

## Retroperitoneal Cystic Lymphangioma: A Report of a Rare Clinical Entity

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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. After a thorough literature search in English we concluded that less than 200 cases of adult retroperitoneal lymphangioma have been reported so far. Apart from being a rare entity it also presents as a diagnostic dilemma and final diagnosis is often made on surgical exploration. Case Presentation: We report a case of retroperitoneal lymphangioma in a 65-year-old male who presented with abdominal distension and dull aching abdominal pain. Radiological imaging revealed a large multilobulated cystic abdominal mass involving the upper retroperitoneum and extending into the pelvis. Surgical removal of the cyst was accomplished without incident. A benign cystic retroperitoneal lymphangioma was diagnosed on histology and confirmed with immunohistochemical stains.

**Keywords:** Retroperitoneum, Cystic, Lymphangioma

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### I. Introduction

Lymphangiomas are rare cystic tumors of the lymphatic system. These are benign, slow-growing lesions characterized by proliferating lymphatic vessels[1]. They frequently affect the neck (75%) and the axilla (20%)[2]. Intra-abdominal lymphangiomas (fewer than 5%) have been reported in the mesentery, gastrointestinal tract, spleen, liver and pancreas[3].

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 especially 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.

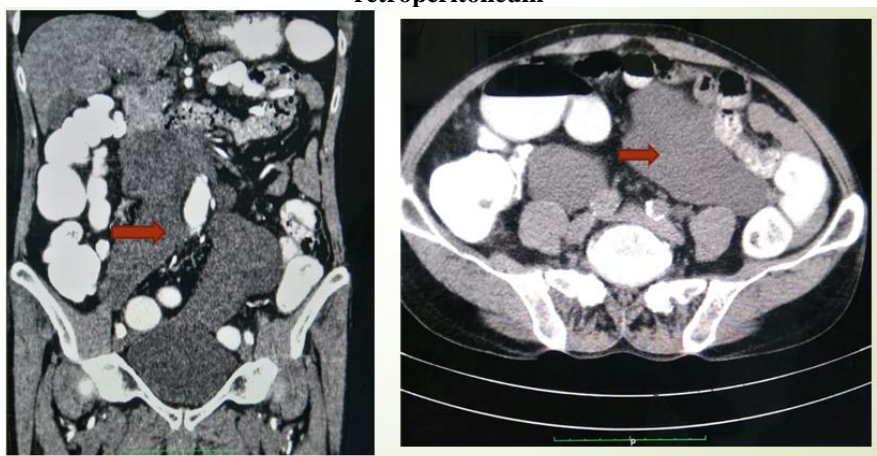
### Observations

A 65 year-old man, who is laundry worker presented to Surgical OPD with complaint of pain in abdomen of 10 days duration. Present complaint started as pain abdomen which is diffuse and constant. There were no other complaints. No significant past, personal or family history. Systemic examination was within normal limits. (Figure -1)



