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ORIGINAL ARTICLE

"Lymphangioma Common Entity at Uncommon Site" – A Case Series

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Abstract

Background: Most lymphangiomas represent malformations rather than true neoplasms and are thought to result from failure of the lymphatic system to communicate with venous system. Three forms exist mainly: capillary, cavernous and cystic. **Materials And Methods:** The present study was undertaken in the Department of Pathology, P.D.U. Government Medical College, Rajkot, Gujarat, India. All the three specimens were fixed in 10% formalin overnight, processed, blocks were made and sectioning was done and stained with Harris Haematoxylin and Eosin stain. **Result:** This study describes the rare case of a cystic retroperitoneal lymphangioma in a 29 and 20-year-old male patient with clinical symptoms of abdominal pain and abdominal distension respectively. Whereas other 12 year old male patient had nausea, vomiting and no passage of stool suggesting intestinal obstruction. Radiological imaging revealed a multiloculated cystic mass with enhancing septations suggesting neoplasm - lymphangioma. Surgical removal was done and sent for histopathological examination. All three cases were then diagnosed as lymphangiomas histopathologically. **Conclusion:** Lymphangioma in adults is a rare, benign proliferation of lymphatic tissue. Surgery is often required to reduce the symptoms, and histopathological analysis is necessary to confirm the diagnosis.

Keyword: Lymphangioma, Benign, Neoplasm.

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Introduction

Lymphangiomas are uncommon, benign malformations of lymphatic system that can occur anywhere on the skin and mucous membranes. Lymphangiomas can be categorized as deep or superficial based on the depth and size of the abnormal lymphatic vessels, or as congenital or acquired.[1]

The deep forms of lymphangioma include two specific well defined congenital entities: cavernous lymphangioms and cystic hygromas.[2] Superficial forms of lymphangioma include

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lymphangioma circumscriptum and acquired lymphangioma, which is also referred to in the literature as lymphangiectasia.
Although both entities share similar clinical and histologic features, the term lymphangioma circumscriptum infers lymphatic channel dilatation due to a congenital malformation of the lymphatic system. Whereas, the term lymphagiectasia, or acquired lymphangioma, denotes dilated lymphatic channels of previously normal lymphatics that have become obstructed by an external cause.
Some lymphangiomas are diffuse and/or multicentric, the condition can be designated as lymphangiomatosis.
Lymphangiomas are rare entities that account for less than 1% of the cases in retroperitoneum.[3]

Materials And Methods
The following cases were diagnosed in Histopathology laboratory, Department of Pathology, P.D.U. Government Medical College and Hospital, Rajkot, Western India.
Specimen were fixed in 10% formalin; following fixation of 12 hours, sections were passed through the steps of dehydration, clearing and impregnation and embedding in paraffin, finally block preparation, cutting done and sections were stained with Harris Haematoxylin and Eosin stain and made ready for microscopic examination.

Results
Case 1
A 29 year old male patient came to Surgical Outdoor Patient Department (OPD) with complain of abdominal pain since 1 day. On deep palpation, lump was faintly palpable.
Ultrasonography of abdomen: Pancreas sized 12 x 5.5 cm with ill-defined multicystic lesion with multiple thick septas and echoes within with evidence of vascularity within septas noted at splenorenal pouch suggestive of possibility of neoplastic etiology – mucinous cystadenoma likely arising from tail of pancreas.
MSCT scan of abdomen with pelvis: An irregular thin walled cystic non-enhancing lesion of size 116 (CC) x 99 (AP) x 88 (TR) mm is noted near splenic hilum. Thin septa noted within the lesion. The lesion is extending near upper pole of left kidney and abutting left supra renal gland. The lesion is extending on inferior aspect of tail of pancreas. The lesion is extending on lateral aspect and posterior aspect of splenic flexure of colon. The lesion is extending on posterior aspect of descending colon up to level of L5 vertebrae. Overall findings suggest possibility of Lymphangioma likely.
Specimens received: One specimen of resected part of lesion with part of transverse colon and proximal descending colon and spleen, and other specimen of resected part of lesion with transverse mesocolon was received at histopathology laboratory, following surgery.

On Gross Examination (Figure 1):
Specimen 1: Tissue received size measuring 22 x 14.5 x 5 cm having multiple cystic structures with largest cyst measuring 6 x 4 x 2.5 cm.
On cut section, clear fluid came out.
Specimen 2: Tissue received size measuring 2.5 x 2 x 1 cm with multiple cysts.
On cut section, clear fluid came out.

