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PANCREATIC CYSTIC LYMPHANGIOMA: DIAGNOSTIC APPROACH WITH MDCT AND MR IMAGING

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Lymphangiomas are rare congenital benign tumors arising from the lymphatic system mostly encountered in the neck and axillary regions of pediatric patients. Pancreatic cystic lymphangiomas very rarely occur in adults. Radiologically, the lesion may mimic pancreatic carcinoma and should be considered in the differential diagnosis of any patient found to have an abdominal cystic mass.
In this article, we present a 50-year-old man who presented with pain in the upper abdomen, nausea, and abdominal swelling. On computed tomography (CT) and magnetic resonance (MR) imaging, a gross septated cystic lesion was detected in the upper abdomen which extended from the pancreatic corpus to the left liver lobe. The patient underwent complete resection of tumor. Pathology revealed a cystic lymphangioma.

Key-word: Lymphangioma.

Case report

A 50-year-old man presented with a 2-week history of upper abdominal pain, nausea, and abdominal distention. The pain was relieved temporarily with analgesics. There was no history of alcoholism, jaundice, fever, vomiting, gastrointestinal bleeding, or weight loss. The patient did not have any prior history of pancreatitis. Physical examination revealed abdominal swelling occupying the upper abdomen. Blood investigation, liver function tests and tumor markers were within normal limits. An ultrasound exam showed a large septated cystic mass that extended

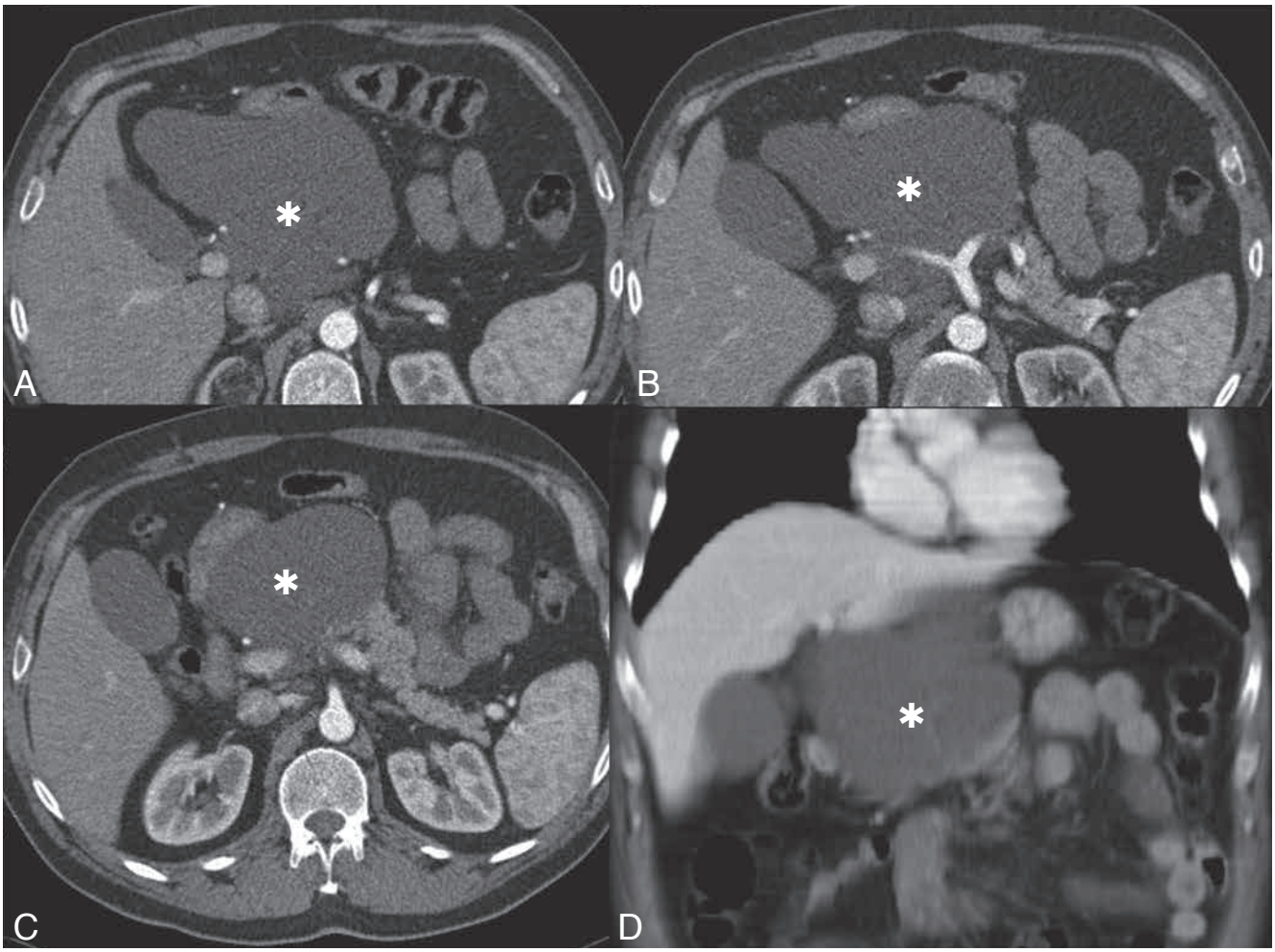

Fig. 1. — Contrast-enhanced MDCT axial (A-C) and coronal reformatted scan(D): a large hypodense multiloculated cystic mass is demonstrated over the pancreas (asterisk).

