Surg Today (2014) 44:1367–1370
DOI 10.1007/s00595-013-0654-x

CASE REPORT

Differentiating a large abdominal cystic lymphangioma from multicystic mesothelioma: report of a case

Hiroshi Nagata · Yutaka Yonemura · Emel Canbay · Haruaki Ishibashi · Makoto Narita · Makio Mike · Nobuyasu Kano

Received: 9 November 2012 / Accepted: 21 January 2013 / Published online: 27 June 2013

Abstract We report a case of retroperitoneal cystic lymphangioma in a 30-year-old woman who presented with abdominal distention and pain. Imaging studies revealed a large, thin-walled multicystic mass occupying the whole abdomen. Based on a preoperative diagnosis of multicystic mesothelioma, we performed laparotomy, which revealed a tumor arising from the gastropancreatic ligament in the posterior wall of the omental bursa. We resected the tumor completely, without the adjacent viscera. The final pathological diagnosis was cystic lymphangioma, based on the immunohistochemical findings of positive CD31 and CD34 expression, the presence of smooth muscle confirmed by smooth muscle antigen and desmin, and negative calretinin, WT-1 and cytokeratins 5/6 expression. Multicystic mesotheliomas and cystic lymphangiomas are so similar in morphology that immunohistochemical staining should be fully utilized to differentiate them.

Keywords Cystic lymphangioma · Multicystic mesothelioma

H. Nagata (✉) · M. Mike · N. Kano
Department of Surgery, Kameda Medical Center, 929 Higashi-cho, Kamogawa 296-8602, Japan
e-mail: hiroshin84@gmail.com

Y. Yonemura · E. Canbay · H. Ishibashi
Department of Surgery, Kishiwada Tokushukai Hospital, 4-27-1 Kamori-cho, Kishiwada 596-8522, Japan

E. Canbay
Department of General Surgery, Kocaeli-Delince Education and Research Hospital, Kocaeli 41900, Turkey

M. Narita
Department of Pathology, Kameda Medical Center, 929 Higashi-cho, Kamogawa 296-8602, Japan

Introduction

Abdominal cystic lymphangiomas and multicystic mesotheliomas are both rare tumors, which are difficult to differentiate since their macroscopic and histological findings are similar. We report a case of retroperitoneal cystic lymphangioma diagnosed in a young woman and review the literature concerning the differentiation of these two pathologies.

Case report

A 30-year-old woman without any remarkable medical history presented to the Emergency Room at Kameda Medical Center with a 1-week history of dull abdominal pain. She had first noted abdominal distention about 1 year earlier and although her abdomen enlarged gradually, she had not experienced any associated symptoms until just prior to this visit. She reported gaining 4 kg, with a 20-cm increase in waist circumference over the past 9 months. She denied any history of abdominal trauma.

On physical examination, her abdomen was moderately distended with mild tenderness of the epigastrium, but no signs of peritoneal irritation. Laboratory data were within normal limits, except for slight elevation of cancer antigen 125, to 69 U/ml (reference range 0–35 U/ml). Contrast-enhanced abdominopelvic computed tomography (CT) revealed a 32 cm × 18 cm multilocular, ovoid mass reaching up to the pelvis and displacing the stomach and pancreatic head to the left (Fig. 1). The tumor encompassed the major vessels of the upper abdomen, including the celiac trunk, common hepatic artery, superior mesenteric artery, and portal vein. No solid components were noted within the cysts, but the fluid within the mass

