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CT and MR Imaging of Retroperitoneal Chylolymphatic Cyst.

J Jayapriya *, I Venkatraman, and M Prabakaran.

Department of Radio-Diagnosis, Sree Balaji Medical College and Hospital, Tamil Nadu, India.

ABSTRACT

Intra-abdominal chylolymphatic cyst is a very rare entity. Retroperitoneal chylolymphatic cyst is rarest. We report a case of young female with swelling and pain in the right hypochondrium with retroperitoneal chylo lymphatic cyst. Early diagnosis prevents complications like rupture and potential risk of chylo peritoneum that increases the morbidity and mortality.

Keywords: retro peritoneum, cyst, chylo lymphatic, rupture, chylo peritoneum.

**Corresponding author*

INTRODUCTION

Chylolymphatic cyst is a rare entity .Retroperitoneal chylolymphatic cyst is even rarer. To the best of our knowledge so far no case report was documented. We have elaborated the findings in x-ray abdomen, ultrasonography, color doppler, computed tomography (CT), and magnetic resonance imaging (MRI), MRCP with intraoperative and histopathological reports correlation.

Case report

A 27 years old female presented with complaints of pain and swelling in upper abdomen for a week, radiating to the back. She had no other relevant past/surgical history or history of trauma. On clinical examination there was a palpable swelling in the right hypochondrium extending to the epigastrium. Biochemical tests were normal. Patient underwent radiological investigations x-ray abdomen, ultrasound, CT and MRCP examinations. X-ray abdomen showed a large ill-defined soft tissue opacity in the right hypochondrium and epigastrium displacing the transverse colon downwards and small bowel loops to the left. (Fig 1). Ultrasound findings- Large well defined thin walled anechoic cystic lesion (13.7 x 12.3 cm) without any internal septations or echoes seen in the region of porta hepatis adjacent to the gall bladder, displacing the liver laterally, elevating and displacing head of pancreas and part of CBD medially, IVC is displaced inferiorly. (Fig 2) Differential diagnosis was given with retroperitoneal cyst as the first diagnosis and Type II choledochal cyst as the second diagnosis .since the cystic lesion was very large the origin whether intra/ retroperitoneum could not be made out in ultrasound. CT abdomen screening showed hypo dense homogenous cystic lesion of near water attenuation with subtle wall calcification. (Fig 3)

Patient was referred again for MRCP to find the cyst communication with common bile duct. MRI images showed T1 hypo intense, T2 hyper intense thin walled large cystic lesion with homogenous signal intensities .This massive cyst was found to produce mass effect on the adjacent structures, displacing the left lobe of liver superiorly, gallbladder laterally, pancreatic head pushed to the left, aorta pushed medially, right kidney and IVC displaced inferiorly .Cyst was seen invaginating between the aorta and L2 vertebra(fig 4 a,b) .MRCP –CBD was draped along the lateral aspect of the cyst. No connection with biliary radicles was seen.(fig 4 c) Imaging diagnosis was given as retroperitoneal cyst (in view of extension of small portion of the cyst between the abdominal aorta and anterior aspect of vertebra at L2 level, separation – splaying of IVC/AORTA and elevation of head of pancreas.) - Probably lymphatic cyst. Hydatid cyst was given as a less likely differential due to the subtle calcification noted in the cyst wall in CT screening. Surgical excision was done and intra operative findings (fig 5 a, b) showed large cystic swelling arising from retroperitoneal region between IVC and abdominal aorta. A large thin walled transparent cystic lesion with whitish fluid within it. Thick and whitish chylous fluid was aspirated. Cyst wall incised and cyst aspirate was sent to histopathology.

Histopathology section (fig 5 c) showed cystic lesion lined by single layer of flattened cells overlying fibrous stroma enclosing lymphoid aggregates .Final report was given as chylolymphatic cyst.



Figure 1: Large ill-defined soft tissue opacity in the right hypochondrium and epigastrium displacing the transverse colon downwards and small bowel loops to the left.

