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Case Report

A Huge Retroperitoneal Cystic Lymphangioma Mimicking an Adnexal Cyst

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Abstract

We present a case of retroperitoneal cystic lymphangioma mimicking an adnexal cyst. The clinical presentation and prognosis of the disease, histological features with unique staining patterns, and differential diagnoses are discussed. A 60-year-old woman presented with acid reflux and a bulging left abdominal mass. Abdominal computed tomography revealed an 18-cm well-defined retroperitoneal tumor occupying the left side of her abdomen with descending colon and aorta displacement. The retroperitoneal tumor was resected; its diameter was approximately 18 cm with up to 2000 mL of the chylous fluid. Histopathological analysis, including immunohistochemical staining, confirmed the definitive diagnosis of retroperitoneal cystic lymphangioma.

Keywords: Adnexal cyst, lymphangioma, surgery

Introduction

Lymphangiomas are rare, benign, slow-growing tumors of the lymphatic system. They are usually asymptomatic and found incidentally on imaging studies. Most are found in the neck (75%) and the axillary region (20%), and only 5% are intraabdominal, including the mesentery, gastrointestinal tract, spleen, liver, and very rarely in the retroperitoneum (approximately 1% of all lymphangiomas).[1,2] Complete resection, rather than drainage alone, is recommended to reduce the recurrence rate.[3]

In this report, we describe the case of a 60-year-old woman with acid reflux and a bulging mass over her left abdomen mimicking a primary ovarian tumor, which was later diagnosed as a huge retroperitoneal cystic lymphangioma.

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Case Report

A 60-year-old postmenopausal woman (G3P1A2) presented with acid reflux and a bulging mass over her left abdomen. She had pain and soreness in the left flank and left lower

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abdominal quadrant. At a local clinic, panendoscopy revealed extragastric compression, and abdominal ultrasonography showed a cystic lesion. She was transferred to our hospital for further examination. Tumor markers were in the normal range (CEA: 0.047 IU/mL, CA-125: 8.7 IU/mL, and CA 19–9 <0.8 IU/mL). Transabdominal and transvaginal ultrasonography revealed a 16-cm left pelvic cystic lesion with well-defined borders, which extended to the left abdomen [Figure 1a], and color Doppler imaging revealed no obvious vascularity [Figure 1b]. Contrast-enhanced abdominal computed tomography (CT) showed a huge well-defined retroperitoneal tumor with a diameter of approximately 18 cm occupying the left side of her abdomen with descending colon and aorta displacement. Stretching and narrowing of the left ureter was suspected of causing hydronephrosis [Figure 2]. Therefore, a urologist was consulted for preoperative double-J catheter insertion to identify the left ureter.

During the operation, no obvious abnormal findings were observed in the uterus or bilateral adnexa. A huge left retroperitoneal cyst was noted [Figure 3a], and its chylous fluid content (2000 mL) [Figure 3b] was aspirated. A cytological report of the fluid was negative for malignancy; red blood cell: 22,882 × 10/9 uL; white blood cell: 3321 × 10/9 uL; L: N: 98%:2%; Rivalta's test: negative. A biochemical report showed total protein: 1.8 g/dl; albumin: 1.2 g/dl; lactate dehydrogenase: 186; total-cholesterol: 29; triglycerides: 30, and a culture report also showed no growth in 5 days. Adhesion of the left ureter to the anterior cystic wall was identified, and the retroperitoneal cystic wall was completely resected [Figure 4a]. Postoperatively, no pus formation or active bleeding was recorded. Subsequently, the drainage tube was removed, and she was discharged under a stable condition. No complications were noted during the postoperative follow-up.

The resected specimen consisted of fibromembranous tissue and had a gross volume of 8.0 cm × 1.5 cm × 0.3 cm. It was white and elastic. Histologically, it was a retroperitoneal lymphangioma comprising congested fibroadipose tissue with multiple cystic spaces [Figure 4b]. Immunohistochemical analysis with anti-CD34 antibodies revealed dilated angiolymphatic channels [Figure 4c]. Cytokeratin [Figure 5a] and calretinin [Figure 5b] staining were both negative, which ruled out mesothelioma.[4]