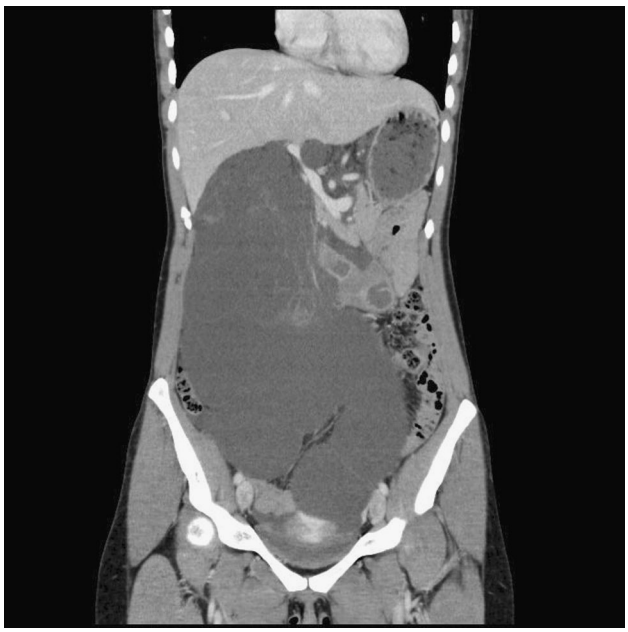

Fig. 1 Computed tomography revealed a large multicystic tumor reaching above the pelvis

demonstrated uniform T1 hypointensity and T2 hyperintensity on magnetic resonance imaging (MRI). Her ovaries were intact bilaterally. Positron emission tomography (PET) demonstrated slight FDG accumulation on the septa (standardized uptake value, 1.83–2.78), but no metastasis.

Based on an assumed clinical diagnosis of multicystic mesothelioma, the patient underwent laparotomy. The peritoneal cavity was fully occupied by the mass, which arose from the gastropancreatic ligament, in the posterior wall of the omental bursa (Fig. 2). There was no evidence of ascites or peritoneal nodules. As intimate attachment to the lesser curvature of the stomach, the pancreas and major vessels were separated easily from the tumor by careful dissection. Complete removal was accomplished without resection of the adjacent organs. After an uneventful recovery, the patient was discharged on postoperative day 6. She suffered mild pancreatitis 2 months after the surgery, but recovered fully within several days.

Cross-section of the tumor revealed multiple cysts filled with serous fluid (Fig. 3). Histological sections showed variably sized cysts lined by a single layer of flattened cells without cytological atypia (Fig. 4). The cells of the inner surface of those cysts were positive for CD31, CD34, D2-40, mesothelin, thrombomodulin, and HBME-1 (faintly); and negative for calretinin, WT-1, and cytokeratins 5/6. Smooth muscle antigen (SMA) and desmin revealed a layer of smooth muscle in the cystic wall (Fig. 5). Based on these findings, we made a final diagnosis of cystic lymphangioma.

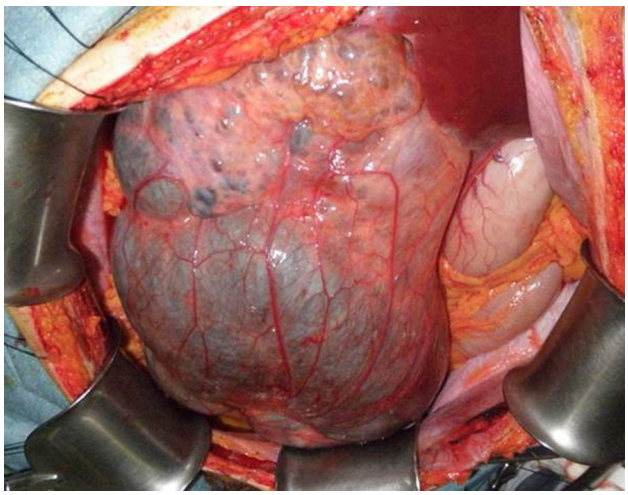

Fig. 2 At laparotomy, the peritoneal cavity was found to be fully occupied by the mass, which arose from the posterior wall of the omental bursa