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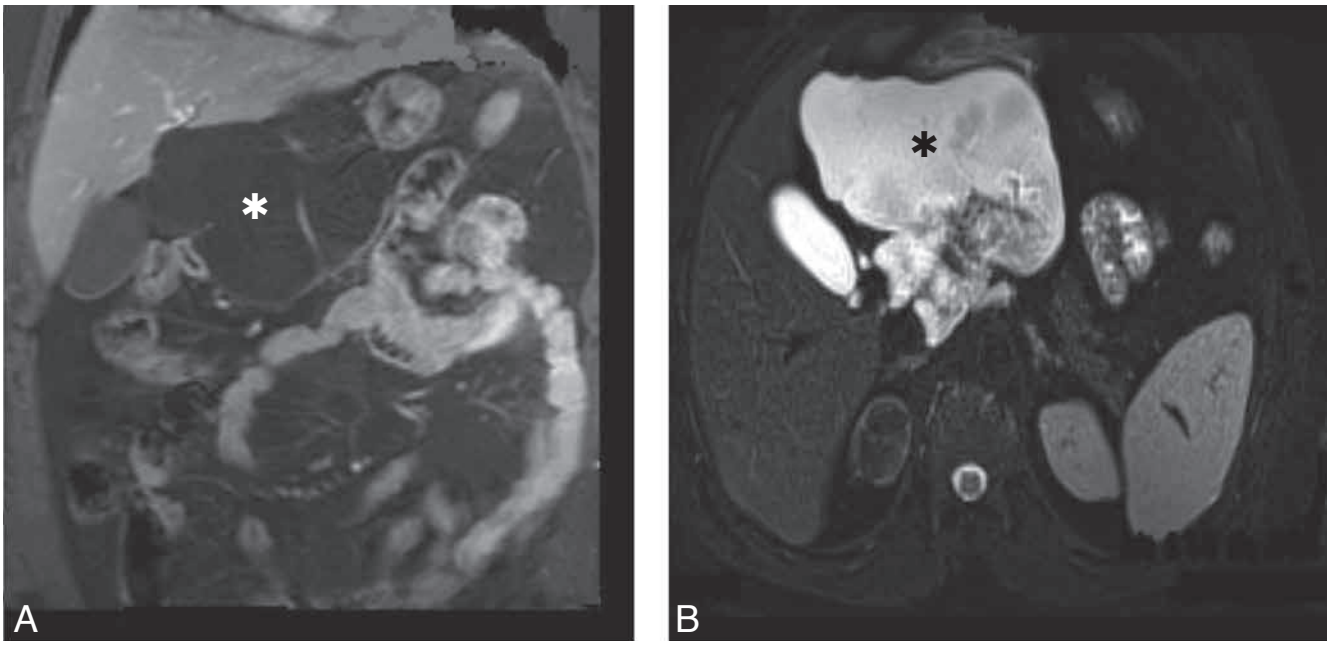

Fig. 2. — Lobulated pure cystic lesion without any solid component is seen on coronal plane contrast-enhanced, fat saturated SE T1 image (A). B: Lympangioma shows heterogeneous-hyperintense signal on fat- saturated TSE T2 image. Granular pattern due to high internal density at the posterior component of the cystic mass on fat- saturated TSE T2 image is noted.

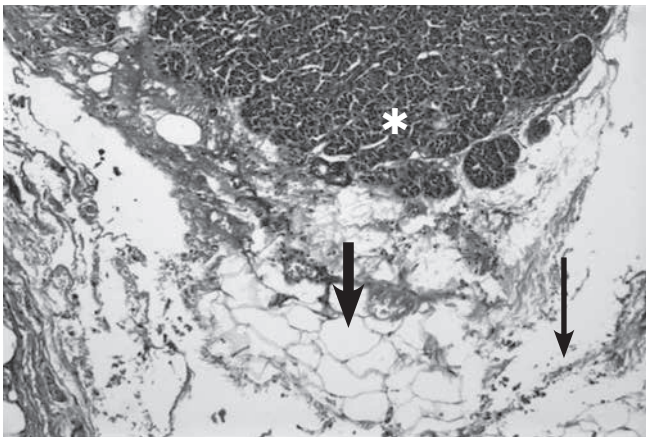

Fig. 3. — Histology shows pancreatic tissue (star), lymphatic vessels (thick arrow), and aggregates of lymphoid cells (thin arrow).

form the left lobe of the liver to the splenorenal angle, lying over the pancreas and aorta. The pancreas could not be identified separately from the mass.

