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Case Report

Thoracic-abdominal MDCT: A one-stop-shop procedure for diagnosis of lymphangioleiomyomatosis

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ABSTRACT

A 62-year-old asymptomatic woman was referred for an abdominal multidetector CT (MDCT) scan to evaluate a renal angyomiolipoma. Upper MDCT images revealed multiple, bilateral pulmonary cystic lesion in the costo-phrenic sulci and a thin-section MDCT scan of the chest was consequently performed. Thin-section MDCT showed several thin-walled cysts uniformly distributed throughout the lung parenchyma consistent with lymphangioleiomyomatosis (LAM). This radiological approach secured a definite diagnosis of LAM, thus avoiding further invasive procedures.

1. Introduction

Lymphangioleiomyomatosis (LAM) is an uncommon disease of unknown etiology, affecting almost exclusively middle-aged women. LAM is characterized by idiosyncratic smooth muscle cell proliferation (LAM cells) which leads to lung cysts, systemic lymphatic abnormalities, and abdominal tumors such as angyomiolipoma (AML). LAM can occur without evidence of other disease (sporadic LAM) or in association with tuberous sclerosis complex (TSC).[1]

The initial imaging modality technique used for the evaluation of patients with suspected LAM is the chest radiograph. If clinical data and/or chest radiography findings are supportive of LAM (young non-smoking women with unexplained pneumothoraces, chylous effusions, obstructive functional impairment etc.) a thin-section computed tomography (CT) study is indicated and may be supplemented. There is less certainty on to whether patients with renal AMLs should be screened for pulmonary LAM, although some authors advocate this.[1,2]

Herein, we present a case of a postmenopausal woman with LAM diagnosed by an abdominal multidetector CT (MDCT) scan performed to evaluate a renal AML.

2. Case report

A 62-year-old asymptomatic woman (with an unremarkable past medical history) was referred for an abdominal MDCT scan to evaluate a renal AML, which was suspected based on a previous abdominal ultrasound. Abdominal MDCT scan showed a 4 cm low-attenuating (fatty) renal mass arising from the lower pole of the left kidney consistent with AML; however, the upper MDCT images revealed also the presence of multiple bibasal pulmonary cysts (Fig. 1). Therefore, a thin-section MDCT scanning of the chest was performed, and showed several thin-walled cysts uniformly distributed throughout the lung parenchyma consistent with LAM (Fig. 2). This patient denied any family history or signs or symptoms of tuberous sclerosis complex.

3. Discussion

As in our report, LAM is increasingly recognized in post-menopause women, in whom a definite diagnosis is often challenging because tissue biopsy may be difficult to obtain.[3–5] Although the MDCT appearance of pulmonary LAM may be striking and characteristic, it may still overlap with that of other cystic diseases, such as lymphoid interstitial pneumonia (LIP), as well as pulmonary emphysema, indeed warranting a surgical lung biopsy.[6] The gold standard for the diagnosis of LAM is thoracopic lung biopsy. In patients unable to undergo a thoracopic lung biopsy, the diagnosis can be made on the basis of a transbronchial biopsy, especially in conjunction with immunohistochemical staining with human

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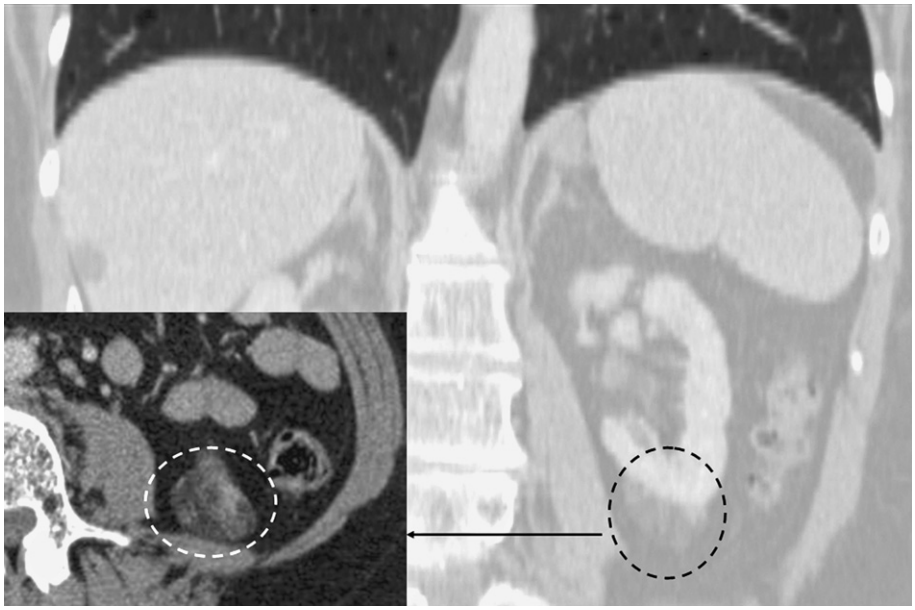

Fig. 1. Coronal MDCT reformation of the upper abdomen shows a 4-cm mass (black circle) arising from the lower pole of the left kidney; the zoomed axial image, reformatted with a soft-tissue window setting, better defines the fatty tissue containing mass (white circle), consistent with AML. Small cysts are clearly displayed in the lower lung parenchyma included in the standard abdominal MDCT scan.

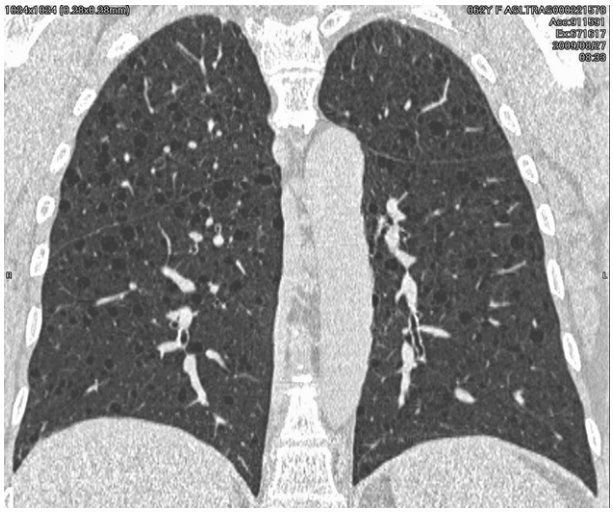

Fig. 2. Coronal reformation of thin-section MDCT of the chest showing thin-walled small cysts randomly distributed throughout both lungs. Such appearance is typical of LAM.

melanoma black (HMB)-45.[7] However, the striking combination of classical appearance of pulmonary LAM and renal AML on MDCT in a female patient allows a definite diagnosis of LAM, thus avoiding a tissue biopsy.[8]

4. Conclusions

Our report highlights the importance of both extending down to the kidneys the MDCT scan of the chest when evaluating patients with suspected LAM, and looking carefully the basal lung parenchyma included in abdominal MDCT scan undertaken for the evaluation of renal AML.

Conflict of interest

The authors have no conflict of interest.

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