

Retroperitoneal Lymphangiomyoma in a Patient with Pulmonary Lymphangiomyomatosis: Case Report¹

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Retroperitoneal lymphangiomyoma is rare abdominal finding of lymphangiomyomatosis (LAM). We report here on a case of retroperitoneal lymphangiomyoma and this is the first case that's been seen in the Korean literature. Retroperitoneal lymphangiomyoma associated with pulmonary LAM has rarely been reported in radiologic literature. The characteristic findings of this case are a prominent solid component and retroperitoneal lymphadenopathy mimicking a malignant tumor. A 45-year-old woman was admitted for further evaluation of a lower abdominal mass that could be palpated for 2 weeks. US findings showed relatively well-defined mass with septated cystic portions and echogenic solid portions in the lower abdomen. CT findings revealed a well-demarcated retroperitoneal mass with septated cystic portions and enhancing solid portions at the right lower abdomen, and there were multiple retroperitoneal lymphadenopathy at the lower abdomen. The patient underwent a mass excision. The pathologic findings were retroperitoneal lymphangiomyoma with multiple lymph node involvement.

Index words : Lymphatic system, diseases

Lymphatic system, US

Lymphatic system, CT

Lymphangiomyomatosis

Lymphangiomyoma is a rare abdominopelvic manifestation of lymphangiomyomatosis (LAM) (1). LAM is characterized by a proliferation of abnormal smooth muscle cells in the lungs and in the lymphatic system of the thorax and retroperitoneum, and it is almost exclusively seen in the premenopausal women (1 - 4). The retroperitoneal lymphangiomyoma showed a thick, echogenic rind that surround a central hypoechoic cystic area on ultrasonography (US), and a thin or thick walled cystic mass that contains material that is low in

attenuation (3-25HU) on computed tomography (CT). To our knowledge, retroperitoneal lymphangiomyoma associated with pulmonary LAM has rarely been reported in the radiologic literature. This is the first case that's been seen in the Korean literature. The characteristic findings of this case are a prominent solid component and retroperitoneal lymphadenopathy mimicking malignant tumor. We report here on a case of retroperitoneal lymphangiomyoma associated with pulmonary LAM and describe the characteristic US and CT findings.

Case Report

A 45-year-old woman was admitted to our hospital via a local OB/GY clinic for further evaluation of soft, palpable lower abdominal mass that had been observed for 2

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weeks. The neurological examination and family history were nonspecific.

US showed a well-defined mass about 8.5 cm in size, with septated cystic portions and echogenic solid portions in the longitudinal scan of the right lower abdomen (Fig. 1A). A follow-up US the next morning shows the change of a mild decrease of the mass at the low abdomen. CT scans revealed a well-demarcated retroperitoneal mass that contained septated cystic portions and well enhancing solid portions in front of the

right psoas muscle and the great vessels at the right lower abdomen (Fig. 1B), and there was multiple retroperitoneal lymphadenopathy around great vessels at the lower abdomen. HRCT images showed variable sized innumerable cystic lesions in the both lung without mediastinal lymphadenopathy and dilated thoracic ducts (Fig. 1C). The patient underwent a mass excision. A photograph of a resected specimen revealed a large ovoid soft tissue mass attached with mesentery, measuring about $8 \times 7 \times 3$ cm. The mass was lobulated and alter-

