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Case Report

Intra-abdominal lymphangioma: A case report

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ABSTRACT

Lymphangioma is a rare, benign congenital malformation of the lymphatic system that usually affects the neck and head in children. Intra-abdominal lymphangioma accounts for less than 5 percent of all cases of lymphangioma. The clinical presentation of intra-abdominal lymphangioma can vary from asymptomatic to nausea, vomiting, and abdominal pain. The diagnosis of intra-abdominal lymphangioma is based on imaging modalities and histopathological examination. The definitive treatment is surgical resection. Here we describe the interesting and rare case of a 29-year-old woman with lymphangioma of the retroperitoneum extending to the root of the mesentery. We focus on the diagnosis and management of this rare tumor by the application of radiological modalities and pathological analysis.

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Introduction

Lymphangiomas are uncommon, noncancerous lymphatic malformations composed of thin-walled cysts. While lymphangiomas are most often found in the neck, head, and axillary regions, in fewer than 5 percent of lymphangiomas can present intra-abdominally, including the mesentery, greater omentum, and rarely in the retroperitoneum. They can occur at any age but predominantly appear in pediatric populations with a great prognosis [1,2]. Intra-abdominal lymphangioma is often asymptomatic but in some cases present with nausea, vomiting, or acute abdominal pain due to bowel obstruction.

They can be identified by imaging modalities such as sonography and computed tomography but the histological exam is essential for confirming the diagnosis. Complete surgical resection is the recommended treatment with a low risk of recurrence [3–5].

Case report

A 29-year-old woman presented with a 3-month history of intermittent diffuse abdominal pain that was not relieved by the antacid or eating a small portion of meals. It was accompanied

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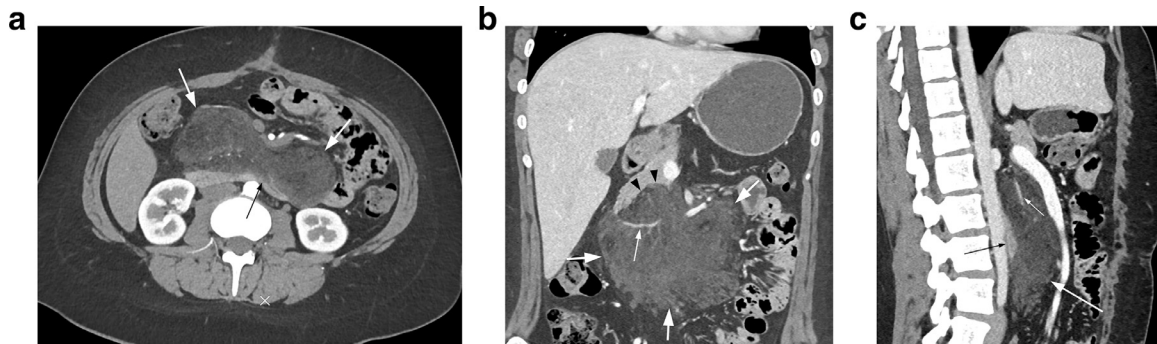


Fig. 1 – Contrast-enhanced arterial phase axial (A) and venous phase (B) coronal and (C) sagittal reformatted computed tomography images show a relatively ill-defined heterogeneous mass of mixed solid and cystic attenuation (large white arrows) in the retroperitoneum extending along the small bowel mesentery measuring up to 13 cm. The mass abuts the superior mesenteric artery and vein and their branches, and completely encasing the posterior inferior pancreaticoduodenal artery (small white arrow). The mass exerts mass effect on the duodenum (black arrow) and uncinate process of the pancreas (black arrowheads). There is suggestion of duodenal wall involvement on axial image (black arrow) (A).

by nausea, vomiting of undigested food an hour after oral intake not related to specific foods, and a 15-pound weight loss. She denied fatigue, fever, or bowel movement changes. A CT scan was performed at an outside institution that showed a large mixed solid and fatty retroperitoneal mass compressing the duodenum. Based on the location and appearance, liposarcoma was suspected, and the patient underwent exploratory surgery with curative intent. However, because of excessive vascular involvement, the tumor was not resectable. Multiple excisional biopsies of the mass were performed, with the frozen evaluations were not diagnostic for malignancy but leaning toward benign pathology, and only appendectomy and gastrojejunostomy bypass were performed with the remainder of the surgery aborted. The final pathology of the excisional biopsy was consistent with lymphangioma. Two weeks after the operation, the patient began to experience intermittent episodes of nausea and vomiting again. She denied any abdominal pain or weight loss in this episode. She underwent colonoscopy due to concern for colon involvement, which was the normal per report.

