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Case Report

Retroperitoneal lymphangioma in a tertiary care hospital of North India

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ABSTRACT

Less than 200 cases of retroperitoneal lymphangiomas (RL), a rare type of benign cystic tumour of the lymphatic system that makes about 1% of all lymphangiomas, have been documented to date. Their rarity makes preoperative diagnosis challenging. 95% of lymphangiomas are found in the head, neck, and axilla where they are most frequently found. We are presenting a 30 years old woman, who presented to outpatient department (OPD) of general surgery with symptoms of recurrent left lumbar pain, radiating towards inguinal region for one month. The contrast computed tomography (CT) scan of abdomen and pelvis defined a large thin walled homogenous non enhancing hypodense cystic lesion in left side of abdomen in retroperitoneal location from left lobe of liver till pelvis on left side craniocaudally. The patient was planned for exploratory laparotomy. Midline laparotomy was performed. A cystic lymph-filled tumour was detected in the abdominal cavity adjacent to the bowel loops. Histologically the mass contained variable-sized cystic spaces lined by flattened endothelium consistent with lymphatic vessels, which on immunostaining were positive for CD31 and negative for Pan CK. The patient was discharged satisfactorily. In conclusion, retroperitoneal lymphangioma presents vaguely as abdominal pain or mass and grown to large sizes without causing any significant symptom. Tumour markers for the excised tissue are important for diagnosis. Laparotomy or laparoscopy are the two surgical excision methods used to treat retroperitoneal cystic lymphangioma and it responds well to excision.

Keywords: Retroperitoneal lymphangioma, CD31, Pan CK, Lymphatic cystic tumour

INTRODUCTION

Less than 200 cases of retroperitoneal lymphangiomas (RL), a rare type of benign cystic tumour of the lymphatic system that makes about 1% of all lymphangiomas, have been documented to date. Their rarity makes preoperative diagnosis challenging.[1] 95% of lymphangiomas are found in the head, neck, and axilla where they are most frequently found. The kidney, bone, adrenal glands, lungs, and mediastinum contain the remaining 5%. The mesentery, gastrointestinal tract, retroperitoneum, spleen, liver, and pancreas are less frequently encountered locations.[2]

The most common clinical symptoms of retroperitoneal cystic lymphangioma are back or abdominal pain, fever, exhaustion, weight loss, and haematuria, albeit they can go for a long time without showing any symptoms.[2] Tumour masses with cavernous, cystic, or uniseptal capillaries may resemble them.[3]

CASE REPORT

We are presenting a 30 years old woman, who presented to OPD of general surgery with symptoms of recurrent left lumbar pain, radiating towards inguinal region for one month. The pain was insidious in onset, progressive in

nature, dull in character with 8/10 on visual analogue scale, relieving on medication. The patient also complained of nausea and vomiting post prandially and generalised heaviness in the left quadrant. On physical examination abdomen was soft and non-tender. No abdominal mass was palpable and the bowel sounds were diminished in all quadrants. All laboratory results were in normal ranges. The contrast computed tomography (CT) scan of abdomen and pelvis defined a large thin walled homogenous non enhancing hypodense cystic lesion in left side of abdomen in retroperitoneal location from left lobe of liver till pelvis on left side craniocaudally (71×106×182 mm, AP×TR×CC). No evidence of calcification/post contrast enhancement seen. No soft tissue content noted within it. Superiorly, lesion was abutting left lobe of liver. Inferiorly, it was extending till the pelvis (Figure 1). Medially, it was abutting the fundus of stomach, pancreas, small bowel and pushing the bowel towards right with maintained fat planes. Laterally, the lesion was abutting spleen with maintained fat plane and posteriorly abutting the left kidney. The attenuation coefficient of the mass was between +10 and 15 HU (Figure 2). The preliminary diagnosis was retroperitoneal lymphangioma.

