A case of recurrent retroperitoneal lymphangioleiomyomatosis treated with progesterone therapy

Sir,

Lymphangioleiomyomatosis (LAM) is a rare, progressive disease of unknown etiology that generally affects women of childbearing age. It is characterized by the abnormal proliferation of smooth muscle cells causing gradual obstruction of small airways, resulting in the formation of thin-walled pulmonary cysts, leading to respiratory failure and death.[1] LAM is predominantly a lung disorder; however, extrapulmonary involvement, such as renal angiomyolipomas (which occurs in the presence of tuberous sclerosis [TSC]) and retroperitoneal masses, can occur in up to 75% of cases. No therapy is of proven benefit in LAM. Some studies support hormonal manipulation.[2] We report here a case of recurrent retroperitoneal LAM in a patient without any pulmonary involvement, who clinically responded with resolution of symptoms and objective ultrasound findings within a month, after receiving progesterone therapy.

A 24-year-old female patient presented to the emergency room with acute severe left sided abdominal pain. Physical examination and laboratories studies were unremarkable. A noncontrast computed tomography (CT) of the chest, abdomen and pelvis revealed multiple retroperitoneal, mesenteric, and pelvic sidewall hypodense masses with a sub-carinal mass of 3.2 × 3.5 cm with no lung parenchymal involvement [Figure 1]. Ultrasound of the abdomen [Figure 2a] revealed para-aortic and mesenteric masses with decreased echogenicity. The initial impression was retroperitoneal lymphadenopathy. A laparoscopic excisional biopsy of the mesenteric mass showed histologic findings consistent with LAM. Her histopathology [Figure 3a and b] revealed a network of lymphatic spaces lined by a single layer of endothelium, surrounded by fascicles and bundles of smooth muscle cells and occasional lymphoid follicles and congested vascular spaces are noted among the smooth muscle bundles. The patient received two doses of intramuscular (IM) medroxy-progesterone (Depo-Provera) 150 mg at 3 months interval. A 6 months follow-up ultrasound showed resolution of masses. However, the patient was lost follow-up for 2 years until the time she presented again with acute abdominal pain and retroperitoneal masses on ultrasound scan [Figure 2a]. She was given the Depo-Provera 150 mg IM. The patient clinically and radiologically responded within a week [Figure 2b].

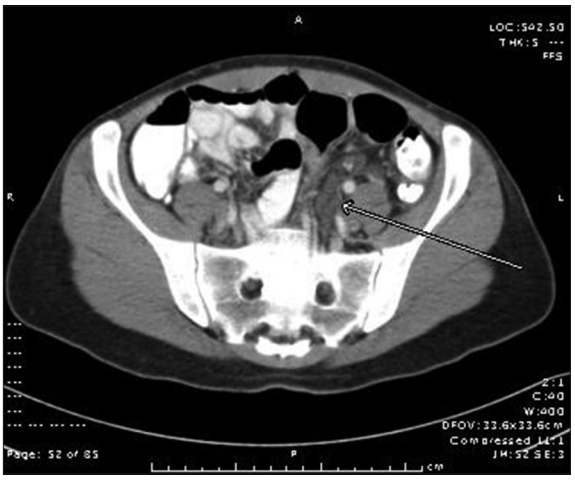

Figure 1: Computed tomography scans revealing evidence of retroperitoneal masses

Lymphangioleiomyomatosis, is a rare, idiopathic disorder involving the lungs, axial lymphatics in the thorax and rarely abdomen.[1,2] Extrapulmonary LAM as the initial presentation of the disease is highly unusual. Extra pulmonary LAM is usually associated with renal angiomyolipomas (54% of cases), enlarged abdominal lymph nodes (39%), and lymphangiomyoma (16%). Less commonly, ascites (10%) and hepatic angiomyolipoma (4%) may be present.[3,4] The clinical presentation of retroperitoneal LAM can vary from abdominal pain to shock due to bleeding from renal angiomyolipomas. LAM can occur without other disease ("sporadic" LAM) or in association with TSC.[1-4] Histopathology with immunohistochemical study of tissue is crucial in the diagnosis of this rare disease entity. Sonographic features and CT scan findings can be difficult to differentiate from malignancy. Supportive data for treatment of LAM is limited. Most therapies such as progestational, antiestrogen agents, rapamycin, and sirolimus provide improvement or stabilization of disease.[5] Our case illustrates a patient with