Two months later, the patient came to our hospital for a second opinion. CT at this time showed a relatively ill-defined heterogeneous retroperitoneal mass in the mid-abdomen measuring approximately $13 \times 4.5 \times 12$ cm extending along the small bowel mesentery (Fig. 1A, B, C). The mass had mixed fat and fluid density components and had a lace-like appearance. The mass was closely contacting and exerting mass effect on the uncinate process of the pancreas, and the third and fourth portions of the duodenum. There was suggestion of duodenal wall involvement (Fig. 1A). The mass abuts the superior mesenteric vein and artery and its proximal jejunal branches up to 180 degrees. The mass completely encased the posterior inferior pancreaticoduodenal artery. Multiple prominent small bowel mesenteric lymph nodes were seen. Three-dimensional cinematic rendering CT images show a low attenuated large mass in the retroperitoneum and root of the mesentery with displacing vessels and bowel with fatty appearance (Fig 2). The patient was consented to exploratory laparotomy and tumor resection and underwent open excision



Fig. 2 – Contrast-enhanced venous phase 3-D CT image of mesenteric lymphangioma with cinematic rendering in the coronal plane shows a low attenuated large mass in the retroperitoneum and root of the mesentery with displacing vessels and bowel with fatty appearance (white arrows).

of the retroperitoneal mass, resection of third and fourth portions of the duodenum, Roux-en-Y duodenojejunostomy, and jejunojunostomy, and pyloric exclusion. The post-surgery pathology revealed a $21.3 \times 13.9 \times 5.2$ cm mass attached to the duodenum with numerous dilated lymphatic spaces that confirmed the diagnosis of retroperitoneal lymphangioma which involved the duodenal submucosa, muscularis propria, and mesenteric adipose tissue (Fig. 3A, B). The patient tolerated the procedure well and was transferred to the intensive care unit for hemodynamic monitoring and support. She was discharged one week after surgery in good condition without pain or any complications and with written instructions encouraging protein intake and avoiding foods extremely high in fat and/or fiber at the beginning of her recovery phase, and scheduled follow-up appointments.

