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A Case of Atypical Cystic Lymphangioma with Fatty Content

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Abstract **Case Report**

Cystic lymphangioma is a congenital anomaly typically detected in early childhood. It predominantly affects the cervical region but can also manifest in other areas such as the mesentery, omentum, and retroperitoneum. Complications, influenced by the lesion's location and size, underscore the critical need for early diagnosis to prevent potential abdominal complications like mesenteric ischemia, volvulus, and bowel obstruction. While radiological imaging may raise suspicion, histological examination remains the definitive diagnostic approach. Treatment primarily involves surgical excision. Here, we present a case of an intra- and retroperitoneal cystic lesion with fatty content on CT and MRI, initially suggestive of either a modified cystic lymphangioma or a teratoma. Subsequent histopathological analysis confirmed the diagnosis of cystic lymphangioma.

Keywords: Cystic lymphangioma; CT, MRI; adipose tissue, dermoid cyst.

INTRODUCTION

Lymphangiomas are benign vascular lesions displaying lymphatic differentiation, can present clinically in both pediatric and adult populations and occur in various anatomical sites. While the majority (95%) are located in the neck and axillary regions, the remaining 5% are distributed across locations such as the mesentery, retroperitoneum, abdominal viscera, lung, and mediastinum [1-2].

Intra-abdominal cystic lymphangiomas are rare entities posing diagnostic challenges radiologically, constituting less than 5% of all lymphangiomas. Mesenteric and retroperitoneal cystic lymphangiomas are even rarer and present a wide array of differential diagnoses when encountering cystic lesions in the mesentery [3].

PATIENT AND OBSERVATION

We report the case of a 57-year-old chronic smoker who presented with abdominal pain. Abdominal CT scans, both with and without contrast injection, revealed a roughly oval intra- and retroperitoneal lesion with well-defined borders, thickened walls in some areas, and spontaneous isodensity. The scan also showed regions of fat density (-50UH) and peripheral enhancement after contrast injection (Figure 1).

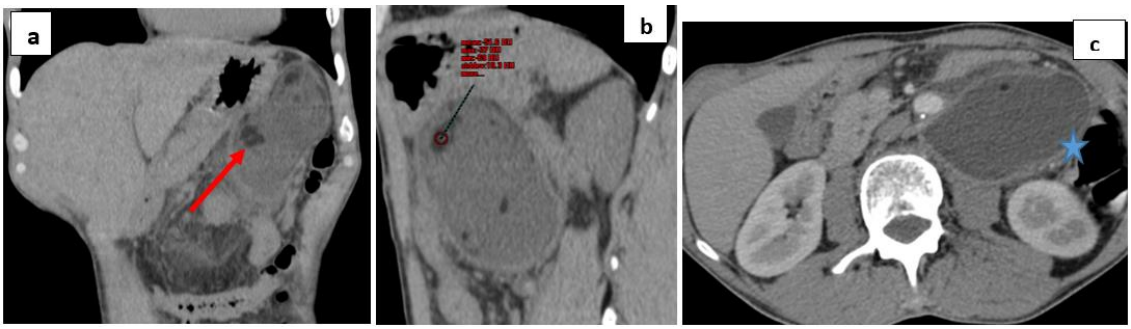

Figure 1: Abdominal CT scan without (a, b) and after iodinated contrast administration (c)

- intra- and retroperitoneal lesion, roughly oval in shape, well limited, isodense, with some areas of fat density (red arrow), thick walled, enhanced after iodinated contrast administration (blue star)

Subsequently, an additional abdominal MRI was conducted, confirming hyperintense signals on T1 and T2 sequences due to fat content. These signals diminished after fat saturation, along with infiltration of adjacent fat and diffuse densification of mesenteric fat (Figure 2).