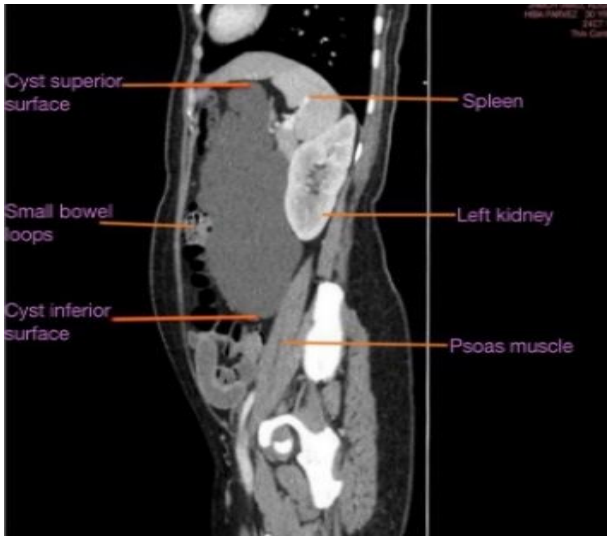

Figure 1: Extending till the pelvis.

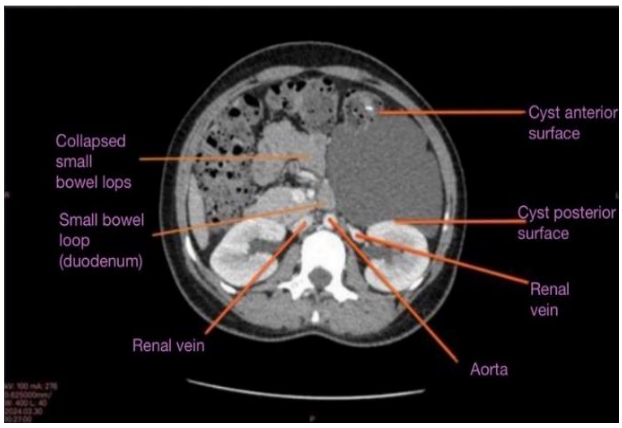

Figure 2: Attenuation coefficient of the mass.

The patient was planned for exploratory laparotomy. Midline laparotomy was performed. A cystic lymph-filled tumor was detected in the abdominal cavity adjacent to the bowel loops. Removal of cyst was done using careful blunt and sharp dissection. The tumor was dissected from left lobe of liver, small bowel, left Gerota's fascia, spleen and gastric fundus and was removed in its entirety (Figure 3). Punctured cyst revealed clear fluid about 800 cc in volume (Figure 4). No extravasation of the lymph flow was observed after removal of the cystic lesion. Abdomen was closed after achieving hemostasis and patient kept nil per oral for 2 days. The patient was discharged post operatively on day 4 in satisfactory condition.

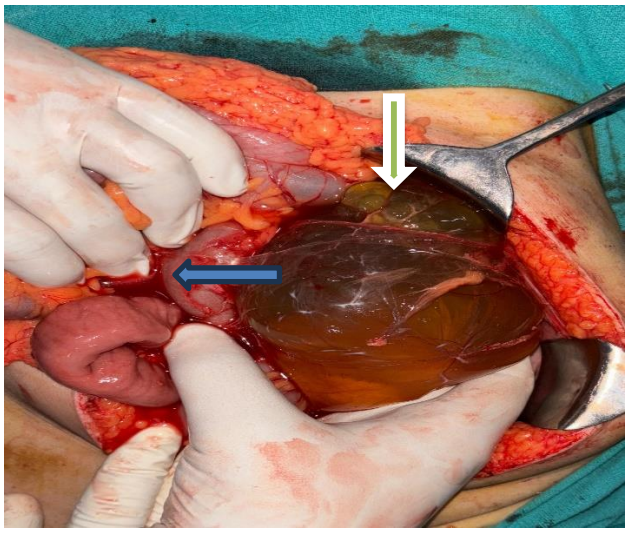

Figure 3: Cystic lesion filled with homogenously clear fluid in the peritoneal cavity extending from left lobe of liver upto pelvis (grey arrow), adjacent small bowel loops with mesentery (blue arrow).

Figure 4: Punctured cyst taken out after laparotomy with blunt and sharp dissection with clear (20×6 cm fluid filled cystic lesion).

