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Original Research Paper

Intra Abdominal Lymphangioma- A Case Report

Pathology

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ABSTRACT

Lymphangiomas are the commonest tumours of infancy and 75% occur in the cervical region. However, intra-abdominal lymphangiomas are rare, usually slow growing and manifest in childhood and early adulthood with varied clinical manifestation and imaging characteristic. Therefore intra abdominal lymphangiomas should be considered in the list of differential diagnosis of any abdominal lump. We present a case of intra abdominal lymphangioma in a seven year old female child because of its rarity.

Introduction:

Lymphangioma is a benign vascular neoplasm. It manifest in early childhood and develops in head, neck and axillary region commonly.1 Intra abdominal lymphangiomas are very rare affecting mesentry, greater omentum, retroperitoneum.2 The lesion is frequently misdiagnosed preoperatively because of its varied and confusing clinical picture. Therefore, histopathological analysis is important for diagnosis of lymphangioma. We report this case because of its rarity.

Case Report:

A seven year old female child presented with history of gradual distension of abdomen since six months and pain in abdomen since seven days. There was no history of nausea, vomiting or malena or altered bowel or bladder habits. On physical examination, abdomen was slightly distended with mild tenderness in right lumbar region. A vague soft mass was palpated in the right lumbar region. General examination revealed no other abnormality. Other routine investigations were well within normal limits.

Ultrasonography (USG) showed a hyperechoic lesion of 6×4×4 cm in right lumbar region extending medially and up to right lobe of liver.

Computed Tomography (CT) scan showed hyperdense, moderately enhancing lesion of 6.4×4×4.4cm in right lumbar and epigastric region pushing bowel loops peripherally, reaching posteriorly upto paravertebral plane suggesting possibility of lymphoma.(Fig 1)

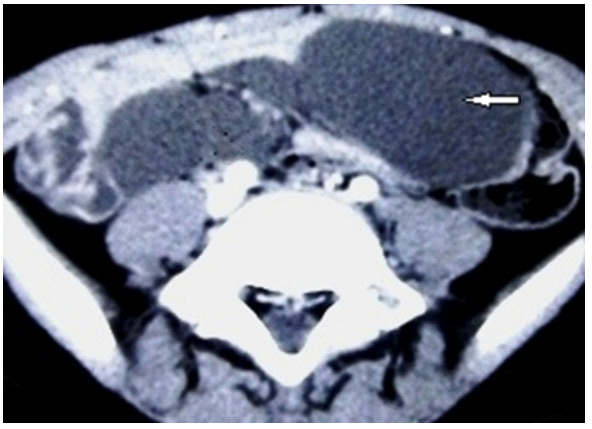

Fig 1: CT scan showing intra abdominal showed hyperdense, moderately enhancing lesion (indicated by arrow) of 6.4×4×4.4cm in right lumbar and epigastric region.

Considering the possibility of lymphoma, clinicians decided to take a biopsy to rule out the same. Biopsy was taken under all aseptic precautions and with due consent of parents.

We received a biopsy of 0.5×0.2×0.1 cm. Histopathological section revealed cystic spaces lined by single layer of endothelial cells. Cysts were filled with fluid at places and separated from each other by loose fibrous stroma showing focal and diffuse lymphocytic collection. There was no evidence of malignant lymphoid cells to be labeled as lymphoma. Considering these findings we offered a diagnosis of Intra abdominal Lymphangioma. (Fig 2)