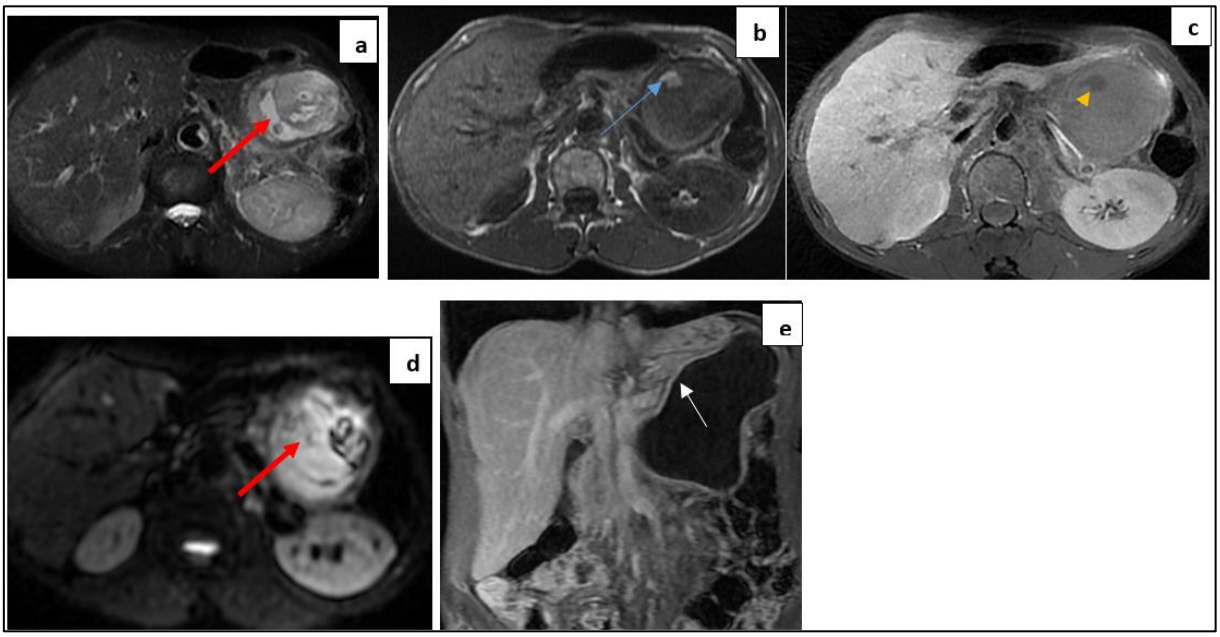

Figure 2: Abdominal MRI in axial and coronal sections with weighted sequences: T1 (b), T2(a), T1 Fatsat (c), diffusion (d) and T1 after injection of gadolinium (e) :

- Intra- and retroperitoneal lesion, roughly oval in shape, well limited, thickened wall in places, heterogeneous T2 and diffusion hypersignal (red arrow), seat of unenhanced partitions, containing some areas of fatty signal; T1 hyper signal (blue arrow) fading after fat saturation (arrowhead), peripherally enhanced after injection of gadolinium (white arrow)

The initial differential diagnoses considered were cystic lymphangioma, initially favored, or teratoma, which was less likely. However, further pathological examination confirmed the diagnosis of an atypical cystic lymphangioma.

DISCUSSION

Lymphangioma is an uncommon, hamartomatous, and congenital malformation of the lymphatic system typically observed in newborns and young children, but occasionally remains undetected until adulthood. Intra-abdominal lymphangiomas, situated in the mesentery and retroperitoneal space, represent less than 5% of all lymphangiomas [4-5].

Intra-abdominal cystic lymphangioma, though rare in adults, is a common type of mesenteric or omental cyst. Typically asymptomatic, it may induce non-specific symptoms such as abdominal distention, pain, nausea, and vomiting. Histologically, lymphangiomas are characterized by multiple dilated cystic spaces separated by minimal intervening stromata, comprising serous or chylous fluid lined with single-layered lymphatic endothelial cells and supporting connective tissue stromata. On ultrasound, cystic lymphangiomas typically present as unilocular or multilocular anechoic masses with smooth and thin or irregular and thick walls [6-8].