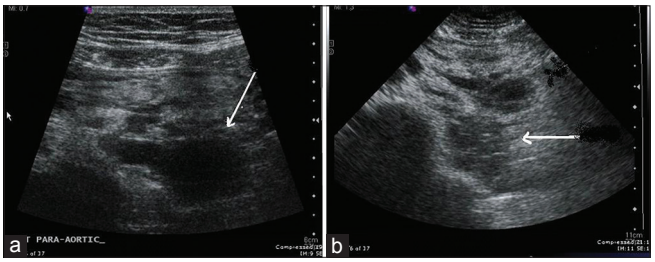

Figure 2: (a) Ultrasound revealing evidence of hypoechoic para-aortic masses (b) Ultrasound repeated after 1 week revealed resolution with decrease in size of the para-aortic hypoechoic masses

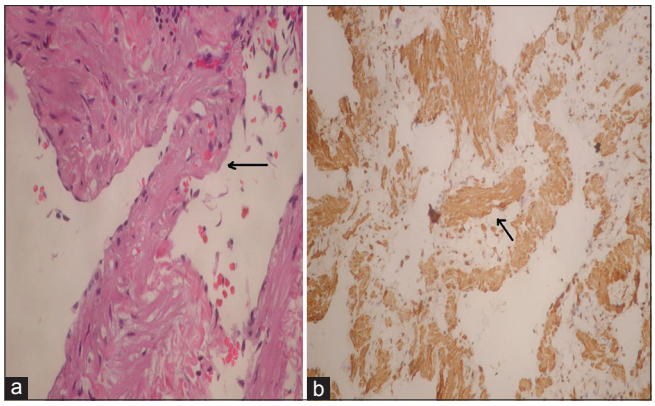

Figure 3: (a) Histopathology (H and E staining) revealed smooth muscle cells that are plump and possess pale to clear cytoplasm. There is also evidence of interspersed vascular spaces (b) Histopathology (HMB-45 staining) revealing granular cytoplasm consistent with retroperitoneal lymphangioleiomyomatosis

recurrent extrapulmonary LAM that had a resolution of symptoms and ultrasound findings after progesterone treatment.

Alina Basnet, Hamid Shaaban[1], William Kessler[2]

Departments of Internal Medicine and [2]Medical Oncology, Trinitas Regional Medical Center, Elizabeth, [1]Department of Hematology and Oncology, Saint Michael's Medical Center, Newark, New Jersey, USA

Address for correspondence:
Dr. Hamid Shaaban,
Department of Hematology and Oncology,
Saint Michael's Medical Center, Newark, New Jersey, USA.
E-mail: hamidshaaban@gmail.com

REFERENCES

1. Sullivan EJ. Lymphangioleiomyomatosis: A review. Chest 1998;114:1689-703.
2. Kelly J, Moss J. Lymphangioleiomyomatosis. Am J Med Sci 2001;321:17-25.
3. Derweduwen AM, Verbeken E, Stas M, Verschakelen J, Coolen J, Verleden G, *et al*. Extrapulmonary lymphangioleiomyomatosis: A wolf in sheep's clothing. Thorax 2013;68:111-3.
4. Gopinath D, Attarbashi S, Reid F, Seif M. Extrapulmonary lymphangioleiomyomatosis complicated by vesicovaginal fistula. J Obstet Gynaecol 2013;33:910-2.
5. Matsui K, Tatsuguchi A, Valencia J, Yu ZX, Bechtle J, Beasley MB, *et al*. Extrapulmonary lymphangioleiomyomatosis (LAM): Clinicopathologic features in 22 cases. Hum Pathol 2000;31:1242-8.

Access this article online	
Quick Response Code:	Website: www.urologyannals.com
	DOI: 10.4103/0974-7796.148664