

CASE REPORT

Lymphangioliomyomatosis: Computed Tomography-guided Tru-cut Biopsy of a Retroperitoneal Mass

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ABSTRACT

A patient with lymphangioliomyomatosis with an unusual presentation mimicking ovarian cancer is described. Computed tomography-guided tru-cut biopsy of the retroperitoneal lymph node established the histological diagnosis. Concomitant computed tomography-guided fine needle aspiration only revealed a large number of small-sized lymphocytes. Tru-cut biopsy of the retroperitoneal lymph node is an alternative to open lung biopsy for patients with lymphangioliomyomatosis with pulmonary disease and retroperitoneal lymphadenopathy.

Key Words: Adnexal and skin appendage neoplasms, Biopsy, Lymphangioliomyomatosis, Retroperitoneal neoplasms

INTRODUCTION

Lymphangioliomyomatosis (LAM) is a rare disorder with the lung as the primary target of the disease pathology. LAM is traditionally diagnosed by open lung biopsy or classical high resolution CT (HRCT) findings with compatible respiratory symptoms. A patient with LAM with an atypical presentation that mimicked ovarian carcinoma with lymph node metastases is described. CT-guided tru-cut biopsy of the retroperitoneal lymph node established the diagnosis without requiring lung biopsy.

CASE REPORT

A 35-year-old Chinese woman presented with a 4-week history of left thigh swelling. She was a non-smoker and did not have any chest symptoms. Ultrasound examination of the pelvis by a private practitioner showed a large, complex, cystic left adnexal mass. She was referred for further treatment.

A repeat ultrasound examination using an Acuson-128XP10 machine (Acuson Corporation, Mountain View, USA) confirmed the presence of a complex left adnexal mass lesion containing both cystic and solid

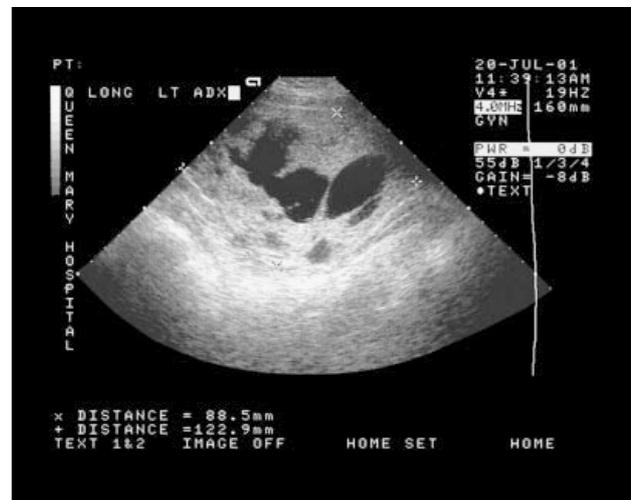


Figure 1. Ultrasonogram of the pelvis showing the complex left adnexal mass with mixed cystic and solid components.

components (Figure 1). Duplex ultrasound examination of the deep veins of the left lower limb did not show venous thrombosis. Contrast-enhanced CT examination of the abdomen and pelvis was performed using a General Electric Lightspeed CT scanner (General Electric Company, Milwaukee, USA). There was an 8 x 11 x 14 cm left adnexal mass containing both solid and cystic components, with the solid components showing heterogeneous enhancement (Figure 2).

Bilateral iliac and retroperitoneal lymph nodes — many containing internal hypodense centres with attenuation numbers ranging from -10 to +20 Hounsfield units

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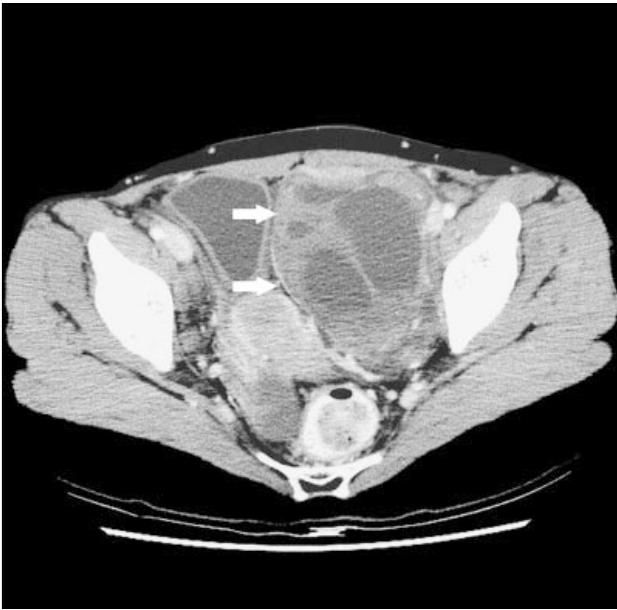


Figure 2. The soft tissue components of the left adnexal mass (arrows) showed heterogenous enhancement on contrast-enhanced computed tomography.

