CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 41 (2017) 315–318

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

An extrapulmonary manifestation of lymphangioleiomyomatosis: A rare case report

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ARTICLE INFO

Article history:
Received 21 September 2017
Accepted 28 October 2017
Available online 9 November 2017

Keywords:
Abdominal neoplasm
Lymphangioleiomyomatosis
Surgery
Radiotherapy
Sirolimus

ABSTRACT

Lymphangioleiomyomatosis (LAM) is a rare and fatal disease which occurs almost exclusively in young women. The disease often affects lungs and most of the patients die from respiratory failure. It is often initially misdiagnosed as asthma or chronic obstructive pulmonary disease. The most common presentations of pulmonary LAM (P-LAM) include dyspnea and coughing. Chylothorax and spontaneous pneumothorax may be seen in advanced cases. Although rare, it may present with extrapulmonary LAM (E-LAM). Renal angiomyolipomas and abdominal lymphadenopathies (LAPs) are common in E-LAM cases. Pelvic retroperitoneal masses are very rare and often require exploratory laparotomy. Herein, we report a 36-year-old female case of a rare extrapulmonary manifestation of LAM who was treated with abdominal and thoracic surgery, radiotherapy and finally sirolimus.

1. Introduction

Lymphangioleiomyomatosis (LAM) is a rare, systemic, progressive disease characterized by proliferation of abnormal, smooth muscle-like LAM cells in lymph vessels, inducing obstruction and cystic formation [1]. It is also regarded as a low-grade, destructive, and rare neoplasm with metastatic potential [2]. In general, LAM occurs sporadically (S-LAM); however, in about%40 of the cases, it affects women with autosomal-dominant inherited tuberous sclerosis syndrome (TSC-LAM) [3]. It mostly affects young women. The thorax is the most commonly affected part of the body and the disease typically presents with pulmonary symptoms (P-LAM). The presentation of abdominal LAM without pulmonary findings is unusual [4], which results in a number of unnecessary interventions due to misdiagnosis or delayed diagnosis.

Although several treatment options have been reported for LAM in the literature, its specific treatment is still unclear. Surgery often yields unsatisfactory results in most of the patients [1] and there is a limited number of data on the efficacy of radiotherapy (RT) [5,6]. In addition, the prognosis is poor and mortality is usually associated with respiratory failure [3].

Herein, we report a 36-year-old female case of a rare extrapulmonary manifestation of LAM who was treated with abdominothoracic surgery, abdominal radiotherapy, and finally sirolimus, a mammalian target of rapamycin inhibitor. This work has been done in line with the SCARE criteria [7].

2. Clinical case description

A 36-year-old female patient was admitted to an external center due to the intermittent claudication in the left leg and pelvic abdominal pain for the past six months. The patient was previously operated with the preliminary diagnosis of a malignant mass originating from the left ovary. Based on the presence of an unusual cystic mass surrounding the left iliac artery and vein during surgery, the operation was terminated and the patient was referred to our clinic with the diagnosis of a retroperitoneal tumor.

Abdominal computed tomography (CT) showed a homogeneous, well-limited mass ($10 \times 9 \times 7$ cm) located next to the left ovary, surrounding the left iliac artery (Figs. 1 and 2). An accompanying conglomerated lymphatic tissue originating from the paraaortic space and extending through the left renal hilus was also detected. Chest X-ray showed normal findings, except for sequelae fibrotic changes. An ultrasound (US)-guided fine-needle aspiration cytology (FNAC) was performed for the cystic mass, which was closely adjacent to the vein, upon aspiration of non-bloody fluid, and CT-guided fine-needle aspiration biopsy (FNAB) was performed for the definite diagnosis. The result of the FNAC was reported as a low-grade mesenchymal neoplasm and the result of the FNAB was reported as an extra-renal angiomyolipoma and possible retroperitoneal leiomyomatosis due to strong positive immunohistochemical reaction for actin, desmin, and HMB-45.

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https://doi.org/10.1016/j.ijscr.2017.10.057

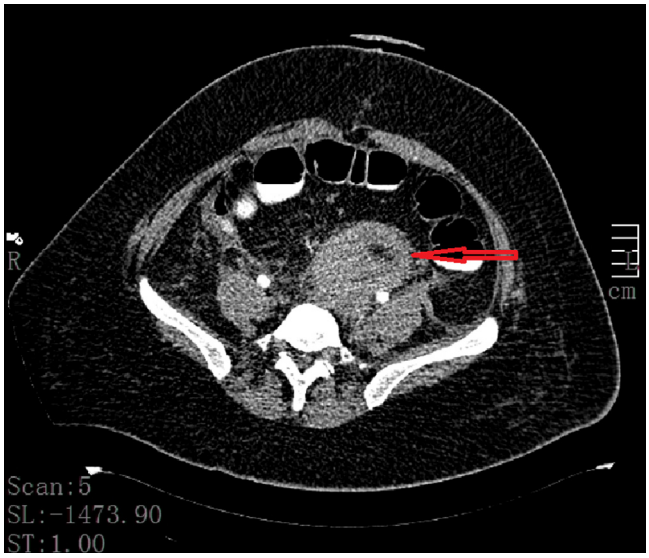

Fig. 1. A 10 × 9 cm cystic mass surrounding left iliac artery appearance on axial computed tomography (red arrow).