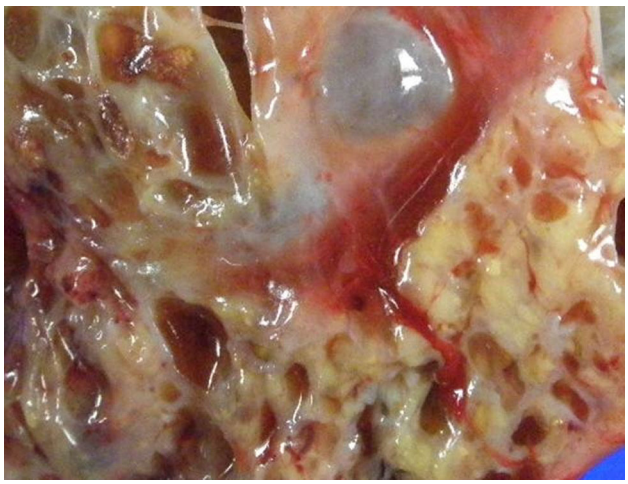

Fig. 3 The tumor was composed of multiple cysts filled with serous fluid

Discussion

Abdominal cystic lymphangiomas are rare benign tumors, with a reported incidence ranging from 1 in 20,000 to 1 in 250,000 [1, 2]. Most lymphangiomas develop in the neck (75 %) and axilla (20 %), with less than 5 % affecting the intra-abdominal contents such as the mesentery, greater omentum, and retroperitoneum [2, 3]. Losanoff et al. [2] defined four different types of mesenteric cystic lymphangiomas, including the pedicled type (type 1), the sessile type (type 2), the retroperitoneal type (type 3), and the multicentric type (type 4). Su et al. [4] reported that most patients with retroperitoneal cystic lymphangiomas were, like our patient, female and younger than those with mesenteric lymphangiomas (median age: 29 vs. 50 years).

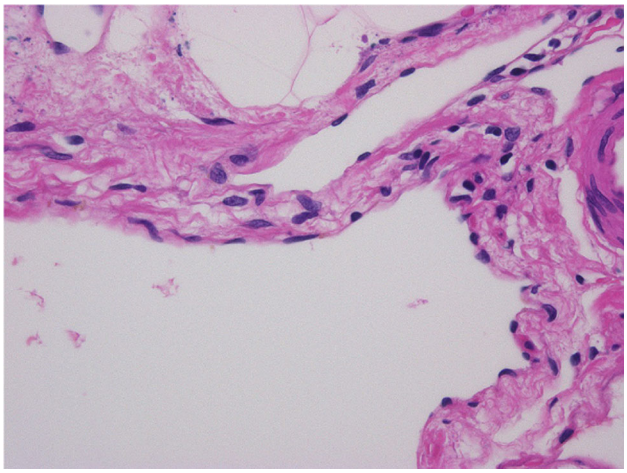

Fig. 4 Hematoxylin and eosin (HE) staining revealed variably sized cysts lined by a single layer of flattened cells without atypia

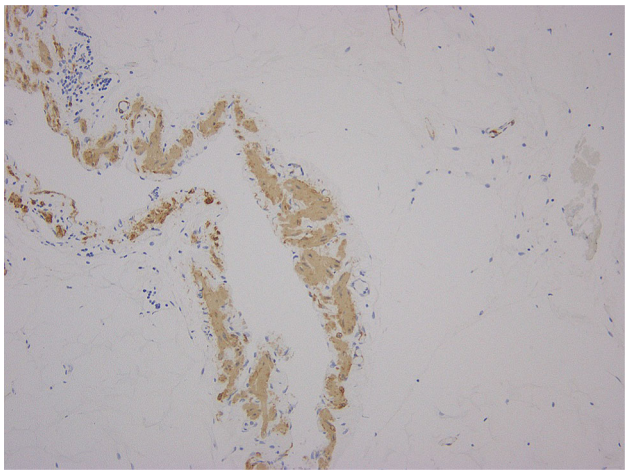

Fig. 5 Immunohistochemical staining with smooth muscle antigen revealed a layer of smooth muscle in the cystic walls

The etiology of cystic lymphangiomas is not fully understood [5]. The most common hypothesis is that lymphangiomas are congenital tumors arising from the sequestration of embryonic lymphatic vessels that have failed to communicate with the rest of the lymphatic or venous systems [6–8]. This theory is supported by the fact that the disease is diagnosed before the patient is 15 years old in over 60 % of cases [9]. The other theory is that the tumor is caused by obstruction of the lymphatic channels by bleeding or inflammation [10].