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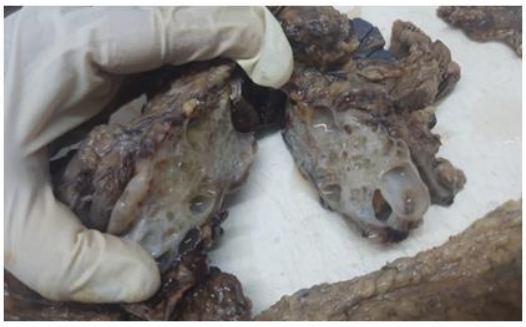

Figure 1: shows multiple cystic structures attached to splenic hilum

Microscopic Examination (Figure 2): Studied sections show large lymphatic channels growing in loose connective tissue. There is large collection of lymphocytes in the stroma. Overall findings are suggestive of **LYMPHANGIOMA**.

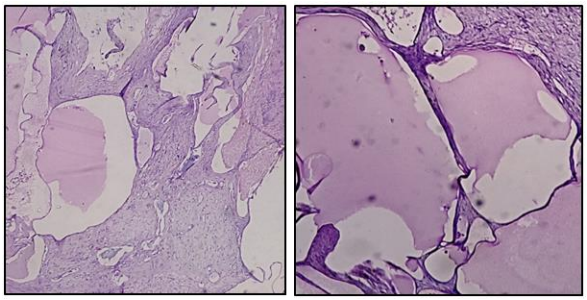

Figure 2

Case 2

A 20 year old male patient came to Surgical Outdoor Patient Department (OPD) with complain of painless lump in abdomen since 2-3 months.

MSCT scan of abdomen with pelvis: A multicystic lesion of size of 106 (CC) x 115 (TR) x 74 (AP) noted in retroperitoneal region of left side on posteroinferior aspect of pancreas abutting/involving body and tail of pancreas.

Possibility of multicystic pancreatic/retroperitoneal neoplasm appears likely.

Excised retroperitoneal mass was received at histopathology laboratory following surgery.

On Gross Examination (Figure 3): Tissue size measuring 11 x 10 x 4 cm.

On cut section, purulent fluid came out, multiple cystic structure found, largest size measuring 3 x 2 x 1 cm.

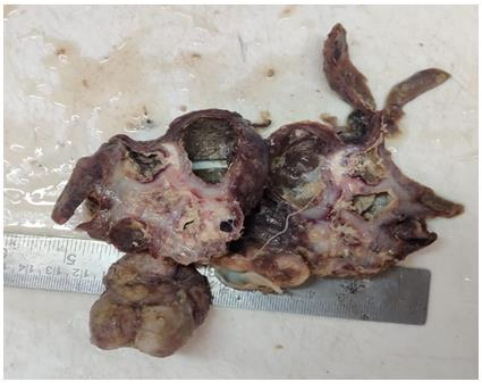

Figure 3: shows multiple cystic structures of retroperitoneal mass

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Microscopic Examination (Figure 4): Studied sections show large, irregular vascular spaces lined by flattened, bland epithelial cells with fibroblastic and collagenous stroma. These cystic spaces are containing clear lymphatic fluid at many places with clustering of lymphocytes seen. Stroma contains large areas of congestion, haemorrhage, necrosis and chronic inflammatory infiltrate.
Overall findings are suggestive of **CAVERNOUS LYMPHANGIOMA WITH SECONDARY CHANGES**.

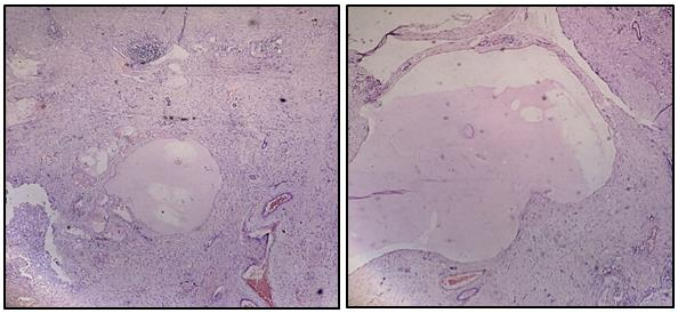

Figure 4

Case 3
A 12 year old male patient came to Surgical Outdoor Patient Department (OPD) with complain of vomiting since 4-5 days, abdominal distension since 4 days and no passage of stool since 2-3 days.
X ray abdomen (standing): Few abnormal air fluid levels are seen in the central abdomen.
USG abdomen and pelvis: Torsion of bowel noted around it's mesentery in the umbilical fossa suggesting intestinal volvulus. Whirlpool sign noted. Multiple mesenteric cysts noted in the pelvis, largest cyst size measuring 50 mm. Overall findings suggest possibility of intestinal obstruction likely.
MDCT scan of whole abdomen: Lobulated hypodense collection seen in pelvis distal to twisting of mesentery measures 98 x 65 x 57 mm in size – mesenteric lymphangioma formation likely.
Resected part of ileum with mesentery was sent to histopathology laboratory following surgery.