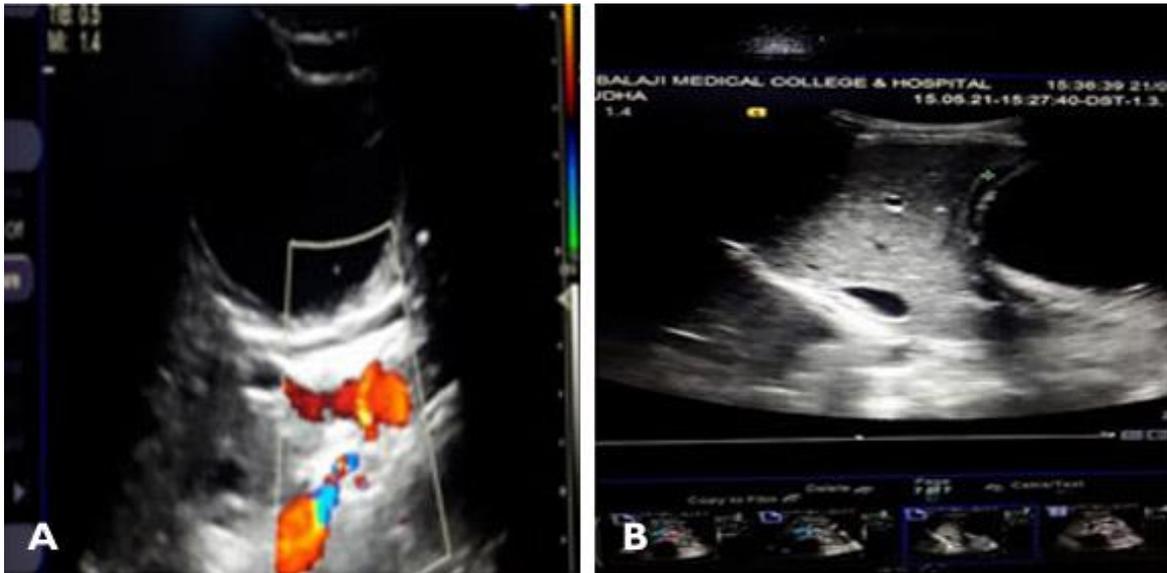


Figure 2: USG Large well defined thin walled anechoic cystic lesion adjacent to porta hepatis

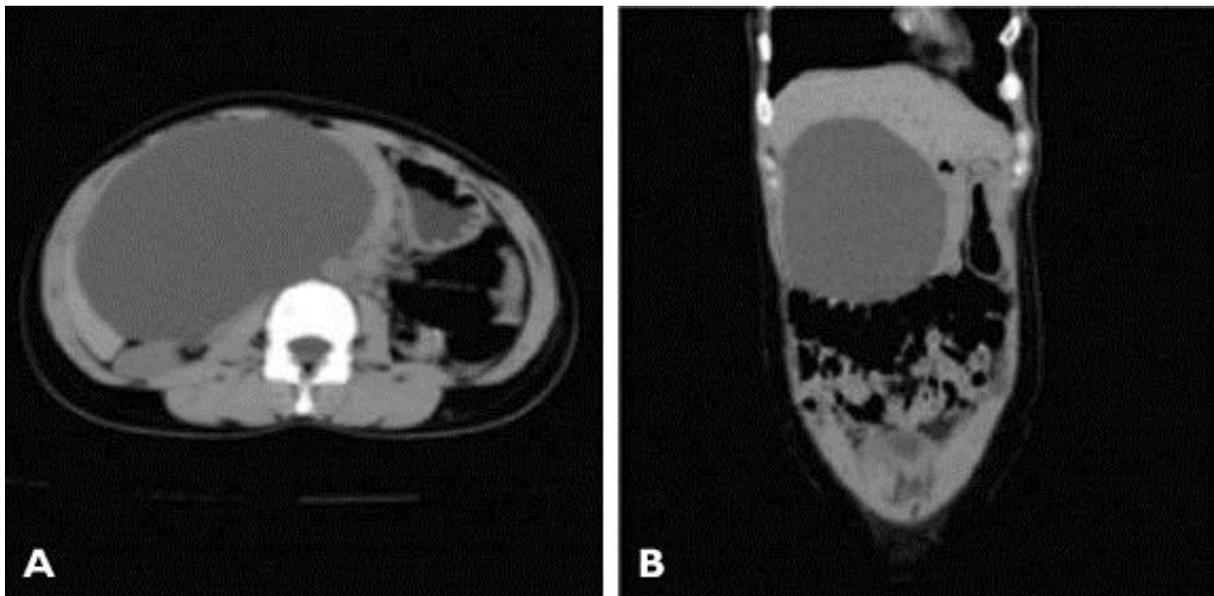


Figure 3: CT abdomen screening showed hypo dense homogenous cystic lesion of near water attenuation with subtle wall calcification.