Histologically the mass contained variable-sized cystic spaces lined by flattened endothelium consistent with lymphatic vessels, which on immunostaining were

positive for CD31 and negative for Pan CK (Figure 5 and 6). Therefore, the diagnosis of a retroperitoneal cystic lymphangioma was histologically confirmed.

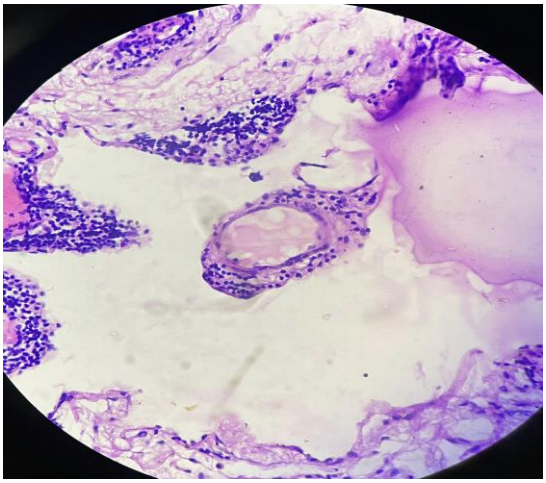

Figure 5: Hematoxylin and eosin stained section from cyst wall shows numerous small to medium sized thin walled dilated vascular spaces lined by benign looking endothelial cells. At places these vessels are surrounded by lymphoid aggregates.

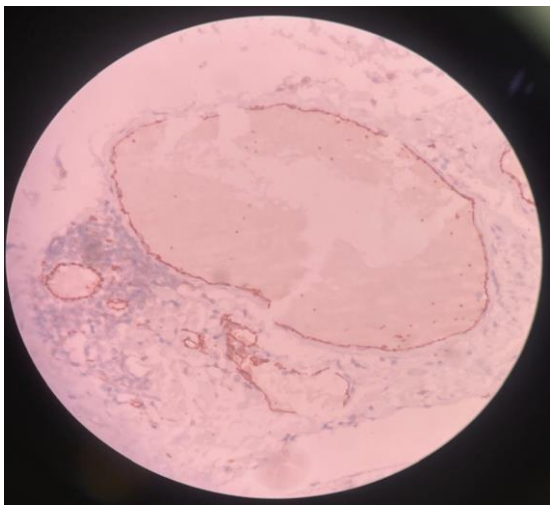

Figure 6: Positivity in the CD 31 marker applied to endothelial cells of dilated vessels.

DISCUSSION

Koch published the first description of lymphangioma in 1913. According to one idea, lymphangiomas originate from a congenital abnormality of lymphatic vessels, which causes lymphatic flow obstruction and lymphangiectasia.[4] There are three different histological forms of lymphangiomas: capillary, cavernous, and cystic. Cystic types of retroperitoneal lymphangiomas, like the one we see here, are the most prevalent.[5,6]

Retroperitoneal lymphangiomas can present with a variety of clinical signs and symptoms, and making the diagnosis is frequently difficult. They are initially asymptomatic, with the first signs being mild abdominal pain, abdominal distention, and abdominal asymmetry brought on by the growing mass.[7] There have also been reports of uncommon clinical symptoms such anemia and back pain. In addition to previous hematuria symptoms, our patient had complaints of recurring left lumbar discomfort.[8]

Laparotomy or laparoscopy are the two surgical excision methods used to treat retroperitoneal cystic lymphangioma.[9] Another therapeutic approach that has been shown to be successful is nonoperative measures such sclerosant agent injections and aspiration of contents.[10] The preferred course of treatment is complete surgical excision in order to prevent bleeding, infection, rupture, progressive growth, and recurrence.[4,11,12] In our situation, too, surgical excision was performed.

CONCLUSION

Lymphangiomas are rarely develop in the retroperitoneum and they are usually asymptomatic. When the retroperitoneal lymphangiomas reach large sizes, symptoms like back pain and hematuria can occur and the method of treatment is total surgical excision.

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