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Retroperitoneal lymphangioma as the final diagnosis of a middle-aged woman with abdominal pain

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Abstract
Lymphangiomas are lesions attributed to the congenital malformations of lymphatic system or acquired chronic obstruction of the lymphatic network due to trauma, radiation, surgical manipulation, inflammation or infection. Overall, lymaphangiomas are rare and particularly retroperitoneal lymphangiomas are far more sparse per reported cases. In this study, we have introduced a middle-aged woman admitted to the gastroenterology unit with a progressive abdominal pain since approximately one month before her visit. She was found to have a retroperitoneal lymphangioma after a precise medical work up.

Keywords: lymphangioma, celiac artery, retroperitoneum, pancreatic cancer

Introduction

Lymphangiomas are rare benign lesions originating from lymphatic system [1]. They can be detected at any age but their infantile type is more common. Any part of the body can be involved but, head, neck and axilla are more commonly affected. Abdominal lymphangiomas in particular retroperitoneal forms are found to be rare. It is estimated that less than 1% of overally detected lymphangiomas are located in retroperitoneum [2, 3]. Their clinical presentation is dependent entirely on the place they have arised from. The compressive effect of the tumor on the adjacent organs determines the symptoms. Of note, retroperitoneal forms are usually manifested by abdominal pain and palpable masses as well [4]. Ultrasound (US), computed tomography (CT), and magnetic resonance imaging (MRI) are modalities used for evaluation of these tumors which clearly manifest their cystic nature. However, imagings suffer from a poor ability in differentiating these lesions from other cystic lesions. This may be in part due to their inflammatory changes which obscures their lymphatic origin. Given that, histologic confirmation is a necessity for definite diagnosis of lymphangiomas [5, 6]. Complete resection of these lesions is introduced to be the treatment of choice [6].

Case presentation

A 49-year-old woman presented to the gastroenterology clinic of Taleghani Hospital; a tertiary academic hospital in Tehran, Iran with a persistent epigastric pain radiating to the back since 1 months before admission. It was found to be aggravated over time. It was not considered to be altered by fasting or eating. She denied nausea, vomiting, any alteration in bowel habits and appetite. She did not mention any systemic signs and symptoms including fever, sweating and significant weight loss. Her past medical history was unremarkable except for uterine fibroids. Family history was positive for pancreatic ductal adenocarcinoma and gastric adenocarcinoma in her second-degree-relatives. On admission, she was uncomfortable but her vital signs were stable. Her physical examination was only revealed a mild to moderate tenderness in epigastrium without rebound and gaurding. A complete blood count revealed mild anemia with white blood cells (WBC): 5100 cells per cubic millimeter (reference range: 3400-9600 cells/mcl), hemoglobin (hb): 11.2 gr/dl (reference range in women: 12.1-15.1 gr/dl) and platelets (plt): 210000 per microliter (reference range: 150000-450000 per microliter). Further evaluations for anemia demonstrated a normocytic and normochromic anemia with a serum iron profile compatible with iron-deficient anemia. Erythrocyte sedimentation rate (ESR), Renal function tests, liver function tests, pancreatic enzymes, coagulation tests and serum bilirubin level were all within normal limits. Abdominopelvic ultrasound revealed a heterogenous hyperechoic lesion about 48×40mm at the head of the pancreas adjacent to the liver hilum. Color Doppler study of abdominal vessels was unremarkable. Spiral chest CT scan was normal. Abdominopelvic CT scan showed a well-defined hypodense lesion measuring about 45×26 mm in size which resembled a soft-tissue mass of unknown origin without clear enhancement near celiac trunk abutting left gastric artery. Adjacent mesentric vessels were severly engorged. An increased gastric wall thickness was noted and porta hepatis lymph nodes smaller than 10 mm were also detected (Figure-1). To assess a detailed image of the mentioned lesion, an endoscopic ultrasound (EUS) was utilized which demontrated a hypoechoic lesion measuring 43×35 mm lying at the posterior wall of the stomach. Tumor encasement of the left gastric artery and a short segment of celiac trunk without obstruction was detected. Elasto ratio of the lesion was estimated to be 22 (Figure-2). To reach a definit diagnosis and obtain specimens, fine needle aspiration (FNA) was performed which was unfortunately indeterminate (Figure-3). Inconclusive results of FNA made us proceed with a surgical resection of the lesion. Surgical exploration revealed a

retroperitoneal tumor adherent to celiac trunk, pancreas and left gastric artery which was resected and was sent for pathologic evaluation. Damaged arteries were reconstructed. Histologic assessment of the resected mass revealed large lymphatic channels with peripheral lymphoid aggregations embedded in a loose connective tissue stroma diagnostic for a benign vascular neoplasm with foci of hemorrhage compatible with lymphangioma (Figure-4).

