

## MULTI-MODAL IMAGING FEATURES AND LYMPHO-VENOUS SHUNT FOR VAGINAL CHYLOUS FISTULA IN LYMPHANGIOLEIOMYOMATOSIS: CASE REPORT AND REVIEW

M. Huo, Z.-C. Wang, Y.-L. Yue, J. Dong, Z. Wen, J.-F. Xin

Department of Radiology (MH,Z-CW), Beijing Friendship Hospital, Capital Medical University; Departments of MR (Y-LY), Radiology (JD), Radiology-Nuclear Medicine (ZW), and Lymphatic Surgery (J-FX), Beijing Shijitan Hospital, Capital Medical University, Beijing, China

### ABSTRACT

*Vaginal chylous fistula is a very rare lymphatic disorder and lymphangioleiomyomatosis (LAM) is also a rare multisystemic disease. LAM with the development of a vaginal chylous fistula has rarely been reported. Here we report a case of vaginal chylous fistula accompanied with LAM. A 61-year-old woman occasionally experiencing milky vaginal discharge for the last 3 months underwent CT lymphangiography (CTL) after direct lymphangiography (DLG) and lymphoscintigraphy. DLG showed lymph flowing backwards toward the pelvic cavity. CTL revealed abnormal distribution of contrast agent in the pelvic cavity, including uterus, vagina, perineum, bladder, and muscles in the bilateral pelvis. Multiple small thin-walled cystic lesions were present in both lungs. Lymphoscintigraphy demonstrated lymphangiectasia in the pelvic cavity bilaterally with radionuclide tracer flowing back abnormally into the pelvic walls. The vaginal exudate was positive for chyle. According to LAM guidelines, the patient was diagnosed with LAM. This case presents multimodal images of a rare case of vaginal chylous fistula accompanied with LAM and highlights the value of CT lymphangiography after DLG in establishing the diagnosis and providing important guidance for sequential therapy.*

**Keywords:** vaginal chylous fistula, chylous reflux, lymphangioleiomyomatosis (LAM), direct lymphangiography (DLG), CT lymphangiography (CTL), lymphoscintigraphy

Vaginal chylous fistula is a very rare lymphatic disorder and lymphangioleiomyomatosis (LAM) is a rare multisystemic disease that mainly affects females of childbearing age. The pathological diagnosis is based on the abnormal proliferation of LAM cells, an immature smooth muscle-like cell, in the lymphatic system, bronchi, and blood vessels (1,2). Patients with LAM usually demonstrate progressive dyspnea, pneumothorax, chylothorax, and chylous ascites. However, LAM with the development of a vaginal chylous fistula has rarely been reported, and there are no reports of the imaging findings using CT lymphangiography (CTL) after direct lymphangiography (DLG) and lymphoscintigraphy.

### CASE REPORT

A 61-year-old woman reported that approximately 50 to 100 ml of milky white liquid had discharged from her vagina five times in the previous 3 months. Each episode of discharge stopped spontaneously. The patient had a history of chest pain, shortness of breath, and dyspnea after exercise without



*Fig. 1. CTL after DLG. (1a) Axial section through the perineum demonstrates abnormal distribution of the contrast agent in the perineum (arrow). (1b) Axial section through the pelvis demonstrates abnormal distribution of the contrast agent in the vagina (arrow), bladder, and the muscles of the pelvis. (1c) Axial CT scans of the chest demonstrate multiple thin-walled cystic lesions were present in the lungs bilaterally.*

a known cause beginning 12 years previously. Examination revealed right pleural effusion with bloody chyle but no tuberculosis or tumor was found. The patient underwent drainage of the pleural cavity two to three times weekly (approximately one liter per time) but her symptoms were not alleviated. Therefore, the patient underwent pleurodesis for chylothorax; however, hydrothorax was still present at the follow-up examination, although it had decreased in volume. The patient then developed intermittent abdominal distension and pain of unknown cause for 10 years with symptoms becoming aggravated in the most recent 3 years. However, these symptoms, including the abdominal distension and chest pain, were alleviated after the development of the vaginal discharge in the previous 3 months.

