

# Huge Retroperitoneal Lymphangioma Presenting with Duodenal Obstruction: A Case Report

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## ABSTRACT

Lymphangiomas are benign tumors that develop due to congenital proliferation of lymphatic tissue causing an obstruction of the lymphatic ducts. Lymphangiomas may present with a variety of symptoms from mild abdominal discomfort to abdominal obstruction and DIC. A 22-year-old female presented to us with intermittent abdominal pain and distention. On examination a large mobile mildly tender swelling extending towards the right side of the abdomen was palpated. CT scan revealed signs of partial duodenal obstruction. On Ultrasonography, a large cystic mass with septations was observed with extension in the right side of the abdomen. Laparoscopic assisted removal of the mass was performed. The patient recovered uneventfully and was discharged home with a healthy six-month follow-up with no signs of recurrence.

**Keywords:** Cystic Retroperitoneal Lymphangioma.

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## Introduction

Lymphangiomas are benign tumours caused by the congenital proliferation of lymphatic tissue, causing an obstruction in the lymphatic ducts, producing fluid filled cysts.<sup>1</sup> Histologically they are polycystic, with cysts divided by thin septae lined with endothelial cells. Most common presentation of cystic lymphangiomas are in the neck (75%) and axillary region (20%) and only 5% are intra-abdominal, while retroperitoneal lymphangiomas account for only 1% of cases.<sup>2</sup> It is more common in children with males affected more than women, while women have shown to encounter pancreatic lymphangiomas more than men.<sup>3</sup> Often retroperitoneal lymphangiomas are asymptomatic but due to their large size they might cause a wide variety of symptoms from mild abdominal distention/discomfort and back pain to duodenal obstruction, sepsis and DIC.<sup>4</sup> Diagnostic techniques are ultrasound, CT scan and MRI but often it is found accidentally and definitive diagnosis is made post-operatively. Treatment of choice is complete surgical resection with cystectomy.

## Case Report

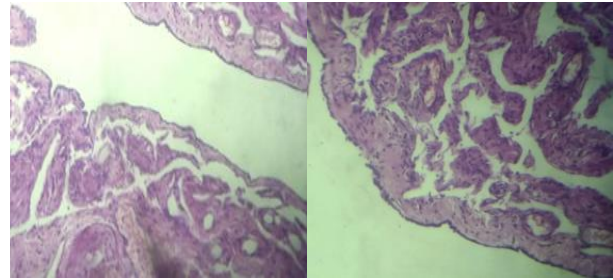
A 22 year old female presented with on and off vomiting, abdominal distension and pain for a period of two years. There were no co-morbidities or significant family history. During abdominal examination a large, palpable, mildly tender intra-abdominal mass, occupying almost all of the abdomen. Her blood tests showed no anaemia or inflammation and her amylase, lipase, bilirubin and tumour markers were within normal limits. Initially reporting to the gastroenterology department, she underwent oesophageal-gastro-duodenoscopy which showed duodenal obstruction due to an extrinsic compression. CT scan was done which showed a large cystic mass occupying almost all of the abdomen, more on the right side. Ultrasound showed a very massive multiloculated cystic mass mostly on the right side extending above the superolateral surface of liver superiorly covering the right lobe and into pelvis inferiorly. Displacement of the bowel and kidneys can also be seen. (Figure 1)

Laparoscopic assisted excision of cyst was performed and cyst was removed through right upper transverse incision. It appeared to have a small connection with pancreas which was carefully dissected and sutured. Gall bladder was closely adherent to cyst wall therefore

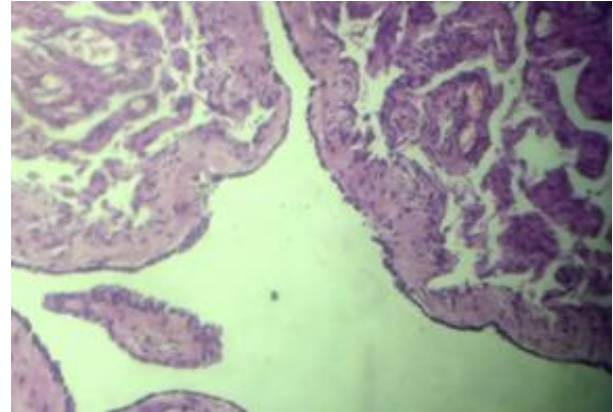
cholecystectomy was also done. A drain was placed in the retro pancreatic area. Patient made an uneventful recovery and was discharged on 6<sup>th</sup> postoperative day. At six months follow up patient is fully recovered and healthy and no signs of recurrence.



**Figure 1. CT scan showing large cystic Lymphangioma occupying almost all of the abdomen and covering the right lobe of the liver.**



**Endothelium lined walls**



**Lymphoid aggregates**

**Figure 2. Histopathology showing classical picture of Lymphangioma**

Large lymphatic channels in loose connective tissue, focally disorganized smooth muscle. Increased mast cells, classically present in lymphangiomas.