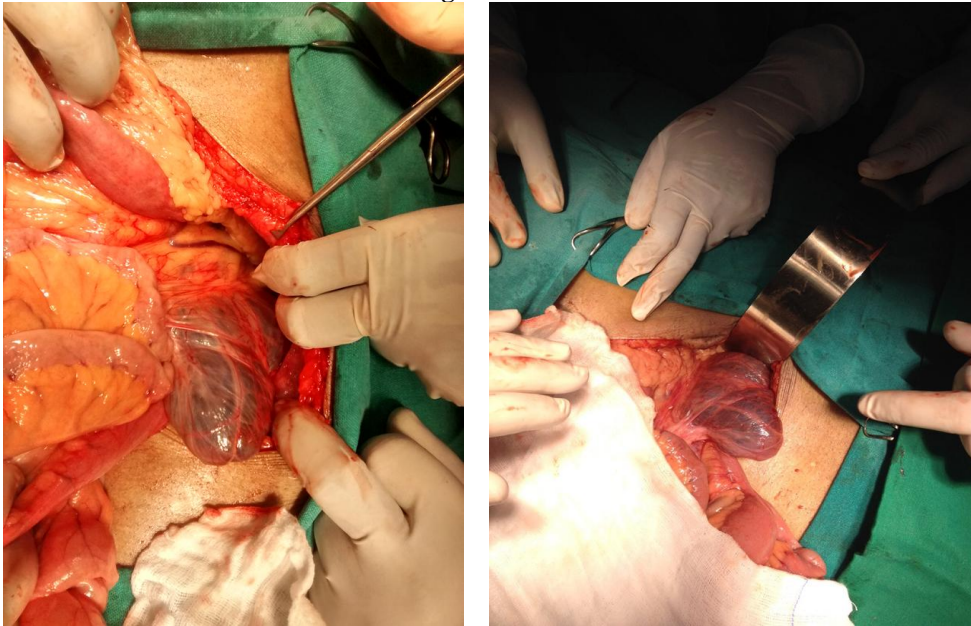
CT scan of abdomen showed a large , lobular , thin walled cystic lesion seen extending from 3rd part of duodenum into pelvis upto urinary bladder inferiorly along the anterior aspect of aorta and IVC with multiple lobulations measuring approximately 17cmx13.6 cm. ( Fig-2 & 3)

**Fig -2 & 3 Abdominal computed tomography scan showing a large multilobular mass in the retroperitoneum**

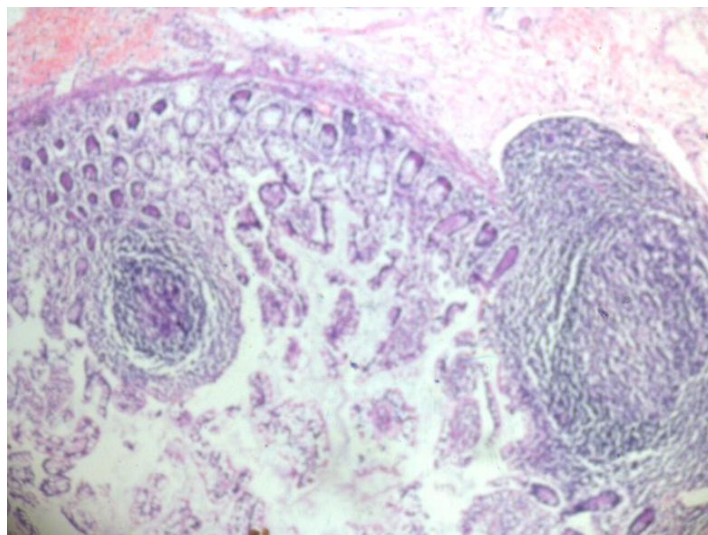
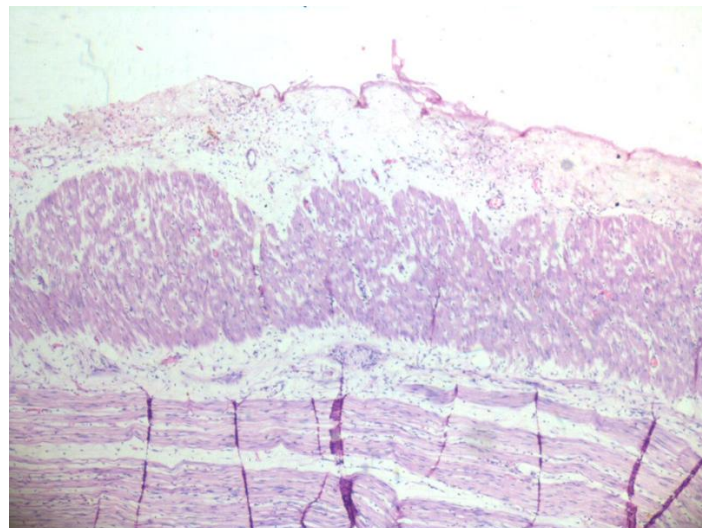