The clinical course is usually indolent and most patients present with vague abdominal symptoms such as pain, tenderness, nausea, vomiting, and abdominal distension. Acute abdomen in an adult with this disease is seldom reported and is usually of the mesenteric type [4]. Occasionally, the diagnosis is made incidentally at surgery or on imaging studies, which play an important role in the detection, diagnosis, and preoperative evaluation of this tumor. The typical imaging appearance of lymphangioma is that of a large, thin-walled, single or multiloculated cystic mass with contents of water-to-fat attenuation on CT and of high signal intensity on T2 weighted MR images [3]. The anatomical location and organ involvement are indispensable information for preoperative evaluation before complete surgical excision [11] because of the high tendency of local invasion and recurrence. Hancock et al. [12] reported recurrence rates of 12 and 53 % when the tumors were completely or partially resected, respectively.

In our patient, multicystic mesothelioma and cystic lymphangioma were the main differential diagnoses [13]. Multicystic mesothelioma is an unusual, multilocular cystic tumor, which frequently occurs in young to middle-aged women with a history of abdominal or pelvic surgery, trauma, or inflammation. It most commonly arises from the pelvic surfaces of the peritoneum [14]. Unlike pleural mesothelioma, this condition is not asbestos-related. Multicystic mesotheliomas are so similar to cystic lymphangiomas in morphology that it is sometimes difficult to differentiate the two by imaging studies and even pathological examination.

Takiff et al. [1] proposed the following standard histologic criteria for the diagnosis of cystic lymphangiomas: the cyst is lined by flat endothelium rather than cuboidal or columnar epithelium; there are small lymphatic spaces in the cyst wall; lymphoid tissue is abundant in the cyst wall as diffuse collections of lymphocytes or configurations similar to true lymph nodes; foam cells containing lipoid material are present in varying numbers; and smooth muscle is present in the cyst wall. We confirmed the first and last of these features in the present case.

In contrast, a layer of cuboidal or columnar lining cells covers the cysts of multicystic mesothelioma and focal adenomatoid structures or squamous metaplasia or mural proliferation are evident [15, 16]. However, as superimposed reactive and inflammatory changes can obscure the nature of, and the ability to differentiate these tumors, immunohistochemical staining is very important [17, 18]. The endothelial cells of lymphangiomas are reported to be positive for CD31 (88–100 %), CD34 (0–57 %), and Factor VIII-R antigen (82 %) [13, 17]. Although the sensitivity and specificity of immunohistochemical markers have not yet been established in cystic mesothelioma, mesothelial cells are presumed to be positive for calretinin (100 %), WT-1 (71–95 %), cytokeratin 5/6 (53–100 %), D2-40 (93–96 %), mesothelin (100 %), HBME-1 (85 %), and thrombomodulin (73–77 %), based on the findings of peritoneal epithelioid mesothelioma [19, 20]. D2-40, which is considered to be a highly sensitive marker of mesothelioma, cannot be used to distinguish multicystic mesothelioma and cystic lymphangioma because the antibody is selectively expressed in lymphatic endothelium. Many

articles have reported that endothelial cells of lymphangiomas are also positive for D2-40 [17].

Mesothelin is inconclusive in this situation because even though it is commonly and strongly expressed in normal mesothelial cells as well as mesotheliomas, it is positive in many malignancies [21, 22]. For example, the specificity of mesothelin used for differentiating between mesothelioma and ovarian serous carcinoma is only 5 % [19]. Although the literature does not refer to the rate of mesothelin positivity in lymphangiomas, we assume that immunostaining with this antibody has limited value in distinguishing multicystic mesothelioma and cystic lymphangioma.

Initially, we diagnosed multicystic mesothelioma in our patient because her clinical picture was compatible with abdominal mesothelioma; however, after discovering that the CD31 and CD34 were positive, that the presence of smooth muscle was confirmed by SMA and desmin, and that calretinin, WT-1 and cytokeratins 5/6 were all negative, we finally diagnosed cystic lymphangioma.

Conflict of interest We have no potential conflicts of interest.