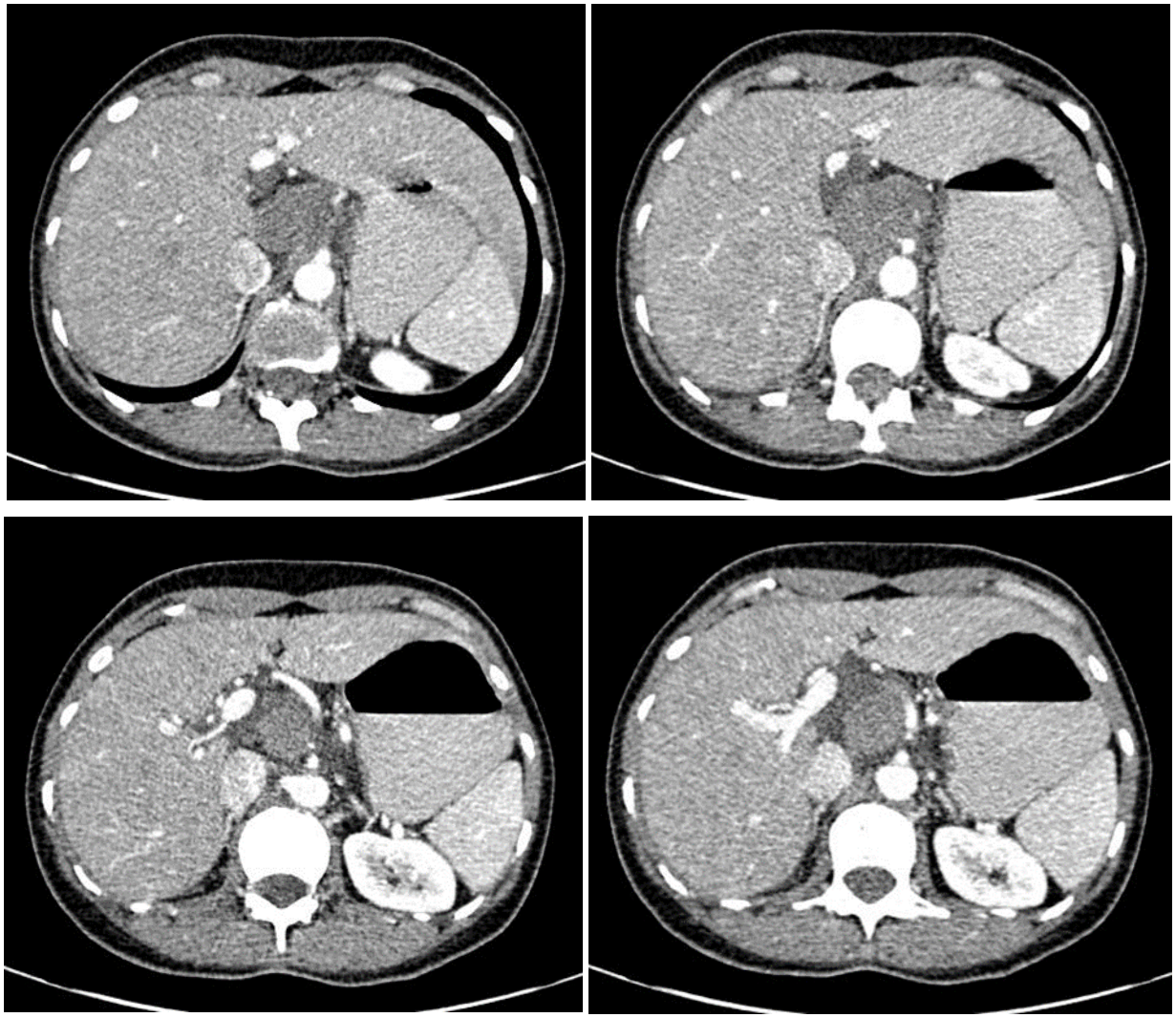
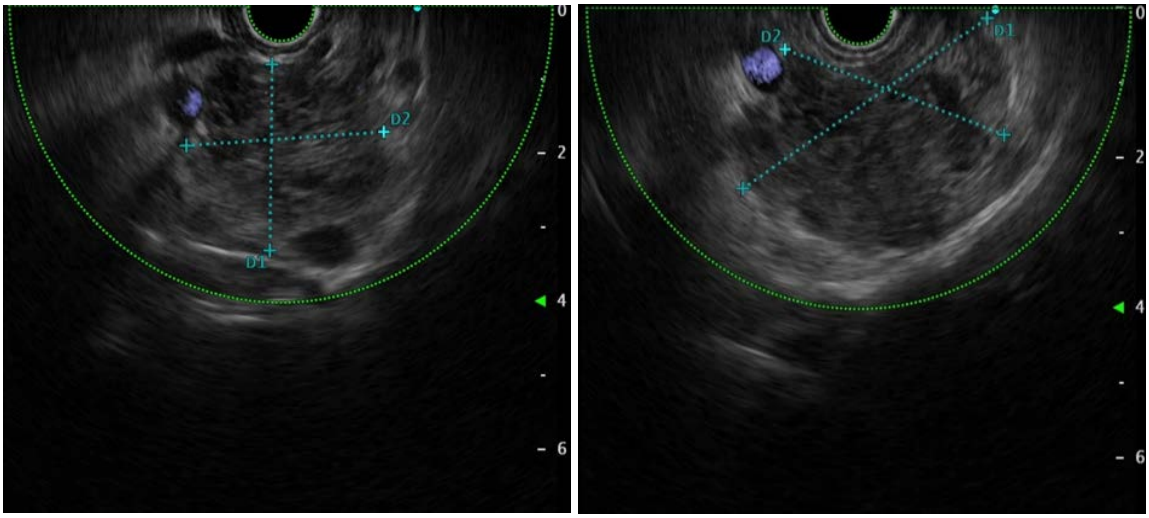