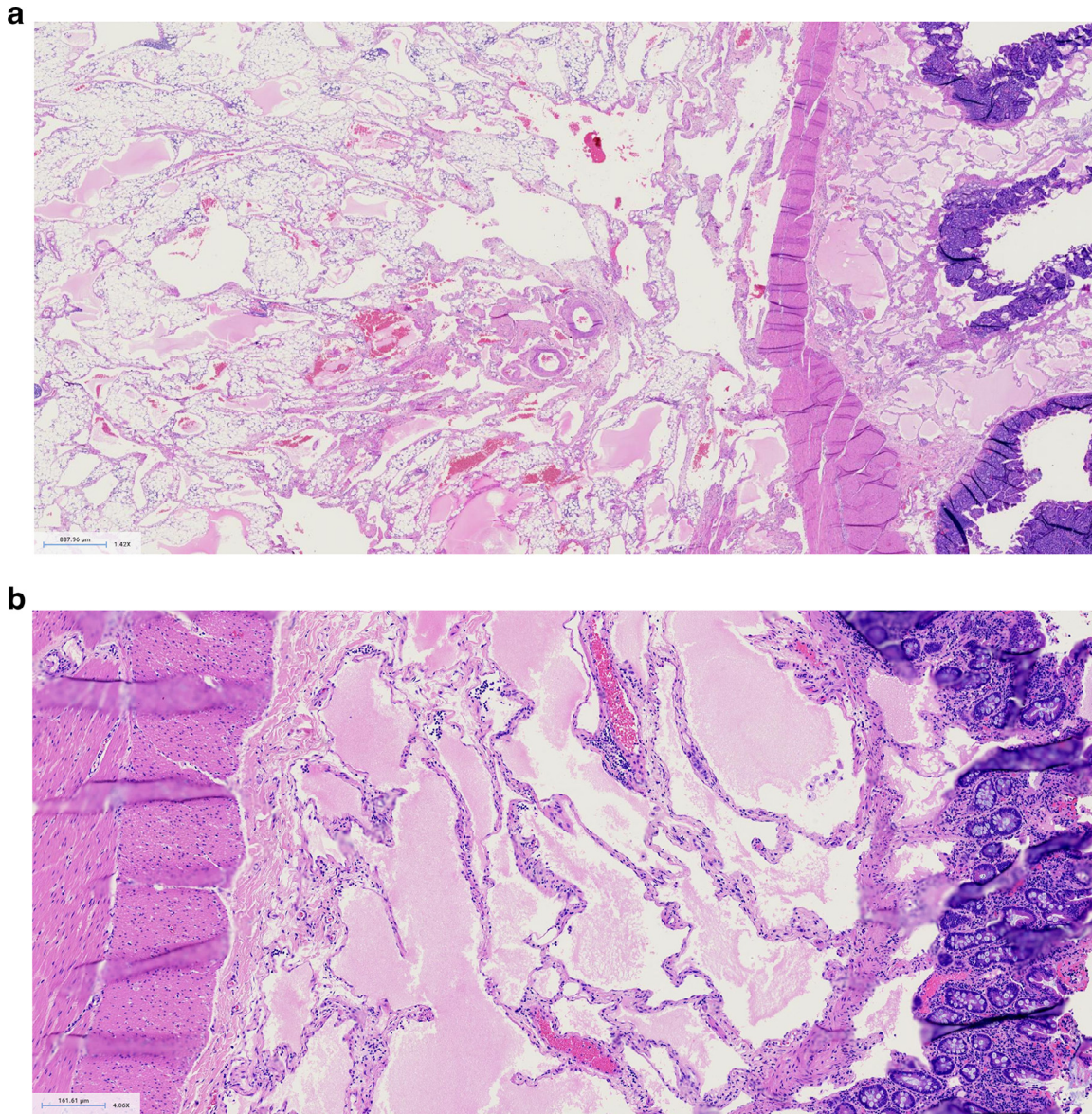


Fig. 3 – Histopathologic evaluation of the retroperitoneal lesion. (A) Low power view of a hematoxylin and eosin (H&E) stained section from the tumor showing that the mass involves the duodenal mucosa, submucosa, muscularis propria, and mesenteric adipose tissue. (B) Higher magnification H&E stained section shows the lesion is composed of variably sized anastomosing vascular spaces containing eosinophilic proteinaceous fluid, confirming the diagnosis of lymphangioma.

Discussion

Lymphangioma is characterized as an uncommon, benign fluid-filled cystic anomaly originating in lymphatic vessels with an excellent prognosis that often appears in the head and neck with high prevalence in children and young adults with equal occurrence between males and females. Intra-abdominal lymphangioma is one of the very rare intra-abdominal tumors that accounts for only 5% of all lymphangiomas. Although the exact etiology is not clear, infection, lymphatic obstruction, and surgery could be risk factors for the development of this anomaly of the lymphatic system [6,7]. They are classified into different subtypes based on

microsporocyte characteristics. Capillary lymphangiomas are made of small, thin-walled lymphatic vessels located in the skin. The cavernous type consists of dilated lymphatic channels of various sizes that keep their connection with normal lymphatic vessels. The cystic type is the most common form of lymphangioma and was consistent with our case. It is composed of large cyst-like cavities filled with straw-colored, protein-rich fluid.

