic lymphangioma. US demonstrates the internal structure of lymphangiomas, particularly septations with clear fluid. CT may differentiate retroperitoneal and mesenteric lymphangiomas from adjacent bowel loops, and can also distinguish parapelvic renal lymphangiomatosis from hydronephrosis. In our case Ultrasonogram finding shows large globular cystic lesion of size 175mm x 160mm x115mm having well defined margin in the right side of abdominal cavity extending from subhepatic area to the right iliac region. It appears separate from the right kidney, liver and major vessels. Bowel loops appears to be displaced by the lesion. CT scan showed a well defined simple cystic lesion of size 17x15cm noted in the right side of abdominal cavity. It appeared separate from all other visceral organs. Histological diagnosis of lymphangioma is based on well established criteria [8]. These include a well-circumscribed, cystic lesion, with or without endothelial lining, a stroma composed of a meshwork of collagen and fibrous tissue, and a wall containing focal aggregates of lymphoid tissue. Immunohistochemical markers used in the diagnosis of lymphangioma are lymphatic vessel endothelial receptor-1, vascular endothelial growth factor-3, monoclonal antibody D2-40 and prox-1. Surgical excision in totality is the treatment of choice because of its potential to grow and invade surrounding organs [10]. Complete surgical excision is often difficult to achieve because of local invasiveness which may lead to encasement of structures like major vessels and ureter. Incomplete excision often leads to recurrence and redo surgery is quite challenging [11]. Retroperitoneal dissemination during surgery is very rare but potentially fatal [12]. Hauser et al. suggested that isolation and ligation of the cystic lymphangioma's peduncle as well as ligation of lymph channels can prevent recurrences and chylascos [13]. Although marsupialisation

aspiration, drainage, and irradiation of the lymphangioma have been described but they give a poor result and are not recommended [1]. Treatment by argon beam ablation and sclerotherapy has also been reported in a patient with a life threatening total abdominal lymphangiomatosis [14]. In our case the cyst was completely excised without injury to any other viscera and dissemination. The patient recovered well post operatively and till now no evidence of recurrence found.

### Conclusion

Retroperitoneal lymphatic cyst is a rare clinical entity in adults and can be due to trauma to lymphatics due to previous surgery. surgery is required for definitive diagnosis and to ameliorate the symptoms.

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