The patient underwent exploratory laparotomy through a midline incision. ( Fig-4) These revealed 2 cystic lesions of about 12x8 cm and 14x8 cm are present lateral to the aorta extending from second part of duodenum upto dome of diaphragm superiorly and inferiorly upto urinary bladder containing clear fluid. Posterior peritoneal wall is opened and both cysts are excised in entirety after separating it from peritoneum as well as retroperitoneum by a combination of blunt and sharp dissection ( Fig-4 &5). About 1 litre of clear fluid is drained from the cyst. Excised cyst is sent for histopathological examination. Fluid from cyst was sent for gram staining and culture and sensitivity that did not show any organism or growth. Histopathology revealed cyst wall which is thrown into papillary folds lined by flattened epithelium and edematous stroma containing blood vessels and focal lymphocytic collection , suggestive of features of cystic lymphangioma.( fig-6 & 7). The patient has been on regular follow up for the last 5 months and there has been no evidence of recurrence , clinically as well as on imaging.

**Figure 4 & 5**



**Fig-6 & 7 : Photomicrograph of cystic lymphangioma**



## II. Discussion:

A lymphangioma is a benign proliferation of lymphatic tissue believed to originate from early sequestration of lymphatic vessels that fail to establish connections with normal draining lymphatics at about 14- 20 weeks of intrauterine life<sup>6</sup>. Lymphangiomas are therefore considered congenital rather than acquired tumor. After birth they become markedly dilated as a result of both the collection of fluid and the budding of pre existing spaces<sup>7</sup>. They frequently affect neck (75%) and the axilla (20%). Lymphangiomas at retroperitoneal collections are rare (<1%). The retroperitoneum is the 2<sup>nd</sup> most common location for abdominal lymphangiomas after mesentery of small bowel. 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 3 categories

- 1) Lymphangioma simplex (Capillary Lymphangioma) : It has small thin walled lymphatic channels and is not commonly found intra abdominally.
- 2) Cavernous lymphangioma : It has larger thin walled channels and is more commoner than capillary lymphangioma, but still are rare intra abdominally and may undergo malignant transformation.
- 3) Cystic lymphangioma : It is always benign, composed of large cystic spaces lined with flat epithelium.

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. Although retroperitoneal lymphangiomas may sometimes be asymptomatic, they usually present as abdominal mass<sup>8</sup>. They become symptomatic if they become large enough to impose on surrounding structures. They might be complicated by intracystic haemorrhage, cyst rupture, volvulus or infection<sup>9</sup>.

Differential diagnosis : cystic mesothelioma, teratoma, undifferentiated sarcomas like liposarcoma and leiomyosarcoma, duplication cysts, pancreatic pseudocysts, cystic metastases from ovarian or gastric primaries, retroperitoneal haematoma.

Pre operative diagnosis of retroperitoneal lymphangioma is difficult. US, CT, MRI appear to be complimentary to each other. Differentiating cystic lymphangiomas from other cystic growths by imaging studies alone is often inconclusive and surgery is most frequently required for definitive diagnosis. The final diagnosis of lymphangioma is achieved by pathological examination of specimen. Surgical excision in totality is the treatment of choice because of its potential to grow and invade surrounding organs<sup>2</sup>. Incomplete excision often leads to recurrence and redo surgery is quite challenging.

Although marsupialisation, aspiration, drainage, irradiation of lymphangioma have been described they give poor result and are not recommended<sup>10</sup>. Treatment by argon beam ablation and sclerotherapy has also been reported in a patient with life threatening total abdominal lymphangiomatosis.

## III. Conclusion

Cystic lymphangioma, a malformation of lymphatic vessels, should be considered among the many different possible diagnosis of retroperitoneal cystic lesion. Retroperitoneal lymphangioma is an uncommon lesion in adults and radiological investigations provide important preoperative diagnostic information for effective surgical approach and management. These rare tumors have excellent prognosis, with symptomatic relief and cure achieved with complete surgical excision.

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