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Case Report

A rare case of intra-abdominal cystic lymphangioma: a case report

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ABSTRACT

An intra-abdominal cystic lymphangioma is a rare benign tumour in adults' uncommon location, originating from lymphatic vessels. Here we present the rare case of 73 year old female presented with right upper abdominal pain, routine blood and radiological investigations done with contrast CT scan showed gastro intestinal stromal tumour over 2nd part of duodenum posteriorly, electively operated and solid cystic mass excised, histopathology showed cystic lymphangioma. The rare looking cystic mass in rare location showing gist/hydatid cyst on CT scan surgical excision showed cystic lymphangioma, where treatment is to excise the mass to prevent recurrence or complications.

Keywords: Benign, Lymphatic vessels, Intra-abdominal cystic lymphangioma, Surgery, Recurrence

INTRODUCTION

An intra-abdominal cystic lymphangioma is a rare benign tumour of lymphatic system. Its a congenital connection between lymphatic and venous channel during embryogenesis, mostly it remains asymptomatic depending upon the location more common in children in head and neck region less common in abdomen.[1]

CASE REPORT

A 73-year-old female presented with right upper quadrant pain since 2-3 months, associated with nausea on and off for 1 month. On examination per abdomen was found soft non-tender. On investigation patient was a known case of ischemic heart disease hypothyroidism and hypertensive for which she was on regular medication.

Ultrasonography

Approximately 27×32×35 mm well defined solid cystic lesion with internal septation and solid component shows internal vascularity noted in relation to along the medial aspect of 2nd part of duodenum gist likely lesion causes mild compression of IVC.

CECT

Approx. 30×44×43 mm sized (ap×tr×cc) lobulated predominantly exophytic. Heterogeneously enhancing solid cystic lesion with internal non-enhancing necrotic. areas and thin internal septations noted arising from posterior wall of 2nd part of duodenum without causing significant luminal narrowing, it abuts rt renal artery 2nd part of duodenum and inferior vena cava. Suggestive of duodenal mass lesion-GI stromal tumor likely.

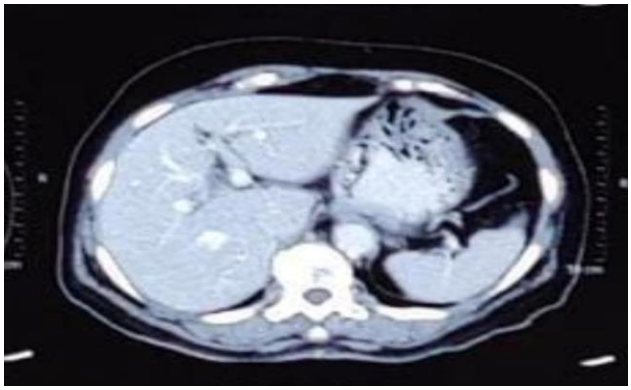

Figure 1: CECT showing heterogeneous mass lesion.

Patient underwent exploratory laparotomy with midline vertical incision and retro duodenal exophytic solid with cystic lump of approx. 4×3 cm found with adhesion to duodenum anteriorly and IVC, aorta posteriorly, lump excised without duodenal attachment intraluminal bluntly separating form surrounding vascular structures which was sent for histo pathological examination.

Suggestive of intra-abdominal cystic lymphangioma: multilocular cyst consisting of large channels with thin and irregular muscular coat collagenous stroma and smooth muscles, lumina containing are eosinophilic fluid RBC and lymphocyte collection in stroma.

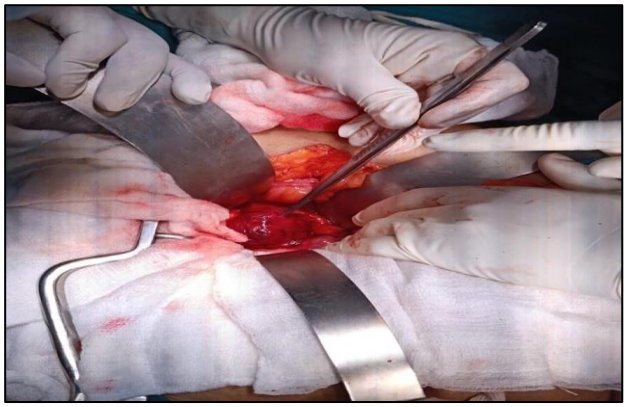

Figure 2: Cystic lymphagioma.