References

1. Takiff H, Calabria R, Yin L, Stabile BE. Mesenteric cysts and intra-abdominal cystic lymphangiomas. Arch Surg. 1985;120:1266–9.
2. Losanoff JE, Richman BW, El-shrif A, Rider KD, Jones JW. Mesenteric cystic lymphangioma. J Am Coll Surg. 2002;196(4):598–603.
3. Leland HA, Lee JT, Tan JH, Romine LE, Bansal V. Cystic lymphangioma of the lesser curvature of the stomach. J Radiol Case Rep. 2011;5(5):31–7.
4. Su C, Yu M, Chen H, Tseng J, Jan Y, Chen M. Single-centre results of treatment of retroperitoneal and mesenteric cystic lymphangiomas. Dig Surg. 2007;24:181–5.
5. Weeda VB, Booij KAC, Aronson DC. Mesenteric cystic lymphangioma: a congenital and an acquired anomaly? Two cases and a review of the literature. J Pediatr Surg. 2008;43:1206–8.
6. Mayer M, Fartab M, Villiger A, Yurtsever H. Cystic lymphangioma of the transverse mesocolon. Chirurg. 1994;65:561–3.
7. Godart S. Embryological significance of lymphangioma. Arch Dis Child. 1966;41:204–6.
8. Perkins JA, Manning SC, Tempero RM, Cunningham MJ, Edmonds JL Jr, Hoffer FA, Egbert MA. Lymphatic malformations: current cellular and clinical investigations. Otolaryngol Head Neck Surg. 2010;142:789–94.
9. Caro PA, Mahboubi S, Faerber EN. Computer tomography in the diagnosis of lymphangiomas in infants and children. Clin Imaging. 1991;15:242–6.
10. Lee FC. Large retroperitoneal chylous cyst: report of a case, with experiments on lymphatic permeability. Arch Surg. 1942;44:61–71.
11. Bezzi M, Spinelli A, Pierleoni M, Andreoli G. Cystic lymphangioma of the spleen: US-CT-MRI correlation. Eur Radiol. 2001;11:1187–90.
12. Hancock BJ, St-Vil D, Luks FI, Lorenzo MD, Blanchard H. Complications of lymphangiomas in children. J Pediatr Surg. 1992;27:220–6.
13. Chung JH, Suh YL, Park IA, Jang JJ, Chi JG, Kim YI, et al. A pathologic study of abdominal lymphangiomas. J Korean Med Sci. 1999;14:257–62.
14. Pitta X, Andreadis E, Ekonomou A, Papachristodoulou A, Tziouvaras C, Papapaulou L, et al. Benign multicystic peritoneal mesothelioma: a case report. J Med Case Rep. 2010;4:385.
15. Cavallaro A, Berretta M, Lo Menzo E, Cavallaro V, Zanghi A, Di Vita M, et al. Cystic peritoneal mesothelioma: report of a case. Surg Today. 2011;41:141–6.
16. Sawh RN, Malpica A, Deavers MT, Liu J, Silva EG. Benign cystic mesothelioma of the peritoneum: clinicopathologic study of 17 cases and immunohistochemical analysis of estrogen and progesterone receptor status. Hum Pathol. 2003;34:369–74.
17. Hornick JL, Fletcher CDM. Intraabdominal cystic lymphangiomas obscured by marked superimposed reactive changes: clinicopathological analysis of a series. Hum Pathol. 2005;36(4):426–32.
18. Toma GD, Nicolanti V, Plocco M, Cavallaro G, Amato D, Letizia C. Cystic peritoneal mesothelioma: report of a case. Surg Today. 2000;30:98–100.
19. Takeshima Y, Amatya VJ, Kushitani K, Inai K. A useful antibody panel for differential diagnosis between peritoneal mesothelioma and ovarian serous carcinoma in Japanese cases. Am J Clin Pathol. 2008;130:771–9.
20. Ordóñez NG. The diagnostic utility of immunohistochemistry and electron microscopy in distinguishing between peritoneal mesotheliomas and serous carcinomas: a comparative study. Mod Pathol. 2006;19(3):417–28.
21. Ordóñez NG. Application of mesothelin immunostaining in tumor diagnosis. Am J Surg Pathol. 2003;27(11):1418–28.
22. Ordóñez NG. Value of mesothelin immunostaining in the diagnosis of mesothelioma. Mod Pathol. 2003;16(3):192–7.