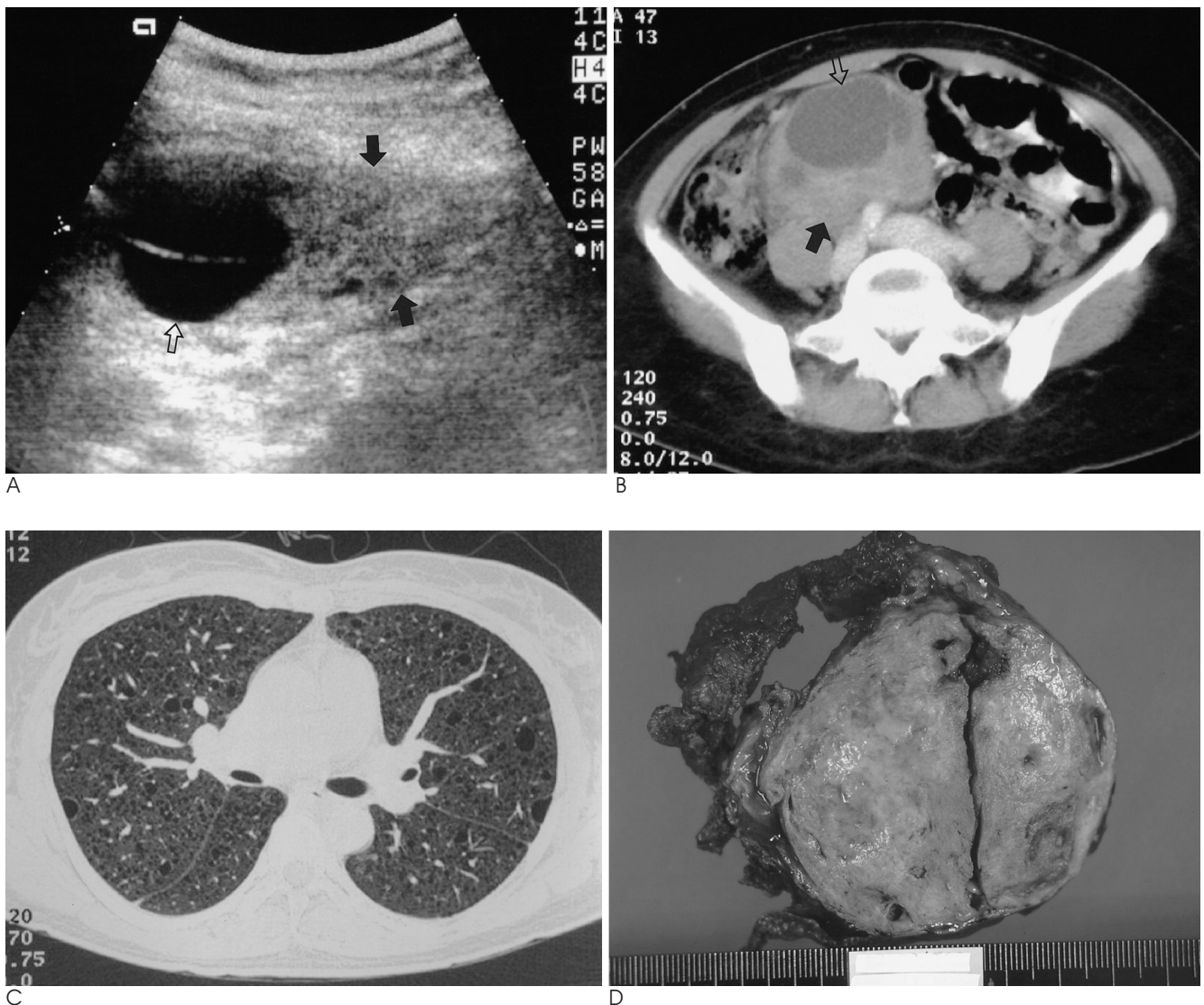


Fig. 1. A. Ultrasonography shows about an 8.5 cm sized, well-defined mass with a septated cystic portion (open arrow) and an echogenic solid portions (solid arrows) in the longitudinal scan of the right lower abdomen.

B. CT reveals a well demarcated retroperitoneal mass that contains septated cystic (open arrow) and also well enhancing solid portions (solid arrow) in front of the right psoas muscle and the great vessels at the right lower abdomen. Multiple retroperitoneal lymphadenopathies at the middle and lower abdomen are noted. (Not demonstrated).

C. HRCT shows variable sized, innumerable cystic lesions in both lungs without mediastinal lymphadenopathy and dilated thoracic ducts.

D. Photograph of a resected specimen reveals a large ovoid soft tissue mass attached with mesentery measuring about $8 \times 7 \times 3$ cm. The mass is lobulated and alternated by solid and cystic portions.