#### *Examination of vaginal exudate*

The vaginal exudate was positive for chyle. Microscopic examination revealed fat globules stained by Sudan III.

#### *Radiological examinations and manifestations*

##### 1. Direct lymphangiography (DLG)

1.1. Method: A mixture of 1 ml of methylene blue and 3 ml of 2% lidocaine was subcutaneously injected between the first and third toes of the left foot. The skin of the

first metacarpal space along the dorsal aspect of the left foot was cut and blue-dyed lymph vessels were identified. After puncture, 20 ml lipiodol (Guerbet, Laboratories, Aulnay-sous-Bois, France) was injected into a lymph vessel at a speed of 6 to 8 ml/h, and the patient was observed under digital subtraction angiography.

1.2. Imaging manifestations: The number of lymph vessels in the left inguinal area and left pelvic cavity was increased. The lymph vessels were circuitous and extremely dilated. The lymph flowed back toward the contralateral inguinal area and pelvic cavity, and it was difficult for the contrast agent to advance upstream. No contrast agent was present above the second lumbar vertebra.

##### 2. CT lymphangiography (CTL) after DLG

2.1. Method: Immediately after DLG, a CT scan ranging from the pelvic cavity to the neck was performed using a Somatom Sensation 16 (Siemens, Munich, Germany). The abdomen and pelvis were reconstructed at a 5-mm section thickness, and the thorax was reconstructed at a 2-mm section thickness.

2.2. Imaging findings: CTL showed abnormal distribution of the contrast agent in the pelvic cavity, including the uterus, vagina, perineum, bladder, and muscles in the bilateral pelvis (Figs. 1a,b). Dilated, circuitous, and dysplastic lymph vessels were found in the bilateral inguinal region, pelvic



Fig. 2.  $^{99m}\text{Tc}$ -dextran lymphoscintigraphy. Lymphangiectasia was present in the pelvic cavity bilaterally, and the radionuclide tracer flowed back into the pelvic walls (arrow) abnormally.

cavity, and retroperitoneum. However, no contrast media was present above the level of the second lumbar vertebra. Multiple thin-walled cystic lesions were also present in both lungs (Fig. 1c), and a small amount of effusion was present in the right pleural cavity.

### 3. $^{99m}\text{Tc}$ -dextran lymphoscintigraphy

3.1. Method:  $^{99m}\text{Tc}$ -dextran lymphoscintigraphy was performed using a SPECT gamma camera (Forte; Philips, Amsterdam, Netherlands).  $^{99m}\text{Tc}$ -labeled radionuclides (111-185 MBq, equivalent to 0.10-0.15 ml) were subcutaneously injected into both feet at two points: between the first and second toes and between the fourth and fifth toes. Images were obtained at 10 min, 1 hour, 3 hours, and 6 hours after injection, respectively.

3.2. Imaging findings: Lymphangiectasia was present in the pelvic cavity, and the bilaterally radionuclide tracer flowed back abnormally into the pelvic walls (Fig. 2). The tracer moved cephalad slowly. Only a

small amount of tracer was detected above the level of the second lumbar vertebra indicating obstruction of the initial segment of the thoracic duct.

According to the LAM guideline published by the European Respiratory Society in 2010 (1), the patient was diagnosed with LAM because of the typical thoracic CT manifestations and chylothorax.

3.3 Surgery: The patient underwent anastomosis of the left external iliac vein and lymph vessel. During surgery, many dilated, circuitous, and structurally disordered lymph vessels were found in the pelvis bilaterally. One dilated lymph vessel was isolated and an end-to-end anastomosis between the proximal part of the lymph vessel and left external iliac vein was performed. No vaginal chyloous fistula was observed for 2 years postoperatively.