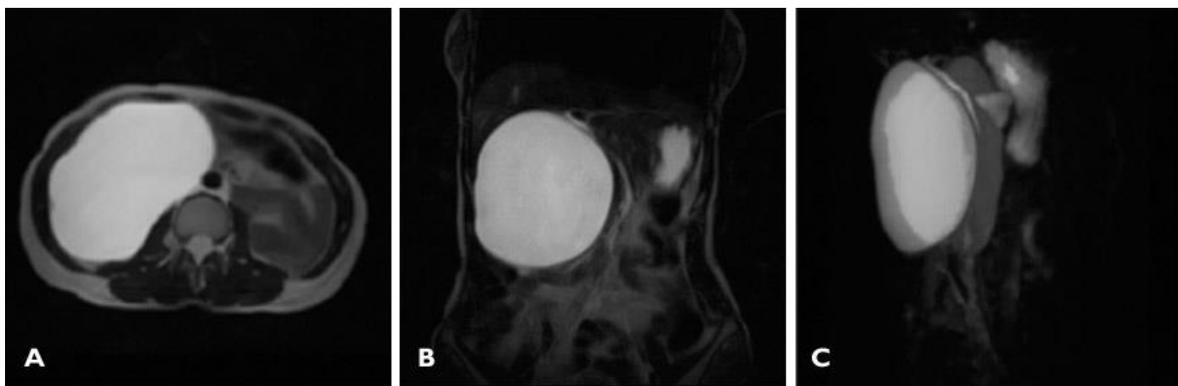


Figure 4: MRCP-CBD draped along the lateral aspect of cyst. Cyst was seen invaginating between the aorta and L2 vertebra



Figure 5 a, b-Intra operative images

Figure 5c-Histopathological slide - lymphatic cyst

DISCUSSION

Simple lymphatic cyst [1] are congenital, usually small (1 to 5 cm), unilocular, asymptomatic and it may contain clear or chylous fluid. Can be intraperitoneal or retroperitoneal.

CHYLOLYMPHATIC CYST

Chylolymphatic cyst arises from congenitally misplaced lymphatic tissue with no efferent communication with the lymphatic system. It is lined with thin endothelium and filled with lymph and chyle with varying consistency. It is unilocular or multilocular and may grow to very large size. It has different blood supply from intestine hence enucleation is possible without bowel resection [2] (Very rare .only 300 cases worldwide) [2].

Mesenteric chylolymphatic cyst is the most common site. [3, 4] these cysts are lined with a thin endothelium or mesothelium and filled with chylous and lymphatic fluid. Imaging findings include CT -simple fluid attenuation, may have minimal wall enhancement. MRI- Hypo/hyper intense in T1WI, Hyper intense in T2WI. Fat fluid level may be identified in case of high fat content. Chemical shift imaging potentially reveals the presence of microscopic lipid in the chylous fluid. Complications [3, 4] includes Super infection, Torsion, Hemorrhage and rupture leading to chyloperitoneum which increases the morbidity and mortality.

DIFFERENTIALS FOR RETRO PERITONEAL CYSTIC LESIONS

Cystic lesions of the retroperitoneum can be classified as either neoplastic or nonneoplastic.[5] Neoplastic lesions include cystic lymphangioma, mucinous cystadenoma, cystic teratoma, cystic mesothelioma, mullerian cyst, epidermoid cyst, tailgut cyst, bronchogenic cyst, cystic change in solid neoplasms, pseudomyxoma retroperitonei, and perianal mucinous carcinoma. Nonneoplastic lesions include pancreatic pseudocyst, nonpancreatic pseudocyst, lymphocele, urinoma, and hematoma.

A cystic retroperitoneal lesion can carry a relatively broad differentials. Retroperitoneal cystic lymphangioma is the closest differential diagnosis to this case. Lymphangioma is a benign proliferation of lymph vessels, producing fluid-filled cysts. It forms 1% of all lymphangioma more common in men, can occur at any age. Most common sites are neck (75%) and axillary region (20%). Retroperitoneal location is very rare. It usually crosses more than one compartment, with uniseptate or multiseptate appearance. CT and MRI features will show large cystic, unilocular or multilocular, thin walled, multiseptate cystic mass that tends to invaginate between rather than displacing the adjacent structures. Its attenuation values vary from that of fluid to that of fat. Low signal intensity on T1 and very high SI on T2 relative to skeletal muscle. Chylous fluid shows fat attenuation on CT. High signal intensity in both T1 and T2. Loss of signal intensity of internal fluid contents from in phase to opposed phase T1 weighted gradient echoes. Occasionally small amount of fat in septations with high T1 signal intensity is seen.



CONCLUSION

Imaging findings and intraoperative findings of the present case will increase the diagnostic confidence of both radiologists and surgeons in evaluating large retro peritoneal cystic lesion.

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