Fig 1: Abdominopelvic CT scan reveals a well-defined hypodense lesion measuring about 45×26 mm in size which resembles a soft-tissue mass of unknown origin without clear enhancement near celiac trunk abutting left gastric artery.



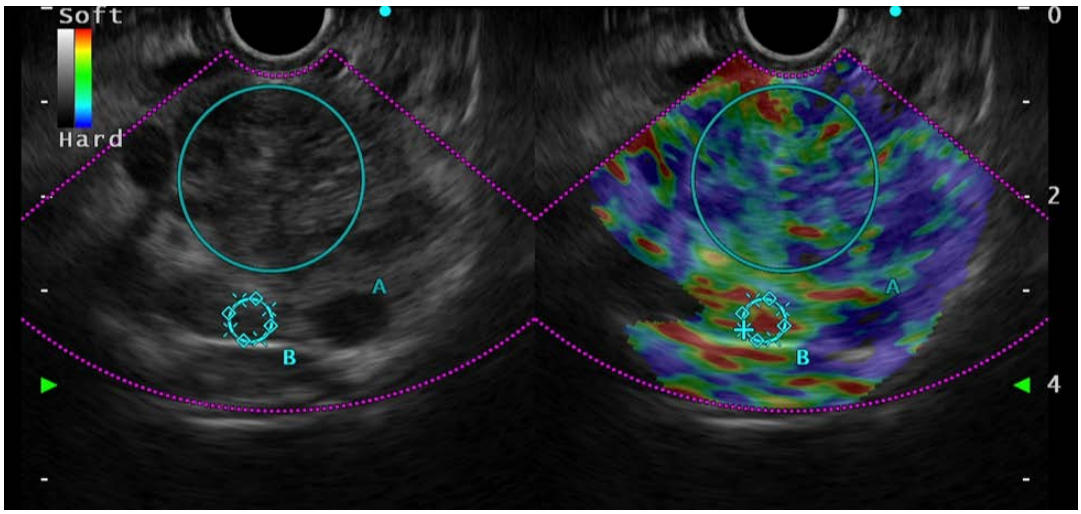


Fig 2: Endoscopic ultrasound study demontrated a hypoechoic lesion measuring 43×35 mm lying at the posterior wall of the stomach. Tumor encasement of the left gastric artery and a short segment of celiac trunk without obstruction was detected. Elasto ratio of the lesion was estimated to be 22.

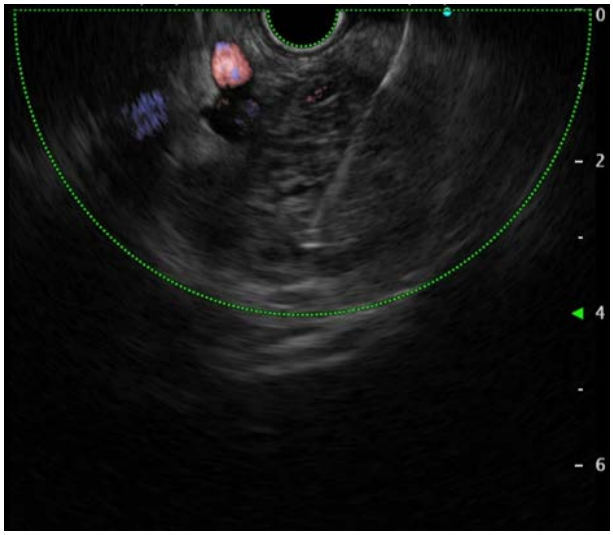


Fig 3: Fine needle aspiration of the lesion.