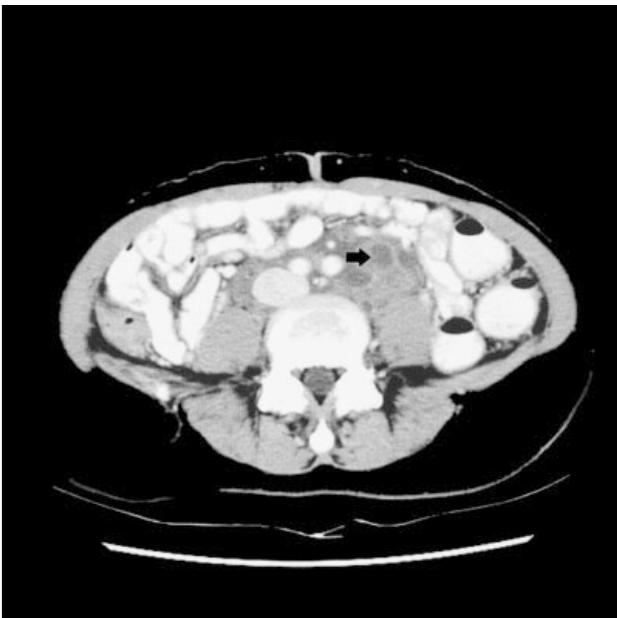


Figure 3. Contrast-enhanced computed tomography showing the enlarged retroperitoneal lymph nodes, many harbouring low attenuation centres (arrow).

(Figure 3) — were observed extending up to the level of the left renal hilum.

The presumptive diagnosis at this juncture was ovarian carcinoma with lymph node metastases or lymphoma. Therefore, CT-guided fine needle aspiration of the retroperitoneal lymph nodes was performed using the posterior approach with the patient in the prone position. A single pass with a 22-gauge Inrad needle (Inrad Inc,



Figure 4. Computed tomography-guided tru-cut biopsy using a posterior approach with the patient in the prone position.

Kentwood, USA) was made. The aspirate showed a large number of small-sized lymphocytes and occasional small-sized lymphoid cells only. CT-guided tru-cut biopsy of the retroperitoneal lymph node was performed in the same setting using a 20-gauge Temno biopsy needle (Allegiance Healthcare Corporation McGaw Park, USA). One pass was made (Figure 4), which showed tumour cells expressing the smooth muscle markers actin and desmin. The tumour also showed focal expression of melanoma-specific antigen (HMB-45), and was focally positive for oestrogen and progesterone receptors. The histological diagnosis was LAM.

Later, high resolution CT of the thorax showed only a few thin walled cystic lesions consistent with early LAM. The woman was treated with oral progesterone, although she stopped taking her medication after only 2 weeks because of breast discomfort.

DISCUSSION

An atypical presentation of LAM mimicking ovarian carcinoma in a 35-year-old Chinese woman is described.

LAM is a rare disease characterised by proliferation of hamartomatous smooth muscle (LAM cells) in the lung and the lymphatic vessels, mainly in the retroperitoneal and mediastinal regions.¹ It is found exclusively in women of late reproductive age. LAM cells are differentiated from other types of smooth muscle cells by means of their reactivity with HMB-45 antibody,² which was also found in this patient. Pelvic LAM has

reportedly been described as an atypical precursor to pulmonary disease on the basis that an affected patient developed chest symptoms 3 weeks after initial presentation.³ Our patient, on the other hand, remains free of chest symptoms at the time of writing, 14 months after her first presentation. A patient described by Matsui et al was found to have mild pulmonary LAM 6 years after surgical removal of an enlarged retroperitoneal lymph node histologically proven to be LAM.² The patient described here did not have cutaneous stigmata of tuberous sclerosis or evidence of renal angiomyolipoma or ascites.

Open or thoracoscopic lung biopsy is usually required to establish the histological diagnosis of LAM.⁴ Abdominal lymphadenopathy is not uncommon: in one series of 80 patients with LAM, 39% had this pathology.⁵ Biopsy of the retroperitoneal lymph node is technically simpler and thus a good alternative to lung biopsy for histological diagnosis.⁶ Tru-cut biopsy should be performed as fine needle aspiration cannot confirm the tissue diagnosis (as in the present case). In fact, the unusual presentation of this patient led to the presumptive diagnosis of ovarian carcinoma, thus prompting fine needle aspiration to be performed.

The presence of low attenuation centres should alert clinicians to the possibility of LAM, particularly in

young women. The retroperitoneal lymph nodes in this patient showed low attenuation centres with -10 to +20 Hounsfield units. These centres were likely to be chylous collections caused by lymphatic obstruction and dilatation secondary to smooth muscle proliferation. Overdistension of this cystic chylous collection could result in chylous ascites, which would be associated with a sudden, marked reduction in the size of the cystic retroperitoneal masses.⁷

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