Letter to the Editor

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Pulmonary Lymphangiomyomatosis and Kidney Angiomyolipomas

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Dear Sir

A woman aged 39, with close antecedents of spontaneous pneumothorax receives medical advice for progressive dyspnea and hemoptysis. X-ray shows bilateral diffuse pattern, and thin-walled aerial cysts of several sizes localized diffusely in both lungs, confirmed with high-resolution CT. Abdominal CT and echography reveal angiomyolipomas in both kidneys and uterine leiomyoma. Brain CT is normal. Transbronchial biopsy and bronchoalveolar lavage are un-specific. Routine blood analysis is normal. Tuberculin skin test, mycobacteria and Löwenstein culture of sputum are negative. Basal gasometry shows hypoxemia. Functional respiratory exploration: severe obstruction and small airway disease, total lung capacity 138%, diffusion lung of CO 32%. Cycloergometry suggests high arteriolar pulmonary resistance. Gallium lung scan is normal. Open lung biopsy is conclusive of lymphangiomyomatosis (LAM). With immunohistochemical techniques high cytoplasmic positivity for desmine, weak for actin and negative for estrogenic receptors are detected. Treatment with medroxyprogesterone acetate and tamoxifen was attempted. Total hysterectomy and bilateral salpingo-oophorectomy was made before the existence of gynecological clinics. The patient is currently undergoing treatment with medroxyprogesterone acetate.

The rare association of kidney angiomyolipomas and LAM has been reported. In this case the symptoms are typical, but without chylothorax. The existence of airway obstruction in a woman in fertile age with bilateral reticulonodular chest X-ray pattern makes us think of this sickness. High-resolution CT suggests the diagnosis [1], although it is easily confused with lung emphysema or histiocytosis. The diagnosis is histological, usually with open lung biopsy. It is difficult to distinguish LAM from tuberous sclerosis (TS). Despite the fact that kidney angiomyolipomas are characteristic of TS, their association with LAM has been reported [2,3], as it happens in our case. TS has been excluded on the base of the absence of heritage component and intracranial or cutaneous lesions.

The possibility of benign metastasizing leiomyoma was eliminated because of the existence of pneumothorax, reticulonodular chest X-ray pattern, severe obstructive ventilatory malfunction and proliferation of fusiform cells in vessels and respiratory airways.

Cases with receptors for progesterone and estrogen in muscular fibres in LAM have been reported in different studies, assuming a hormonal relation with the sickness evolution [4]. Different treatments as oophorectomy, tamoxifen and progesterone have been used separately or in association with similar results [5]. Anyway it is hard to get conclusions due to the short casuistry. The possibility of pulmonary transplant must not be forgotten. Death is described to occur in 10 years.

We present this case because it is very unusual – just a few cases over 100 of LAM all around the world – and for its association with kidney angiomyolipomas.

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