

# Research Journal of Pharmaceutical, Biological and Chemical Sciences

## Retroperitoneal Lymphatic Cyst: Case Report.

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### ABSTRACT

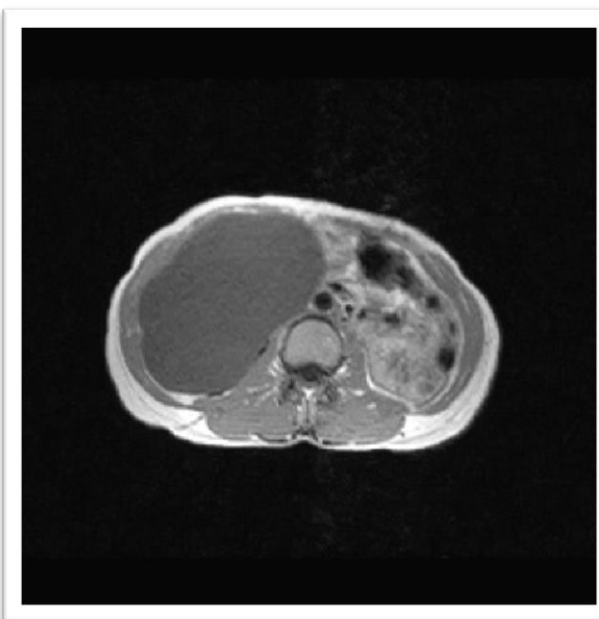
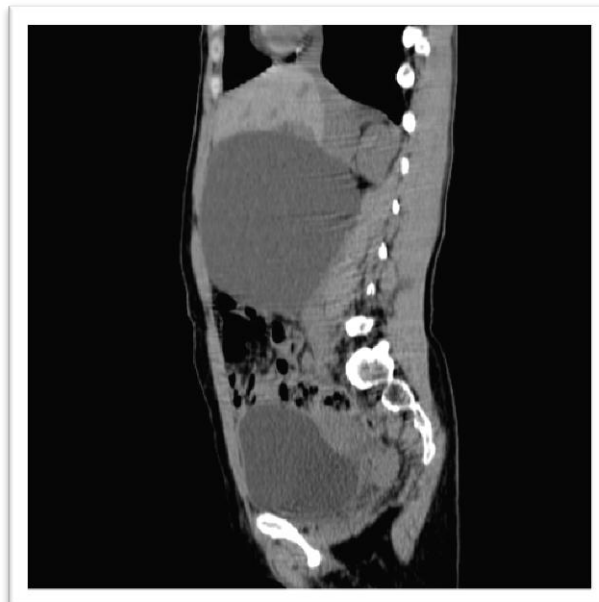
Abdominal lymphangiomas are uncommon benign cystic tumors of the lymphatic system. Retroperitoneal lymphangiomas are even rarer accounting for 1% of all lymphangiomas. It usually manifests in infants. Its quite rare in adults. In our case patient presented with complaints of swelling in the upper abdomen for past 1 month. Excision of the cyst in toto was done and sent for histopathology which showed lymphatic cyst.

**Keywords:** Retroperitoneal, lymphatic cyst.

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**CASE REPORT**

A 29 year old female came with complaints of swelling in the upper abdomen for 1 month in the epigastric region, which was insidious in onset, gradually progressive to attain the present size. No H/o abdominal pain, vomiting, jaundice, loss of weight or loss of appetite. Normal bowel and bladder habits. On examination 10\*12cms swelling extending from right hypochondrium to epigastric region, more prominent in the epigastric region, surface was smooth, firm in consistency, swelling becomes less prominent on head rising, doesn't move with respiration and was dull on percussion. Routine investigation was done. Ultrasound abdomen was done which showed well defined cystic lesion in porta displacing the gall bladder and head of pancreas and part of CBD. MRCP was done which showed a retroperitoneal cyst , in view of extension of small portion of the cyst between the abdominal aorta and anterior aspect of L2 vertebra. Laparotomy was planned, cystic swelling arising from the right hypochondrium, the content was chylous(chalky white). Cyst wall was present retroperitoneal region arising between IVC and Abdominal aorta. The cyst wall was excised and sent for histopathology which showed features of lymphatic cyst. Drainage tube was placed till post-operative day 12 and there was mild chylous fluid collection was present in drainage tube which gradually decreased and tube was removed on post-operative day 12. Patient was discharged on post-operative day 15.