## DISCUSSION

LAM is a rare multisystemic disease, which can occur sporadically or in association with tuberous sclerosis complex (TSC) and is characterized by proliferation of LAM cells in the lymphatic system, bronchi, and blood vessels (1,2). Proliferation of LAM cells in bronchi can lead to pervasive destruction of terminal air cavities, which results in the formation of typical lung cystic lesions; this finding is the main cause of progressive respiratory failure and pneumothorax in such patients (3). Approximately 76% of patients with LAM have positive abdominal CT findings (4), including renal or hepatic angiomyolipoma (AML), enlarged abdominal lymph nodes, retroperitoneal and pelvic lymphangiomyoma (LALM), and ascites (1,2,4). All of these except AML are related to proliferation of the LAM cells in the lymphatic system, which can lead to obstruction of lymphatic vessels (1,4).

Vaginal chyloous fistula is very rare in the clinical setting, and there are no reports of image findings association with LAM (5-7).

Chyle is collected in the lymph vessels of the small intestine and drains to the cisterna chyli through the mesenteric lymph duct to eventually enter the thoracic duct. Chyle is uniquely distributed in these areas physiologically, and it is transported in the lymph into the venous angle through the thoracic duct. Obstruction of the lymphatic system due to various causes can lead to increased pressure in the lymphatic circulation with subsequent chyle backflow, dilation of the lymphatic vessels, and establishment of lymphatic collateral circulation. If this high-pressure state cannot be alleviated over time, it results in leakage or exudation of chyle from the thin lymphatic vessels. Depending on the body region in which this occurs, clinical manifestations include chylothorax, chylopericardium, chylous ascites, chylous sputum, chylous feces, chyluria, vaginal chylous fistula, and lymphedema of the extremities (6,8).

The patient described herein had a 12-year disease history. Based on her DLG, we hypothesized that LAM cells led to obstruction of the thoracic duct which further induced dilation of lymphatic vessels and establishment of collateral circulation in the pelvic cavity with eventual chylous ascites. There are diffusive lymphatic drainage patterns in the uterus and vagina which are connected to adjacent blood vessels and ligaments as well as the bladder and rectum. In addition to effects of gravity, there is a higher pressure in the lymphatic circulation in this area, and the vaginal chylous fistula resulted from rupture of the dilated lymphatic vessels in the uterus and vagina. This abnormality led to identification of a diffused, abnormal distribution of the contrast agent in vagina, uterus, perineum, bladder, and the muscles of the pelvis in CTL. This patient underwent anastomosis of the external iliac vein and lymph vessel to reduce the pressure of the pelvic lymphatic system, which effectively alleviated her symptoms during the two-year follow up. This type of shunt surgery for chyle backflow

disease induced by LAM is worthy of further investigation.

Although we did not directly observe the fistula by DLG in our patient, we observed abnormal aggregation of the contrast agent in the uterus, vagina, and perineum by CTL. DLG and lymphoscintigraphy are dynamic imaging modalities that can be used to obtain a full view of the lymphatic circulation. CTL after DLG is better than DLG and lymphoscintigraphy in terms of spatial resolution, and they can therefore be effectively used to locate chylous fistulae (9). Particularly in patients with LAM, they can be used to simultaneously observe severity of lung involvement, pneumothorax, pleural effusion, abdominopelvic ascites, LALM, hepatic and renal AMLs, and sclerotic bone lesions (SBLs) (10). Obtaining such information is useful to establish diagnosis, examine differential diagnoses, and provide important guidance for sequential therapy. Additionally, the LAM guideline published by the European Respiratory Society suggests that abdominal and pelvic CT should be performed in all patients with LAM (1). Finally, some studies have shown that the use of lipiodol in DLG has a therapeutic effect on chylous fistulae (9,11,12), and this effect may strengthen the recommendation to use DLG followed by CTL from the pelvis to thorax in patients with LAM who develop chylous fistulae.

#### *ETHICAL APPROVAL*

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Informed consent was obtained from this individual participant included in the study.

#### *CONFLICT OF INTEREST AND DISCLOSURE*

All authors declare that no competing financial interests exist.

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**Dr. Zhen-Chang Wang**  
**Department of Radiology**  
**Beijing Friendship Hospital, Capital**  
**Medical University**  
**No. 95, Yong'an Road, Xicheng District**  
**Beijing 100050, China**  
**Tel: +86-10-6318625**  
**Fax: +86-10-63023261**  
**E-mail: cjr.wzhch@vip.163.com**