Sporadic LAM is an extremely rare condition affecting approximately 5 in 1,000,000 adult women. In TSC, LAM occurs in 30%–40% of adult females and exceptionally in men and children. AMLs are more common and often bilateral in TSC-LAM as compared to the sporadic form. HRCT chest reveals multiple usually 2–5 mm but occasionally up to 30 mm sized thin-walled cysts evenly distributed throughout the lung fields surrounded by normal lung parenchyma. Nodules, reticulations, and
The proposed diagnostic criteria for a definite diagnosis are compatible HRCT features with either characteristic histopathology or any of the extrathoracic findings, namely, AML (histological or radiologically confirmed), chylos effusion/ascites, lymphangioleiomyoma of the lymph nodes, or definite or probable TSC. Both our cases met the criteria for definite diagnosis in the absence of lung biopsy as both had a compatible HRCT features and AMLs. In addition, adenoma sebaceum was also present in the 2nd case which indicated a diagnosis of TSC. Sirolimus, an inhibitor of mammalian target of rapamycin, is the preferred treatment in those with abnormal or declining lung functions and symptomatic chylos effusions. Therapeutic drug monitoring is recommended to minimize the adverse events.

CONCLUSION

LAM is an extremely rare cystic lung disease which commonly occurs in association with renal AMLs and may be associated with TSC. Diagnosis is usually made by characteristic radiological and clinical criteria in the absence of lung biopsy. Treatment with sirolimus in selected cases may prevent the disease progression and worsening of lung function.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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