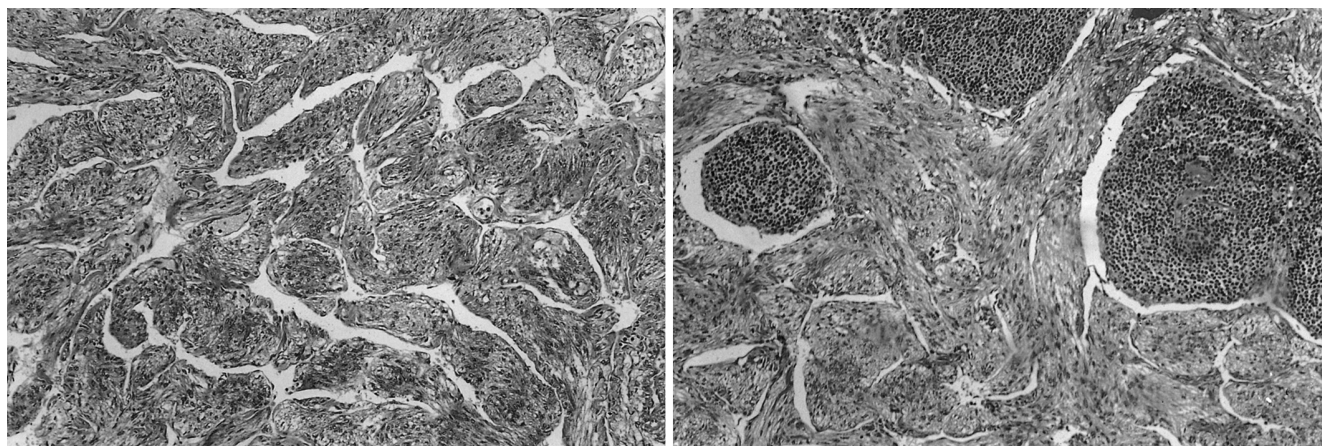


Fig. 1. E. Microscopic examination shows the proliferation of abnormal smooth muscle cells that are arranged in short fascicles around a network of endothelium-lined spaces (lymphatic ducts) without pleomorphism and mitotic figures. ($\times 100$, H & E stain). This appearance is diagnostic of lymphangiomyoma.

F. Microscopic examination shows the involvement of retroperitoneal lymph nodes. ($\times 100$, H & E stain).

nated by solid and cystic portions (Fig. 1D). The microscopic examination showed a proliferation of abnormal smooth muscle cells that were arranged in short fascicles around a network of endothelium-lined spaces without pleomorphism and mitotic figures, and this mass also showed an involvement of the retroperitoneal lymph nodes (Fig. 1E, F).

Discussion

Lymphangiomyoma, a rare finding of abdominal manifestation of LAM, results from the proliferation of smooth muscle cells in the lymphatic vessels, which causes a dilatation and obstruction in the lymph vessels and results in cystic collections of chylous materials (6 - 8). Lymphangiomyoma may lie between and displace vascular structures in the retroperitoneum and, along with abdominal lymphadenopathy, may be misdiagnosed as a neoplastic process such as lymphoma. On CT, the dilated retroperitoneal lymph vessels may have either thin or thick walls (cystic mass) and they may contain a low attenuation material. On US, the multi-septated cystic mass with an echogenic rind that surrounds a central hypoechoic area was shown (1). In this case, the lymphangiomyoma had a thin-walled, septated cystic component and it also had a well-enhanced prominent solid component which was composed of a prominent proliferation of abnormal smooth muscle cells. The latter is a characteristic finding mimicking malignant tumor and this was different from previous

reported findings of lymphangiomyoma.

The differential diagnosis of cystic retroperitoneal and pelvic masses includes various entities. Cystic lymphangiomas are similar to lymphangioleiomyomas in LAM. Lymphangiomas consist of endothelial-lined lymphatic spaces without the muscle or LAM cells in their walls. When a retroperitoneal lymphangioleiomyoma has thick, irregular walls or does not contain fluid, it may be confused with other retroperitoneal neoplasms such as lymphoma or sarcoma. Pelvic cystic lymphangioleiomyomas may be misdiagnosed as cystic pelvic masses (hydrosalpinx or pyosalpinx) or as ovarian cancer. A carefully obtained medical history and the diurnal variation of mass size may help to differentiate lymphangioleiomyomas from other causes (12).

The causes of diurnal variation in size of lymphangioleiomyomas are the increase in chyle production after meals, an increase in lymphatic return due to activity, and the gravity effects of intraluminal pressure. Their characteristic diurnal variation in size upon CT assists in the diagnosis and prevents unnecessary biopsy.

LAM is a rare idiopathic disorder found in premenopausal women and is characterized by a proliferation of abnormal smooth muscle cells in the lungs and in the lymphatic system of the thorax and retroperitoneum (2 - 4). About 76% of LAM had positive abdominal findings. Common abdominal findings included renal angiomyolipoma in 54% of LAM, enlarged abdominal lymph nodes in 39% and lymphangiomyoma in 16%. Less common findings included chylous ascites, dilata-

tion of thoracic duct and hepatic angiomyolipoma (1). In this case, the abdominal findings were a retroperitoneal lymphangiomyoma and multiple enlarged lymph nodes.

