Pulmonary lymphangioleiomyomatosis is easily mistaken for more common lung diseases

To the Editor

I have read with interest the recent article by Shawki et al,[1] who reported the first case of pulmonary lymphangioleiomyomatosis in Iraq. I agree with the authors that pulmonary lymphangioleiomyomatosis (pulmonary LAM) is a rare disease, and due to its rarity, LAM is usually mistaken for more common pulmonary diseases like bronchial asthma, chronic obstructive pulmonary disease, and other forms of interstitial lung diseases.[2-4] This message should be made clear to all physicians particularly those beyond the pulmonary community, as many of these patients are being cared for by primary health physicians and internists. In fact the disease should be a diagnostic consideration in any woman in the childbearing age that presents with chronic pulmonary symptoms, pneumothorax of no obvious cause, or chylothorax. A computed tomography (CT) scan of the chest in combination with clinical presentation is usually sufficient for diagnosis. The CT chest usually shows bilateral diffuse thin-walled cystic changes of various shapes and sizes with or without pneumothorax or chylothorax. I believe that the presence of cystic changes along with confirmed chylothorax in this 38-year old lady was sufficient for the diagnosis of LAM, and I don't think that open lung biopsy was needed to confirm the diagnosis. Important issues like risk of pregnancy, air travel, and prognosis should also be discussed with the patient. I suggest to health authorities in Arab countries to make a registry for LAM to ascertain the incidence of this disease among Arabs and for future studies of this disease. We see a few cases of this disease in Qatar, and an interesting presentation of one case, will be published shortly.

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Reply from the Author

I agree with the remarks of Dr. Wanis from Qatar in that this rare disease may be misdiagnosed as other more common respiratory diseases and we need to increase the awareness of primary health physicians on this subject. The presence of clinical features consistent with LAM, in a non-smoker female, in her childbearing age, and with typical thoracic radiographic changes on HRCT, would preclude lung biopsy as a need for the diagnosis.

These are especially true when there are chylothorax, TSC (tuberous sclerosis complex) and angiomyolipomata. Nevertheless, the rarity of the disease, in addition to the need to register these cases with high accuracy; will guide the concept of carrying out lung biopsy. In the UK registry of LAM cases, out of 50 cases, 41 were finally diagnosed by lung biopsy.[5] It is an interesting idea to have a local registry in Arabic countries for lymphangioleiomyomatosis.

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