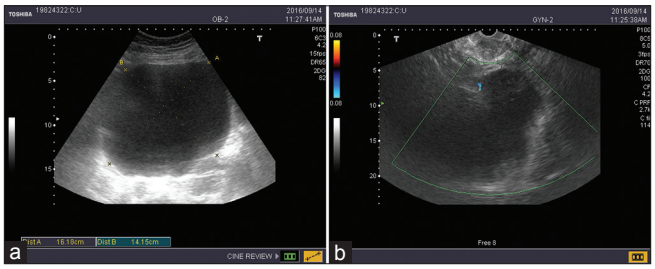

Figure 1: (a) Transabdominal and transvaginal ultrasonography revealed a 16-cm left pelvic cystic lesion with well-defined borders, which extended to the left abdomen. (b) Color Doppler imaging showed no obvious vascularity

Discussion

Since the tumor was slow-growing, our patient presented with acid reflux and left flank pain only when the tumor grew to 16 cm in diameter and produced a mass effect, compressing her stomach and ureter. No specific symptoms of cystic lymphangioma were detected. Thus, cystic lymphangiomas are difficult to diagnosis at an early stage, especially retroperitoneal cystic lymphangiomas.

Retroperitoneal cystic lesions must be differentially diagnosed from numerous diseases, including ovarian tumor growth into the retroperitoneal space, pseudomyxoma retroperitonei with cystic change, perianal mucinous carcinoma with cystic change, pseudocyst, tailgut cyst, retroperitoneal cystic lymphangioma, cisterna chyli, cystic mesothelioma, lymphocele, urinoma from a retroperitoneal bladder rupture, retroperitoneal hematoma with liquefaction, abscess, retroperitoneal hydatid cyst, and retroperitoneal bronchogenic cyst.[5-8] Our patient had no history of trauma, previous retroperitoneal surgery, contact with a wild animal, or history of travel to an area with a high prevalence of echinococcosis, and she presented with no signs of infection. Moreover, contrast-enhanced CT showed the lesion to be located in the retroperitoneal space rather than in the intraperitoneal space, thereby implying a lesion of purely retroperitoneal origin. The bladder was intact during double-J catheter insertion via cystoscopy, and urinoma was ruled out before the incision was made. In addition, the fluid content was chylous, suggesting a lymphatic channel problem.

Lymphangioma is not responsive to radiotherapy or steroids; however, propranolol and sodium tetradecyl sulfate have been shown to be useful in certain patient subpopulations with cystic lymphangioma.[9,10] Sclerotherapy, radiofrequency ablation, percutaneous drainage, or marsupialization can be done, and the cystic lesions might shrink but persist. To avoid recurrence, recent literature recommends complete surgical resection.

Both cystic lymphangiomas and cystic mesotheliomas are rare tumors and are difficult to differentiate because their macroscopic and histological findings are similar.[4] The endothelial cells of lymphangioma are positive for CD31, CD34, and factor VIII-R antigen[11,12] and negative for calretinin, WT-1, and cytokeratins.[5,6] Since a layer of smooth muscle is present in the cystic wall of lymphangiomas, the presence of smooth muscle antigen and positive desmin staining can be used to differentiate them from mesotheliomas. D2–40, mesothelin, thrombomodulin, and HBME-1 may have no benefit in differentiating between the two diseases.[13,14]

Saadi *et al*. reported a case series of five patients with retroperitoneal cystic lymphangiomas, including four female patients and one male patient who underwent surgery between 2004 and 2014. Their medical records were reviewed retrospectively with a mean follow-up of 32.6 months. Follow-up was based on the clinical examinations and abdominal CT scans. The average age was 45 years, and