Renal angiomyolipoma is the most common tumor associated with LAM (4). Non-enhanced CT is essential to visualize the fat content of angiomyolipoma. Moreover, helical CT is more sensitive than conventional CT in demonstrating the fat in angiomyolipoma that are less than 2 cm in diameter (5). US may not be sufficient to depict the angiomyolipoma if it is isoechoic to normal renal parenchyma, and if it does not deform the renal contour, or if it is adjacent to normal renal sinus fat (1).

Abdominal lymphadenopathy may be extensive and it may contain focal low attenuation areas, which indicates chylous lymph collection or fat within the lymph nodes. The previous report showed a positive trend between the severity of lung disease and the presence of abdominal lymphadenopathy. Lung and abdominal involvement are related to disorders of the lymphatic system. Patients who have undergone a biopsy demonstrated the replacement of the lymph nodes with smooth muscle.

LAM can occur without other disease or in association with tuberous sclerosis (9). Tuberous sclerosis complex is an autosomal dominant genetic disorder with pulmonary, renal and lymph node findings similar to those of LAM (10). Although angiomyolipoma occurs in patients of either sex with tuberous sclerosis complex, cystic lung disease is found only in woman (11).

We report here on a rare retroperitoneal lymphangiomyoma with enhancing prominent solid components associated with LAM. When abdominopelvic US or CT shows cystic and solid masses with extensive lymphadenopathy mimicking malignant masses, evaluation of lung and kidney is thought to be useful to diagnose

the retroperitoneal lymphangiomyoma.

Abbreviations

LAM = lymphangiomyomatosis

Lymphangiomyoma = lymphangioleiomyoma

References

1. Avila NA, Kelly JA, Chu SC, Dwyer AJ, Moss J. Lymphangioleiomyomatosis: abdominopelvic CT and US findings. *Radiology* 2000;216:147-153
2. Kitaichi M, Nishimura K, Itoh H, Izumi T. Pulmonary lymphangioleiomyomatosis: a report of 46 patients including a clinicopathologic study of prognostic factors. *Am J Respir Crit Care Med* 1995;151:527-533
3. Taylor JR, Ryu J, Colby T, Raffin TA. Lymphangioleiomyomatosis: clinical course in 32 patients. *N Engl J Med* 1990;323:1254-1260
4. Maziak DE, Kesten S, Rappaport DC, Maurer J. Extrathoracic angiomyolipomas in lymphangioleiomyomatosis. *Eur Respir J* 1996;9:402-405
5. Silverman SG, Pearson GD, Seltzer SE, et al. Small (< or = 3 cm) hyperechoic renal masses: comparison of helical and conventional CT for diagnosing angiomyolipoma. *AJR Am J Roentgenol* 1996;167:877-881
6. Carrington CB, Cugell DW, Gaensler EA, et al. Lymphangioleiomyomatosis. Physiologic-pathologic-radiologic correlations. *Am Rev Respir Dis* 1977;116:977-995
7. Wolff M. Lymphangiomyoma: clinicopathologic study and ultrastructural confirmation of its histogenesis. *Cancer* 1973;31:988-1007
8. Joliat G, Stadler H, Kapani Y. Lymphangiomyomatosis: a clinico-anatomical entity. *Cancer* 1973;31:455-461
9. Smolarek TA, Wessner LL, McCormack FX, et al. Evidence that lymphangiomyomatosis is caused by TSC2 mutations: chromosome 16p13 loss of heterozygosity in angiomyolipomas and lymph nodes from women with lymphangiomyomatosis. *Am J Hum Genet* 1998;62:810-815
10. Jao J, Gilbert S, Messer R. Lymphangiomyoma and tuberous sclerosis. *Cancer* 1972;29:1188-1192
11. Reed WB, Nickel WR, Campion G. Internal manifestations of tuberous sclerosis. *Arch Dermatol* 1963;87:715-728
12. Avila NA, Bechtel J, Dwyer AJ, Moss J. Lymphangioleiomyomatosis: CT of diurnal variation of lymphangioleiomyomas. *Radiology* 2001;221:415-421