On Gross Examination (Figure 5): 10 cm long part of ileum received with attached mesentery with multiple cystic structures, largest cyst size measuring 6.5 x 6 x 3 cm.
On cut section, yellowish white fluid came out, multiple smaller greyish white cystic structures seen.

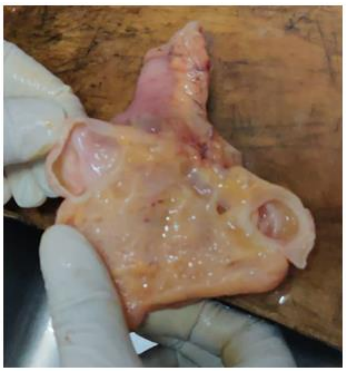

Figure 5: Yellowish white with multiple smaller greyish white cystic structures

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Microscopic Examination (Figure 6): Studied sections show large lymphatic channels in loose connective tissue stroma and peripheral lymphoid aggregate seen.
Overall findings are suggestive of **LYMPHANGIOMA.**

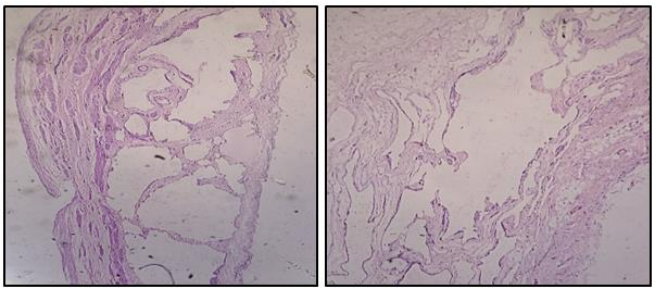

Figure 6

Discussion

Lymphangioma is a rare, benign, congenital malformation of unknown etiology that originates from lymph vessels and this entity was first described by Virchow in 1854.[4]
Approximately 50% of lymphangiomas are present at birth, and almost 90% are diagnosed before the age of 2 years. They can occur in any location where lymphatics are normally encountered. The most frequently affected sites are the head and neck (75%), where these are commonly referred to as 'cystic hygromas', followed by the axilla (20%).[5] The remainder (approximately 5%) of the lymphangiomas are intra-abdominal[6] arising from the mesentery, retroperitoneum or greater omentum,[7] where they are referred to as 'omental or mesenteric cysts'.
Retroperitoneum is the second-most common location for the abdominal lymphangiomas followed by mesentery of the small bowel. Retroperitoneal lymphangiomas are best described as developmental abnormalities of the lymphatics, and are almost always benign.[8]
The majority of the retroperitoneal lymphangiomas are asymptomatic[9] and are discovered incidentally in later life during radiological procedures for other conditions, or during surgery or autopsy.
In the present study, all patients were male and of age group in 2nd and 3rd decade which is rare in contrast with common age group affected of less than 2 years.
Also the site of the two cases in the present study was retroperitoneal and one case was found to be in the ileal mesentery, which is also rare in contrast to most common site affected in head and neck.
The most common clinical manifestation is that of an abdominal lump,[8,10] abdominal pain, nausea and vomiting, no passage of stool, as seen in these cases.
Pre-operative diagnosis of retroperitoneal lymphangioma, in general, is challenging and rare, prior to laparotomy or laparoscopy[11] The diagnosis of lymphangioma, based on the radiological modalities, is generally one of many potential differentials for a multiloculated cystic mass arising retroperitoneally. Our preliminary radiological investigations, however, did include the possibility of a retroperitoneal lymphangioma.
Ultrasound (US), CT and MRI have been shown to be complimentary in the diagnosis of retroperitoneal lymphangiomas. Ultrasonography can demonstrate the cystic nature of the lesion appearing with sharp margins, particularly the septations as scattered internal echoes. CT and MRI can demonstrate uni- or multi-locular cysts with septae, an assessment of the relationship of lymphangiomas to neighboring organs[12]
The final diagnosis of lymphangioma is achieved by pathological examination of the specimen after surgical or laparoscopic examination, and is based on well-established

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criteria.[13] Histopathology shows variable-sized dilated cystic spaces lined by flattened endothelium filled with pale pink proteinaceous material, larger spaces with fascicles of smooth muscles, stroma showing acute and chronic inflammation, oedema and fibrosis along with small lymphoid aggregates.

Immunohistochemical markers that can be used for the diagnosis of lymphangioma include lymphatic vessel endothelial receptor-1, vascular endothelial growth factor-3, monoclonal antibody D2-40, CD31, CD34, and prox-1[14], [15], [16].

Complete surgical resection is the first-line treatment option for all cases, due to the risk of future complications.

Conclusion

Retroperitoneal lymphangioma is an uncommon lesion in adults, and radiological investigations provides important pre-operative diagnostic information for effective surgical approach and management. These rare tumors have an excellent prognosis, with symptomatic relief and cure achieved with complete surgical excision.

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