## Discussion

Retroperitoneal lymphangiomas are benign congenital neoplasms. Three histological types of lymphangiomas are present: cystic, capillary, and cavernous but cystic is the most common.<sup>5,6</sup> On ultrasonography a large anechoic lesion can be identified often with septation and containing some echoic calcified debris. CT scan is used to determine the size and location of the cystic mass. contrast enhanced CT may show enhancement of the walls and septa.<sup>5</sup> Often these cysts are septated, lined with endothelial cells and filled with serous, serosanguinous or chylous fluid. Lymphangiomas commonly present in the paediatric population occurring mostly in the head and neck region.<sup>7</sup> In adults it is extremely rare, presenting commonly in the abdomen where it arises from the mesentery, parts of colon and occasionally the retroperitoneum. Women have shown to have an increased incidence to retroperitoneal lymphangioma compared to men. Though mostly congenital it is now believed that adult lymphangiomas may also be cause due to trauma, fibrosis, radiation therapy and neoplasms.<sup>8</sup>

Most retroperitoneal lymphangiomas are asymptomatic and picked up incidentally on imaging. A variety of symptoms may be seen in patients depending on the location and size of the lesion such as abdominal distention and discomfort, abdominal pain, abdominal asymmetry to bowel obstruction, infection, cyst rupture, sepsis, DIC and haemorrhage within the cyst.<sup>9</sup>

The diagnosis of lymphangioma is made postoperatively on histology, However modalities such ultrasound and CT scan are highly sensitive in identifying intra-abdominal masses.<sup>10</sup> It is difficult to differentiate between pancreatic cystic lesion and retroperitoneal lymphangioma but it can be used to differentiate from liver and kidney pathologies.

Treatment of choice is complete resection via laparotomy or laparoscopy.<sup>11</sup> In complicated cases resection of adherent bowel or gallbladder may also need to be done. Aspiration of cyst contents may also be done but there is high risk of recurrence. The cyst should be sent to pathology for diagnosis.

## Conclusion

Retroperitoneal lymphangiomas are rare tumors often asymptomatic and diagnosed incidentally. CT/ultrasonography are modalities of choice when diagnosing this lesion but definitive diagnosis is made histologically after resection. Complete resection via laparotomy or laparoscopy is the treatment of choice.

## References

1. G. Nuzzo G, Lemmo MM, Marrocco-Trischitta G, Boldrini, and I Giovannini, Retroperitoneal cystic lymphangioma, *Journal of Surgical Oncology* 1996; 61(3): 234–237. [https://doi.org/10.1002/\(SICI\)1096-9098\(199603\)61:3<234::AID-JSO14>3.0.CO;2-7](https://doi.org/10.1002/(SICI)1096-9098(199603)61:3<234::AID-JSO14>3.0.CO;2-7)
2. M. Tripathi, S. Parshad, R. K. Karwasra, A. Gupta, S. Srivastva, and A. Sarwal, "Retroperitoneal lymphangioma in an adult: a case report of a rare clinical entity," *Case Reports in Surgery*. 2015, Article ID 732531, 4 pages, 2015. <https://doi.org/10.1155/2015/732531>
3. Igarashi A, Maruo Y, Ito T, Ohsawa K, Serizawa A, Yabe M, Seki K, Konno H, Nakamura S. Huge cystic lymphangioma of the pancreas: report of a case. *Surgery Today*. 2001 Aug;31(8):743-6. <https://doi.org/10.1007/s005950170084>
3. Gachabayov M, Kubachev K, Abdullaev E, Babyshin V, Neronov D, Abdullaev A. A huge cystic retroperitoneal lymphangioma presenting with back pain. *Case reports in medicine*. 2016 <https://doi.org/10.1155/2016/1618393>
4. Schneider G, Seidel R, Altmeyer K, Remberger K, Pistorius G, Kramann B, Uder M. Lymphangioma of the pancreas and the duodenal wall: MR imaging findings. *European radiology*. 2001;11(11):2232-5. <https://doi.org/10.1007/s003300100846>
5. Wilson SR, Bohrer S, Losada R, Price AP. Retroperitoneal lymphangioma: an unusual location and presentation. *J Pediatr Surg*. 2006;41(3):603-5. <https://doi.org/10.1016/j.jpedsurg.2005.11.057>
7. Kullendorff CM, Malmgren N. Cystic abdominal lymphangioma in children. *Case report, Eur J surgery*, 1993;159:499-501.
8. Enzinger F. M, Weis S. W. *Tumors of lymph vessels, in Soft Tissue Tumors*. St. Louis, Mo, USA: Mosby-Years Book; 1995.679–700.
9. Gayen R, Mahata M, Dasgupta S, Dasgupta J. Giant retroperitoneal cystic lymphangioma- a case report with review of literature. *Journal of Dental and Medical Sciences*. 2015;14(2):69–71.
10. Davidson AJ, Hartman DS. Lymphangioma of the retroperitoneum. CT and sonographic characteristics. *Radiology*. 1990;175:507–10. <https://doi.org/10.1148/radiology.175.2.2183287>
11. Targarona EM, Moral A, Sabater L, Martinez J, Luque P, Trias M. Laparoscopic resection of a retroperitoneal cystic lymphangioma. *Surg Endosc*. 1994;8:1425–6. <https://doi.org/10.1007/BF00187350>