The clinical manifestation of intra-abdominal lymphangioma can vary depending on the location of the tumor. The clinical diagnosis could be difficult due to the lack of specific signs and symptoms, which may include nausea, vomiting, abdominal pain, weight loss, or bowel obstruction [8–10]. Imaging modalities play an important role in the

detection and diagnosis of intra-abdominal lymphangiomas. Ultrasonography is the primary diagnostic modality in showing the size, location, and content of cysts in relation to surrounding tissues. Abdominal contrast-enhanced computed tomography is considered the modality of choice in many studies. Mesenteric lymphangioma lesions may appear as homogeneous cystic masses with wall enhancement, which provides better information about the location and extension of the tumor and degree of vascular involvement. MRI is a more sensitive imaging study in showing intralesional hemorrhage seen as fluid-fluid levels [11–13]. Confirming the diagnosis of mesenteric lymphangioma requires a biopsy revealing thin-walled cystic spaces enclosed with smooth muscle and collagen and positive markers in immunohistology staining like CD31, CD34, and Factor VIII-related antigen. Radical surgical removal of the tumor is the primary treatment since it reduces the rate of recurrence. Surgical removal can be a challenging procedure when the tumor is located in the retroperitoneum or there is extensive vascular involvement and proximity to vital organs [14–16].

Poroos et al. [17] reported a recent case of retroperitoneal lymphangioma. The patient was 17 years old boy with no medical history admitted with right-upper quadrant pain without abdominal distension. An abdominal CT scan showed a retroperitoneal cystic mass infiltrating the mesentery. The diagnosis of cystic lymphangioma was confirmed by pathology and the patient underwent resection surgery without further complications. Chamberlain et al. [18] reported a case of a 57-year-old woman who presented with nausea, abdominal pain, distension, and obstipation. The thickened and dilated small bowel loops with mesenteric edema on the left side of the abdomen were detected in contrast-enhanced CT scans. A laparotomy was performed that revealed chyle throughout small-bowel mesentery with chylous ascites. The diagnosis was confirmed by biopsy of small-bowel mesentery which demonstrated dilated thin- and thick-walled lymphatic vessels, compatible with a cavernous lymphangioma. Nagano et al. [19] described a 40-year-old male with prolonged periumbilical pain. A low-density tumor measuring 45 × 42 mm in the left abdomen located in the peripheral part of the jejunal mesentery was found on an abdominal CT scan. It revealed mesenteric artery involvement and invasion of a mass in the wall of the jejunum. In laparoscopic surgery, the tumor and part of the jejunum were resected. The diagnosis of mesenteric cystic lymphangioma was confirmed by showing positive vascular markers in immunohistochemical staining and cystic thin-wall lesions surrounded by adipose tissue and scattered smooth muscle fibers.

Compared to the available case reports in the literature, our patient was unique in terms of the CT appearance and challenging treatment due to its large size and extensive vascular involvement. Reported cases of retroperitoneal lymphangiomas have typically appeared as a large, thin-walled, unilocular, or multilocular cystic mass on CT [20]. Attenuation of fluid contents is typically homogeneous and of fluid attenuation, but can be of fat attenuation same as that of retroperitoneal fat caused by chyle [20,21]. However, our case was not macrocystic appearance, but predominantly heterogeneous mixed fat and fluid attenuation that caused a lace-like appearance. CT attenuation of such areas ranged from –60 to 0

Hounsfield units depending on the amount of fat component. Therefore, the initial preoperative diagnosis was liposarcoma. It is likely due to the infiltrating nature of this tumor into adipose disuse of the retroperitoneum and mesentery.

Herein, we report the case of a 22-year-old female with no medical history who presented with nausea, vomiting, weight loss, and diffuse abdominal pain. Abdominal CT with contrast as the gold standard imaging study revealed an ill-defined retroperitoneal hypoattenuating mass of mixed fat and fluid attenuation with major vascular encasement. Tumor biopsy confirmed the diagnosis by illustrating numerous dilated lymphatic spaces. Therefore, a combination of radiologic images and pathology evaluation played an essential role in diagnosis. The optimal treatment is radical resection, which minimizes the chance of recurrence [22–24]. Although intra-abdominal lymphangioma is extremely rare, it should be considered among the differential diagnoses of macrocystic or mixed fat and fluid density abdominal mass.

Patient Consent Statement

No specific consent is available for publication. No personal data is presented in the whole text and/or figure, fulfilling anonymity standards.

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