The lymphangiomyomatosis is a rare disease of unknown etiology, almost affecting childbearing women. The typical radiologic manifestations of this disease are pneumothorax, chylothorax and multiple thin-walled cysts throughout the bilateral lung parenchyma, which have been well-documented in the literature. The lymphangiography is a conventional image modality, rarely performed nowadays due to the rapid development of cross-sectional imaging modalities such as computed tomography. Herein we will report a case of lymphangiomyomatosis with the different image modality, lymphangiography.

CASE REPORT

A 45 year-old female suffered from shortness of breath for two weeks. Plain chest X-ray showed massive right pleural effusion revealed to be chylothorax by thoracocentesis. Lymphangiography was requested by the chest surgeon with the suspicion of thoracic duct leakage. The plain films of pelvis five minutes after lipiodol injection into the lymphatic duct of feet were performed and revealed abnormal lipiodol retention along bilateral inguinal and iliac chains (Fig. 1a, 1b). No passage of the lipiodol into the thoracic duct was identified even three days later (Fig. 1c). The follow up abdominal CT revealed multiple cystic lesions along para-aortic, bilateral common, external and internal iliac vessels. There was still abnormal lipiodol retention within the cystic lesions over the para-aortic, bilateral inguinal and iliac regions (Fig. 2a, 2b). These findings suggested these cystic lesions to be the lymphatic structures.

Due to the persistent chylothorax, pleurodesis and thoracic duct ligation were performed. Thoracoscopic wedge resection of right lower lobe of lung was done. The histologic study disclosed variably-sized microcystic spaces and anastomosing vascular channels lined by flattened endothelia surrounded by smooth muscle-like spindle cell, consistent with pulmonary lymphangiomyomatosis (Fig. 3).
DISCUSSION

Lymphangiomyomatosis is a rare disease with unknown etiology, exclusively affecting the childbearing women. It is characterized by the proliferation of abnormal smooth muscle cells (LAM cells) in the pulmonary interstitium and along thoracic and abdominal lymphatics [4, 5]. The radiologic presentations of lymphangiomyomatosis include pneumothorax, pleural effusion with or without chylothorax, and diffuse thin-walled cysts throughout the pulmonary parenchyma, which is the diagnostic clue to this disorder [6]. However, it may appear as retroperitoneal cystic mass, which is found in more than 50% of patients [1]. Sometimes it may mimic metastatic lymphadenopathy.

Figure 1. a. The lymphangiographic film showed lipiodol retention in the lymph nodes and lymphatic ducts over the bilateral iliac chains. b. magnified spot view: multiple abnormally enlarged lymph nodes and the dilated lymphatic ducts were filled with lipiodol. c. Pelvic film performed three days after lymphangiography revealed lipiodol still retention in pelvic lymphatic structures without significant change as compared with images three days ago. The cisterna chili and the thoracic duct was not opacified.

Figure 2. a. Follow up CT scan after lymphangiography showed lipiodol retention within abnormal cystic lesions around bilateral iliac vessels. b. Only small amount of lipiodol in the cystic lesion around para-aortic region. These findings suggested these cystic lesions to be lymphatic structures.
retroperitoneal abscess, tuberculosis, neurofibromatosis or other retroperitoneal tumor [7]. In the reported literature, the lymphangiography of lymphangiomyomatosis showed dilatation of lymphatic channels and slow progression and blockage of the lipiodol within it, similar to our case [8].

Conventional lymphangiography is the most useful method for evaluating lymphatic system. It could demonstrate the internal architecture of the lymph node, which is more accurate than CT [9]. When a patient presents with retroperitoneal cystic mass without clear clinical history or typical manifestation, lymphangiography can be performed to evaluate the etiology, whether it is originated from lymphatic channels or a true neoplasm. In the special situation such as chylothorax, the lymphangiography can demonstrate not only the etiology but also better anatomic demonstration of lymphatic channels for further management [10]. However, the number of lymphangiographic examination has been declined due to the introduction of the cross sectional imaging modalities such as CT and the experiences in both performing and interpreting the lymphangiography are decreasing. This time, we report this disease with a different image modality, lymphangiography, to remind the young radiologists of its manifestation and application.

**REFERENCES**

10. Freundlich IM. The role of lymphangiography in chylothorax: a report of six nontraumatic cases. AJR Am J Roentgenol 1975; 125: 617-627
淋巴管肌瘤增生症的淋巴攝影表現：病例報告

許文遠  曾文盛  吳仁宏

奇美醫學中心永康院區  影像醫學部

淋巴管肌瘤增生症是一個罕見的疾病，幾乎都只影響生育年齡女性。最常見的放射學表現是氣胸，乳糜胸和許多薄壁的囊腫在兩側肺野，這些已經是很常見的表現。淋巴攝影是一個傳統的檢查，隨著電腦斷層的快速發展，這項檢查越來越少被實施。這次我們將介紹一個淋巴管肌瘤增生症的病例並且以不同的影像診斷工具呈現，使用淋巴攝影來表現。