### DISCUSSION

A cystic tumor in the retroperitoneum accounts for widely differing diagnoses, including malignant tumors such as cystic mesothelioma, teratoma, undifferentiated sarcoma, cystic metastases (especially from ovarian or gastric primaries), malignant mesenchymoma, benign tumors such as lymphangioma, cysts of urothelial and foregut origin, microcystic pancreatic adenoma, and other tumors such as retroperitoneal hematoma, abscesses, duplication cysts, ovarian cysts and pancreatic pseudocyst

Cystic lymphangiomas are rare benign malformations of the lymphatic system. Generally, they are common in children and more frequent in male. Approximately 50% of lymphangiomas are present at birth, and almost 90% are diagnosed before the age of 2 years. The most frequently affected sites are the neck (75%) commonly known as 'cystic hygroma' and axilla (20%)[2]. Approximately 5% of the lymphangiomas are intra-abdominal arising from the mesentery or retroperitoneum (nearly 1%)[4]. However they can occur in any location where lymphatics are normally encountered. The frequent development of lymphangiomas within areas of primitive lymph sacs suggests these to be malformations arising from sequestrations of lymphatic tissue that fail to communicate normally with the lymphatic system[4], or developmental defects between the 6<sup>th</sup>-9<sup>th</sup> wk of embryonic development resulting from abnormal budding of the lymphatic endothelium [1]. These abnormal lymphatic channels would dilate, frequently resulting in the formation of uni- or multi-cystic masses that might subsequently be inflamed or obstructed, leading to additional lymphangiomas. The cystic masses are filled with chylous or serous material and lined with a layer of endothelium. It is speculated that lymphangiomas develop due to sequestrations of lymphatic tissue that fail to communicate normally with the lymphatic system. Other potential causes are thought to include abdominal trauma, localized lymphatic degeneration, radiation and lymphatic obstruction. Most retroperitoneal lymphangiomas remain asymptomatic and therefore grow to large proportions. A contrast CT can demonstrate 'water density' fluid within unilocular or multilocular cysts while MRI may delineate additional lesions.

Intra-abdominal lymphangiomas are of four types. Type I, the pedicled type, enlarge rapidly and cause torsion. Type II, the sessile type, is located within the mesentery of the bowel. Type III, the retroperitoneal type, involves the retroperitoneal structures, such as the mesenteric root, the aorta and the vena cava. The type IV multicentric type extensively involves the intra-abdominal and retroperitoneal organs [5].

The treatment of choice is complete surgical resection in order to avoid recurrences or complications. Aspiration and injection of sclerosing agents may be used for symptomatic diffuse lesions which are unresectable due to involvement of vital structures. Histopathology remains the key to definitive diagnosis, sometimes supported by immunohistochemistry. Characteristic finding is dilated lymph vessels lined by endothelial cells along with connective tissue and smooth muscle fibres forming the intervening stroma. The

presence of smooth muscle and lymphatic spaces in the wall of the cyst differentiates it from a chylolymphatic cyst. Histologically lymphangiomas are classified into three types; capillary (or simple), cavernous and cystic. Simple lymphangiomas consist of small, thin-walled, lymphatic channels. Cavernous types are composed of dilated lymphatic channels. Cystic type contains lymphatic spaces with smooth muscle fibres in the stroma but no connection with the normal adjacent lymphatics. Intra-abdominal and retroperitoneal lymphangiomas are generally of the cystic type.

### **CONCLUSION**

Retroperitoneal lymphangiomas are exceedingly rare benign neoplasms which may present as a diagnostic dilemma. Definitive preoperative diagnosis may not be possible. However optimal surgical management can result in remarkably good prognosis.

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