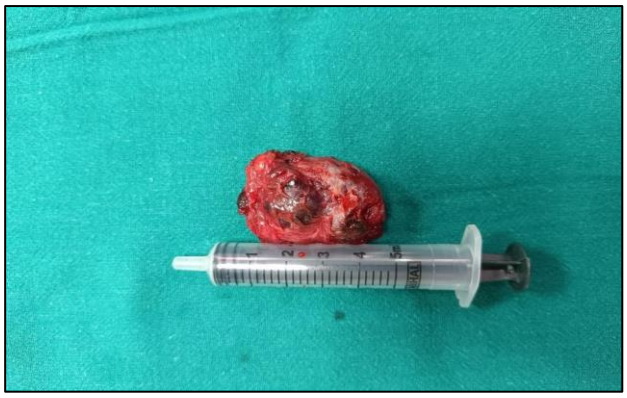

Figure 3: Approx. 3×3 cm solid cystic mass excised.

Endoscopic guided biopsy

Impression-cystic lesion in retroperitoneum hydatid cyst.

There is large lobulated defined nonvascular cystic lesion with daughter cyst in retroperitoneum adjacent 2nd part of duodenum.

DISCUSSION

Abdominal cystic lymphangiomas are extremely rare conditions affecting both paediatric and adult population, their clinical presentation is variable and misleading depending upon the site of involvement.

Hence complex imaging studies such as USG and CT are used in their evaluation. Depending upon size of cysts, they are classified as: macrocystic, microcystic and mixed.[2] Histopathological a lymphangioma is composed of thin-walled cysts lined by single row of flat endothelial cells. they also contain fibrous tissue, smooth muscles and aggregates of lymphoid tissue.

Complications of lymphangiomas include volvulus, intestinal obstruction, and haemorrhage or secondary infection they are divided into three main types according to their histological characteristics: capillaries, cavernous and cystic. The first two types are seen as skin lesions, while the cystic type presents intra-abdominal or retroperitoneal.[3]

With electron microscopy, two types of cells in them have been identified: endothelial and mesothelial. Based on this, they are divided into two groups: those of vascular origin (simple lymphatic cyst and lymphangioma) and those of mesothelial origin (simple mesothelial cyst, benign cystic mesothelioma and malignant cystic mesothelioma) three forms of clinical presentation have basically been established:

Incidental finding-chronic or recurrent picture of abdominal pain and acute abdomen syndrome secondary to complications (intracystic haemorrhage, cyst infection rupture, intestinal obstruction and compression of mesenteric vessels). Ultrasound itself can be used to detect the location, size, division of the cyst, cyst fluid, cystic wall and its relationship with surrounding tissues. the diagnostic protocol is completed with an abdominal tomography that is more sensitive, it helps to differentiate from other entities and also serves for the decision and surgical approach the definitive treatment is the complete excision of the cystic mass.[4]

Complete resection with negative margins on microscopy is the standard treatment for cystic lymphangiomas. Recurrence rates after incomplete and even complete resection are 35% to 64% and 17% to 24%, respectively.[5]

Simple aspiration followed by injection of sclerosing agents is considered ineffective because of infection and recurrence rates are high.[6] However, sclerotherapy has become the mainstay of treatment for macrocystic lesions because it is more effective than surgical resection and morbidity rates are lower. Although sclerotherapy might be a better treatment strategy, our patient underwent resection for a total biopsy, because preoperative assessment using various modalities could not completely exclude the possibility of a pancreatic tumour.

CONCLUSION

Here we described a patient with a rare, abdominal cystic lymphangioma derived from the lymphatic system. Preoperative imaging modalities have limited capacity to distinguish cystic lymphangiomas from pancreatic

tumours. Abdominal cystic lymphangiomas have the potential to grow, invade vital structures, and develop life-threatening complications. Therefore, laparoscopic exploration followed by total resection seems a reasonable strategy for treating peripancreatic cystic lesions that is less invasive than laparotomy.

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