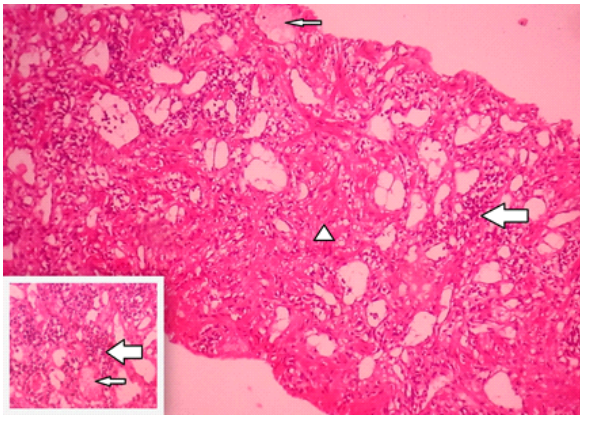

Fig 2: Microphotograph showing cystic spaces filled with fluid at places (small arrow), lined by single layer of endothelial cells with fibrocollagenous stroma (arrow head) showing diffuse lymphocytic collection (large arrow) (HE,100X). (Inset- HE,400X)

Unfortunately our patient didn't show up and we lost follow up of patient.

Discussion:

Lymphangioma is a rare malformative benign tumor of the lymphatic vessels with an incidence of 1 in 20,000-250,000 cases.3 It is described for the first time by Koch in 19134 thought to be resulting from sequestration of lymphatic tissue failing to communicate normally with the lymphatic System.5 It is preferentially located in the head, neck (75%), axillary region (20%) and other parts of body (2-5%) manifesting in childhood and young adults. However, the abdominal location is extremely rare accounting for less than 1%.2 Though benign these are thought to be locally invasive. Their clinical presentation is variable and misleading depending upon the site of involvement. Many patients are asymptomatic and lymphangioma may be an incidental finding.

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Some patients present with abdominal pain, nausea, vomiting and sometimes with abdominal lump and ascitis. [6]

Hence complex imaging studies such as USG and CT are used in their evaluation.7 The USG of intra-abdominal lymphangioma will show a cystic lesion with multiple thin septa (honeycomb or cobweb pattern). On CT imaging, it appears as a uni or multilocular mass with enhancement of the wall and septum by contrast medium.8 However these radiological investigations are insufficient to establish a exact preoperative diagnosis. The definitive diagnosis of lymphangioma is made on histopathological and immunohistochemical grounds. One of the clinically closest mimicker of intra abdominal lymphangioma is mesenteric cyst. Therefore it is important to differentiate between the two. In contrast to the mesenteric cysts which originate from the mesothelial tissue is usually uniloculated having cuboidal to columnar lining and absence of lymphoid elements in the wall9 intra-abdominal lymphangioma is usually a large multiloculated cystic lesion. The cysts may range from few mm to 5 cm containing serous, purulent or chylous fluid. Histopathological sections show cystic spaces line by endothelial cells. These cells have some capacity to proliferate but more importantly these accumulate vast amount of fluid which accounts for its cystic nature. Intervening stroma is loose fibrocollagenous with abundant lymphocytic collection either diffuse or focal. Foam cells and smooth muscles can be present in the wall. Sarno et al proposed that preoperative diagnosis of abdominal lymphangioma can be possible with guided fine needle aspiration cytology (FNAC). FNAC smears will show mature lymphocytes, lymphocytes with degenerative changes, fibrin on a fluid background.6 Immunohistochemistry for lymphatic lineage is ultimately reliable method for diagnosis. It will show immunoreactivity to PROX1 and VEGFR3 antibodies and podoplanin (D2-40). [10]

The definitive treatment for abdominal lymphangioma is radical excision even in asymptomatic cases. Incomplete resection follows local recurrence in about 10% of cases. Therefore follow up imaging with USG or CT scan is always advised. Sclerotherapy using doxycycline, alcoholic solutions of zein or with OK- 432 is another modality of treatment with favorable prognosis. [11]

Conclusion: Intra abdominal lymphangioma is a benign tumor of the lymphatic system, affecting mostly young children. Knowledge on the imaging and the pathologic spectrum of the abdominal lymphangiomas is necessary. Though USG or CT scan complements the diagnosis of intra abdominal lymphangioma, histopathological examination for the confirmation is recommended.

Conflicts of interest- none.
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