

Case Report

A rare case of a retroperitoneal cystic lymphangioma

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ABSTRACT

Retroperitoneal Lymphangiomas are rare and account for only 1% of lymphangiomas. They usually present in infancy, rarely they may present symptomatically in adulthood. We present a case of a 19-year old female with a symptomatic retroperitoneal lymphangioma. It was treated with complete surgical excision. Retroperitoneal lymphangiomas are rare. Imaging alone cannot differentiate them from other retroperitoneal cystic masses. Surgical excision is the treatment of choice and required for final diagnosis.

Keywords: Lymphangioma, Retroperitoneal cyst, Surgical excision

INTRODUCTION

Lymphangiomas are fluid filled cysts produced due to blockage of the lymphatic system. Retroperitoneal lymphangiomas are rare and account for 1% of all lymphangiomas. They mostly present in infancy, less than 200 cases of adult retroperitoneal lymphangiomas have been reported in literature. Other than being rare they cause a diagnostic dilemma with other retroperitoneal cystic masses.

Authors reported a case of a 19-year-old female who presented with abdominal distension, pain and vomiting.

CASE REPORT

A 19-year-old female of Indian ethnicity presented to the general surgery OPD with complains of abdominal distension for 2 months, continuous dull aching abdominal pain since 2 months and vomiting since 1 month. Patient gave no history of trauma, weight loss, altered bowel habits, fever or any urinary symptoms.

A vague mass approximately 15x12 cm was palpable extending from the right hypocondrium, lumbar up to the umbilical region just crossing the midline. It had a smooth surface and ill-defined margins. The lump was firm, non-tender with restricted mobility in all directions. Ultrasonography showed a large cystic lesion with septations in the retroperitoneal region located in the para aortic, pre-caval space.

A CECT of the abdomen (Figure 1) showed a large 12x5.9x11cms cyst extending from the level of the right renal artery upto the supravescical region, compressing the abdominal IVC, right ureter and kidney, displacing the small bowel loops and sigmoid. There was no evidence of invasion of surrounding structures.

With a provisional diagnosis of a retroperitoneal lymphangioma/ mesenteric cyst the patient underwent a Exploratory laprotomy. Intraoperatively a large dumbbell shaped cyst was found arising from the retroperitoneum. It was easily separated from the surrounding structures. The patient tolerated the procedure well and post-

operative course was uneventful. Histopathology confirmed the diagnosis of a cystic lymphangioma.

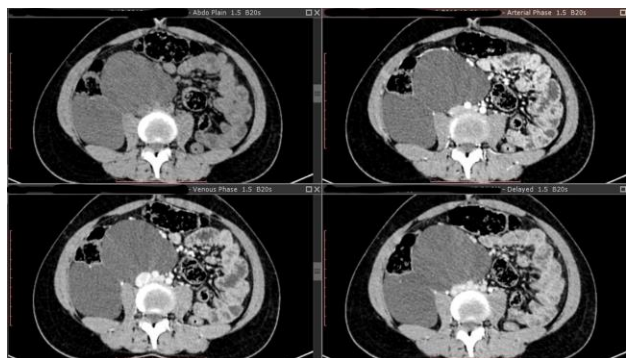


Figure 1: CECT abdomen.

DISCUSSION

Lymphangiomas are believed to originate from early sequestration of lymphatic vessels which fail to establish a connection with the normally draining lymphatic system at 14-20 weeks of life.¹ They are most commonly found in the head and neck (75%), Axilla (20%).² The remainder 5% are intrabdominal arising from the mesentery of the small bowel, retroperitoneum of greater omentum.^{3,4}

The retroperitoneum is the second most common intraabdominal site after the mesentery. In 1877, Wegner histologically divided lymphangiomas into three categories:

- Lymphangioma simplex (capillary lymphangioma) with small, thin-walled lymphatic channels and not commonly found intraabdominally.
- Cavernous lymphangioma with larger thin-walled channels, more common than lymphangioma simplex, but still rare intra abdominally, and may undergo malignant transformation, and
- 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.⁵

A cystic tumour arising from the retroperitoneum is a diagnostic dilemma with a differential diagnosis of teratomas, sarcomas, cystic metastasis, duplication cysts, lymphangiomas, hamartomas and abscesses. It is tough to establish a pre-operative diagnosis based on only USG,

CT, MRI findings only. The diagnosis is confirmed only after laprotomy with histopathological examination.

Although marsupialisation, aspiration, drainage and irradiation have been described, results are poor with high rates of recurrence.⁶ Complete surgical excision is the treatment of choice. Incomplete excision may lead to recurrence and redo surgeries are quite challenging.⁷

CONCLUSION

Retroperitoneal lymphangiomas are a rare clinical entity. Imaging alone is unable to differentiate it from other cystic lesions; thus, surgical excision is required for conformation of diagnosis. They can be easily treated with complete surgical excision.

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