Contrast-enhanced multidetector computed tomography (MDCT) scan revealed 15 x 11 x 10 cm non-enhancing, well-defined lobulated cystic mass with multiple septations located over the pancreas. The attenuation coefficient of the mass was in the +10 to 15 HU range generally. The mass extended from the tail of the pancreas to left liver lobe. Solid component of the lesion was not detected in the mass. The arterial branches of the truncus coeliacus were seen through the lesion without invasion signs. Thus, the mass had not invaded any adjacent abdominal organs such as the liver or kidney. There was no evidence of lymphadenopathy on the MDCT scan neither (Fig. 1 A-D).

Abdominal MRI scan showed a large, multiloculated cystic lesion with typically T1 hypointense, T2 hyperintense signals, Thick fibrous septa were seen hypointense on T1 and T2-weighted images. A granular pattern due to high internal density at the posterior component of the cystic masswas also noted (Fig. 2B).

The patient was operated on with the preoperative diagnosis of pancreatic cystic neoplasia. At laparatomy, upper peritoneal cavity was found to be almost fully occupied by a multicystic, septated mass containing a clear yellowish fluid, arising from the corpus of pancreas. There was no evidence of invasion into other adherent organs and main vascular structures such as portal vein and truncus coeliacus. Distal pancreatectomy including the cyst and the distal pancreatic tissue was performed.

Macroscopically, the surface of the cystic mass was shiny and smooth, and serial sectioning revealed multiple cysts ranging 1 to 3 cm in diameter, filled with yellowish fluid. Histologically, the mass was composed of variable-sized cystic spaces lined by flattened endothelium, consistent with lymphatic vessels. Lymphoid aggregates were detected in stroma and nearly all of them were filled with yellowish proteinous material (Fig. 3). The diagnosis of cystic lymphangioma was confirmed histologically.

Discussion

Cystic lymphangiomas are benign cystic tumors that are the result of congenital lymphatic malformations causing obstruction of the lymphatic flow leading to lymphangiectasia.

These malformations result in blockage of the lymphatic flow; thus, cystic dilatation of lymphatic channels occurs (1, 2). Another hypothesis is an inflammation leading to obstruction along the lymphatic channels (1-3). Lymphangioma is mostly detected in the pediatric age group patients. They are mostly localized in the neck (75%) and the axilla (20%) with cystic and cavernous spaces (1). Abdominal lymphangiomas account for less than 1% of all lymphangiomas and are more frequent in female adults (4). These masses generally present with vague abdominal symptoms such as abdominal distension, pain, nausea, vomiting, and palpable mass if it is big enough. However, months or years may pass from the onset of symptoms until the diagnosis especially in adults. Occasionally, acute abdomen can occur due to the complications such as intestinal obstruction, rupture or internal hemorrhage. Although a lymphangioma is a benign lesion, it can be locally invasive and can recur after incomplete excision of the cysts (5). Pancreatic pseudocysts, mucinous and serous cystadenomas, and congenital cysts should be considered for the differential diagnosis of gross abdominal cystic lesions.

Most reports in the literature describe the body and tail of the pancreas as the most common regions involved by lymphangioma (6). Recent studies showed that tumor size may vary between 3 and 30 cm in diameter (7).

Lymphangiomas typically appear as complex cystic masses on ultrasonography (US) due to internal septa and calcifications. They also may have internal echoes if they are secondarily infected. Rarely, calcifications could be demonstrated by US with acoustic shadow sign (8). Therefore, US can sometimes differentiate various cystic lesions, but ancillary studies are needed.

MDCT is considered superior to US, and provides additional information about the characteristics of the lympangioma. Cystic lympangiomas typically appear as well encapsulated, water-density, multiloculated masses. The septa can usually be seen and rare calcifications may be present. Otherwise, we could not see any calcification in this case.

MR imaging can demonstrate uni- or multi-locular cysts with septa in abdomen, an assessment of the relationship of lesions to neighboring organs with multiplanar imaging. The capability of MR imaging is to provide images in multiple planes without loss of resolution that may demonstrate additional pathologies like as internal hemorrhage, rupture, or other else exactly (9).

T2-weighted images showed multiloculated hyperintense areas which corresponded to dilated lymphatic spaces. The septa of the lesion appeared as hypointense bands which corresponded to fibrous tissue.

Histopathological examination plays an essential role in diagnosis. Cystic lymphangiomas are multi-loculated soft cystic masses composed of a combination of variable size, dilated lymphatic channels divided by thin septa. Flat endothelial cells line are seen in the cyst walls as well as focal aggregates of lymphoid tissue (10).

A complete surgical excision is curative for lymphangioma. Incomplete excision is the only reason for recurrent disease (3). Depending on the tumor location and size, complete excision may require pancreatic resection, such as a Whipple procedure or distal pancreatectomy. In our case, we needed to perform distal pancreatectomy for the removal of the lesion.

In conclusion, it is often difficult to establish a precise diagnosis of a pancreatic cystic masses. However, combined evaluation with MDCT and MR imaging may be helpful for a diagnostic approach to complex pancreatic cystic masses. Although rare, intraabdominal lymphangiomas should be included in the differential diagnosis of abdominal cystic lesions. The exact diagnosis still mainly relies on pathological examination.

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