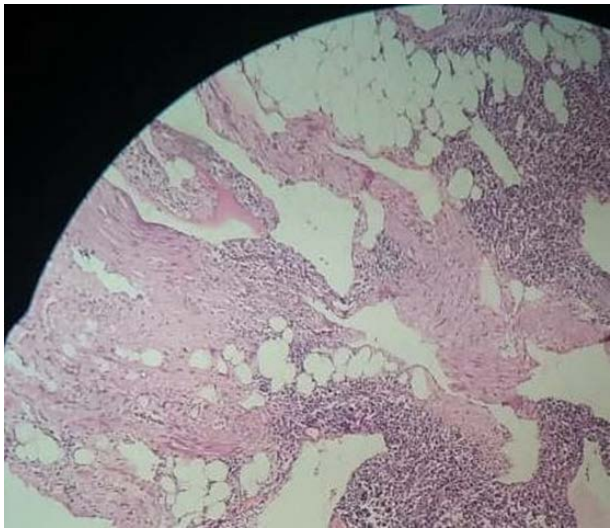


Fig 4: large lymphatic channels in loose connective tissue stroma with peripheral lymphoid aggregations compatible with lymphangioma.

Discussion

Lymphangiomas are slow-progressing tumors which are not reported to harbor malignant potential [1]. The known etilogies for lymphangiomas are the anomalous connection between lymphatic and venous network which is commonly seen in children as well as traumatic degeneration of lymphatic system by radiation, inflammation, infection and surgical resection resulting in chronic obstruction of lymphatic system [2, 7, 8]. Histologically, they can be cystic, capillary or cavernous [7]. Retroperitoneal lymphangiomas are rare and are more commonly seen before 20. Our presented case was a 49-year-old woman with retroperitoneal lymphangioma presented with abdominal pain. Our patient age, her positive family history for pancreatic ductal adenocarcinoma and the placement of the tumor adjacent to the head of the pancreas raised the suspicion of other cystic neoplastic tumors like intrductal papillary mucinous neoplasms (IPMNs), mucinous cystic neoplasms (MCNs) and pancreatic adenocarcinomas. Other benign lesions including pseudocysts, hydatid cysts and cavernous hemangiomas lied in the differential diagnosis of the reported lesion, too. A large number of cases are asymptomatic, so these tumors are incidentally discovered when imagings were utilized for other reasons. Among the rare reported symptoms, abdominal pain is more common and is attributed to the compressive effect of the mass to the adjacent organs. An acute abdomen would be anticipated if complications like intestinal obstruction, cystic infection, intracystic/intraperitoneal/retroperitoneal hemorrhage, torsion or cystic rupture occured [4]. Per the revised literature, hematoma, abscess, duplication cysts, ovarian cysts, tratoma, mesothelioma, cystic metastases, lymphangiosarcoma and pancreatic cystic neoplasms are introduced as differential diagnosis for retroperitoneal lymphangiomas [7]. In the literature, US study of lymphangiomas usually reveals well demarkated unilocular or multilocular cysts with scattered echoes [9]. CT images reveal well-defined homogeneous cysts with prominent walls and distinct septations. These images can accurately provide definite informations on the structure and location of these tumors [10]. MRIs also help to visualuize these cysts as they have distinct characteristics in T_1 and T_2 images [11]. A wide range of

differential diagnosis for the detected lesion on abdominopelvic CT scan of the presented case made us proceed with a more accurate imaging technique like EUS+FNA. But, it did not provide us with a definite diagnosis. Diagnostic challenges for detection of lymphangiomas on imagings make histologic examination of the resected tumor be emerged as a master plan to help the diagnosis of the lesion. Surgical resection of the tumor was performed and histologic examination on resected tumor revealed large lymphatic channels in loose connective tissue stroma with peripheral lymphoid aggregations which were compatible with lymphangioma. Recurrences have been reported after complete and incomplete resection of about 7% and 50% of the cases, respectively. So, free margins of surgically resected tumor is the cornerstone of the treatment [12].

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