A written informed consent was obtained from the patient and she was operated due to a symptomatic mesenchymal tumor, and the cystic mass around the iliac veins was removed. The patient was discharged on the third postoperative day.

The permanent pathological examination revealed a low-grade (Grade I) vascular leiomyosarcoma. Meanwhile, the patient was re-admitted on the ninth postoperative day with dyspnea and chest X-ray showed massive pleural effusion in the right hemithorax. A tube thoracostomy was performed. Thoracic CT showed small, well-limited air cysts in the parenchyma, suggesting LAM (Fig. 3). There was a complex fluid collection in the abdomen as assessed by US. Since the pleural drainage did not decrease, a ligation of the thoracic duct and a mechanical pleurodesis was performed through thoracic surgery. The patient was referred to another hospital which specializes in thoracic surgery, since the drainage was unable to be controlled with these interventions. She underwent partial diaphragmatic resection and mechanical pleurodesis and was put on a fat-free diet. Five months later, she presented to our department with dyspnea and underwent left total pleurectomy due to left chylothorax. Since pleural drainage did not decrease following surgery, a decision was made to perform abdominal RT based on limited reports [4,5]. A three-dimensional conformal RT was applied in 17 fractions (total 3060 cGy), beginning from the left renal hilus and extending to the left paraaortic lymph nodes with an upper margin crossing the L1 vertebra and a lower margin at the level of the left capitis femoris. Following the first RT session, pleural drainage stopped. After three months of the fat-free diet, she was re-admitted with recurrent pleural effusion and re-underwent pleurodesis. After fat-free diet for three years, she was recommended a normal diet. At 129 months of follow-up, the patient had no chylothorax. However, at 97 months, she suffered from respiratory insufficiency requiring oxygen support. At 104 months, in the light of the recent reports in the literature, we initiated sirolimus treatment at 2 mg/day to maintain the blood concentration at 8–12 ng/mL. Bilateral hand eruption spontaneously disappeared during follow-up. However, statin therapy was initiated due to hypercholesterolemia. At 24 months of treatment, the patient showed a favorable clinical response and is currently functionally better in her daily activities (Table 1). At

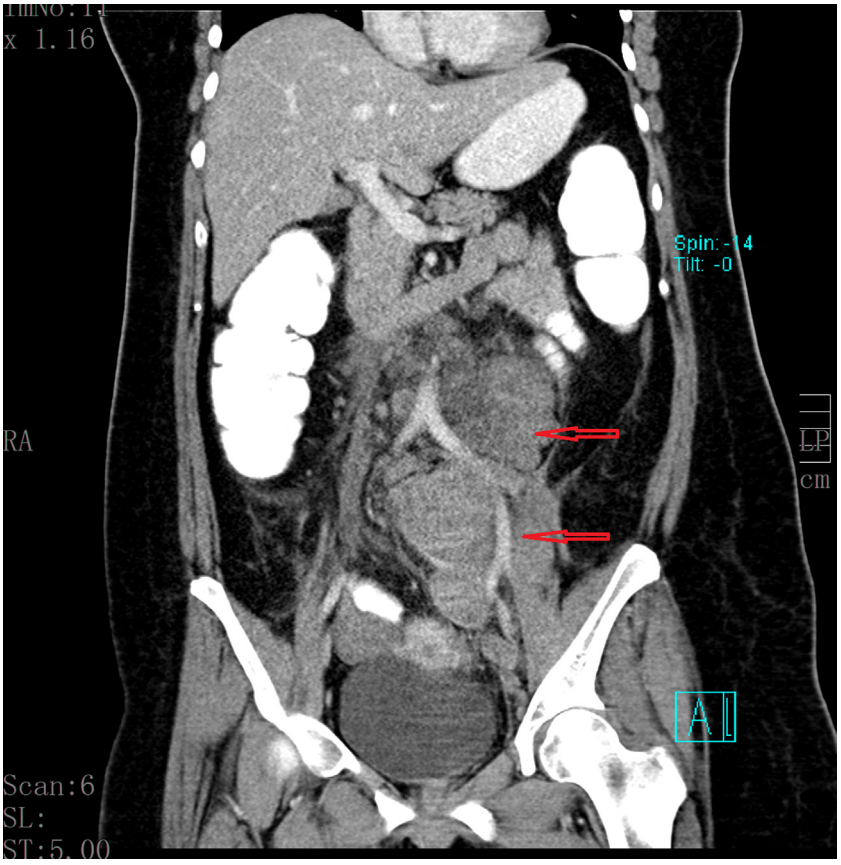

Fig. 2. A 10 × 9 cm cystic mass surrounding left iliac artery appearance on coronal computed tomography (red arrow).