CT imaging usually reveals a smooth-margined multilocular cystic mass with homogeneous fluid attenuation. However, CT density of the fluid varies depending on factors like hemorrhage and lipid content, ranging from -4 to 34 HU. Complicated cystic lymphangiomas, such as those with infection or hemorrhage, may exhibit heterogeneous inner densities with high attenuation, fluid-fluid level, or calcification. On MRI, cystic lymphangiomas typically demonstrate low signal intensity in T1-weighted images and high signal intensity in T2-weighted images [9]. However, they can exhibit high signal intensities in both sequences due to varying ratios of fluid, fat, and hemorrhage within each cyst.

Differential diagnoses for predominantly cystic mesenteric lesions encompass various non-neoplastic and neoplastic entities. Non-neoplastic cysts include pseudocysts, enteric duplication cysts, mesothelial cysts, and dermoid cysts. Neoplastic cystic lesions comprise cystic mesotheliomas, cystic pancreatic neoplasms, and solid tumors with cystic degeneration like leiomyomas or leiomyosarcomas. Some of these conditions may exhibit a fat component in their cystic portion, including dermoid cysts, cystic lymphangiomas, lymphoceles, and liposarcomas with cystic degeneration. Identifying fat on CT scans is relatively straightforward if present in sufficient quantities to reduce CT attenuation

significantly below water density, but it can be challenging to discern when mixed with fluid [10-11].

For definitive identification of small quantities of lipids, comparing in-phase and opposed-phase images is optimal. The opposed-phase effect is twice as pronounced as fat saturation in tissues with minimal lipid content, as lipid signal interferes destructively with water signal in opposed-phase images [12]. Conversely, predominantly fatty tissues exhibit little signal reduction in opposed-phase images, as the fat signal persists even after water signal offset. In our case, fat signal detection was facilitated on CT due to negative density and on MRI with signal decrease on T1 sequences following fat saturation. This enabled us to determine the significant fat and water content within the mass.

The differential diagnosis of a cystic mass containing fat includes dermoid cyst, cystic lymphangioma, and lymphocele. Dermoid cysts typically manifest as thin-walled unilocular masses containing cholesterol, lined with squamous epithelium. They often exhibit homogeneous hypoattenuating fluid material on CT or heterogeneity due to various germinal components. Presence of fat attenuation within the cyst, with or without wall calcification, is diagnostic on CT. The cyst rim commonly enhances post-contrast administration. Dermoid cysts may appear hyperintense or isointense relative to muscle on T1-weighted MRI and hyperintense on T2-weighted images.

Differentiating fat from hemorrhage within a cystic lesion is crucial and can be achieved using fat saturation or chemical shift MR imaging techniques. Multilocular cysts with septa are more suggestive of cystic lymphangiomas, whereas dermoid cysts typically present as unilocular lesions originating from various skin appendages. Distinguishing between cystic lymphangiomas and dermoid cysts can be challenging, especially in cases of hemorrhage or infection [13].

Lymphoceles, fluid-filled cysts lacking an epithelial lining, often develop as post-operative complications following pelvic or retroperitoneal surgery, such as renal transplantation. On CT, lymphoceles appear as hypoattenuating masses typically located in the retroperitoneal or pelvic cavity [14]. While uncomplicated lymphoceles may exhibit low densities, complicated cases may demonstrate higher CT densities. Unlike cystic lymphangiomas, lymphoceles lack enhanced walls post-contrast injection. Surgical history can aid in lymphocele diagnosis.

CONCLUSION

MR imaging techniques, such as chemically selective fat saturation and chemical shift imaging, are valuable for determining the fat content of cystic lymphangiomas. When encountering a cystic mass with fat content in the abdomen, differential diagnosis should include consideration of cystic lymphangiomas alongside dermoid cysts.

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