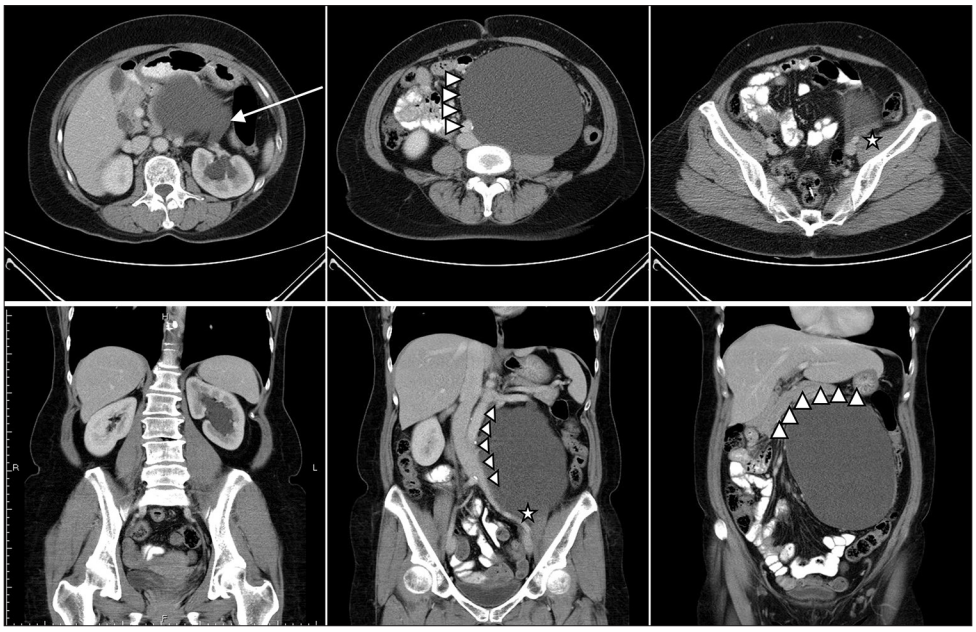

Figure 2: Abdominal computed tomography showed a huge well-defined homogeneous retroperitoneal cyst (arrowheads) in the retroperitoneum (star). Left hydronephrosis was noted with ureteral stricture (arrow)

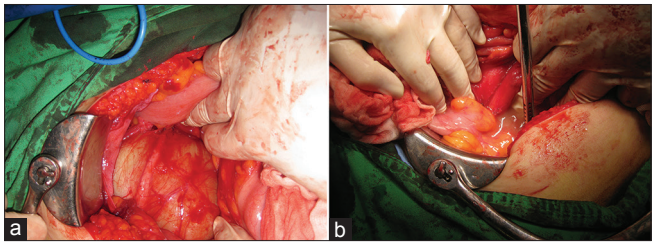

Figure 3: (a) A huge left retroperitoneal cyst was noted. (b) Chylous fluid content was aspirated during laparotomy

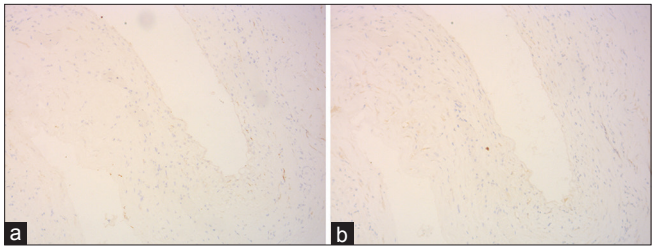

Figure 5: Negative staining for (a) cytokeratin (×100) and (b) calretinin (×100)

the most common symptoms indicative of retroperitoneal cystic lymphangioma were pain and/or an abdominal mass. Abdominal CT was the most useful diagnostic test. Total resection was immediately achieved in four patients and one patient underwent nephrectomy. No recurrence or complications were noted in any of the five patients.[15] Chen and Xia reviewed 11 case reports and concluded that lymphatic channel anomalies were more common in women ($n = 9$) than in men ($n = 2$). The major symptoms were atypical and included abdominal pain, distention, and discomfort, and in general, laboratory data revealed no abnormal findings.[16]

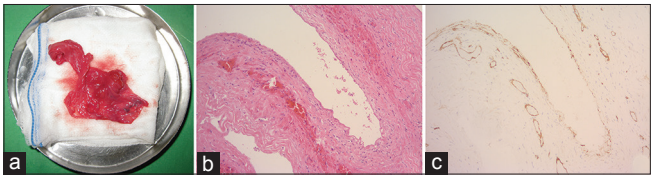

Figure 4: (a) Gross appearance of retroperitoneal lymphangioma. (b) Photomicrograph of the surgical specimen, with fibroadipose tissue seen (H and E, ×25). (c) Immunohistochemistry showing endothelial cells (anti-CD34 antibodies, ×100)

In summary, establishing a definitive diagnosis of retroperitoneal lymphangioma preoperatively using imaging alone can be difficult, especially when the tumor is located near the ovaries. In our patient, the treatment was surgical resection, and the final diagnosis was confirmed through postoperative pathological analysis, including immunohistochemical studies.

Consent

A consent for the publication of this case report and any additional related information was taken from the patient involved in the study.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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