Abdominal Findings in Lymphangioleiomyomatosis: A Report of Two Cases

Patología abdominal asociada a la linfangioleiomiomatosis

To the Editor,

Lymphangioleiomyomatosis (LAM) is a rare interstitial lung disease that mostly affects women, and is characterized histologically by the proliferation of atypical smooth muscle cells in the lymphatic system. Radiologically, LAM in the lung parenchyma is characterized by the presence of multiple pulmonary cysts (Fig. 1a). Abdominal findings associated with LAM are less well known, although they have been described in up to 70% of patients. The most common is renal angiomyolipoma (AML), which can appear in 20%–54% of cases. Other findings are lymphangioleiomyomas, lymphadenopathies and chylous ascites.1

To draw attention to these abdominal findings, and to recognize their clinical importance, we present 2 cases of LAM associated with abdominal disease.

The first case is that of a 42-year-old woman who presented clinically with a pneumothorax that required a chest drain. During the same procedure, cysts were removed from the apex of the lung, with a histological diagnosis of LAM cysts. A subsequent chest computed tomography (CT) incidentally detected a right renal mass (Fig. 1b). The mass was exophytic, with homogeneous density, measuring about 4-cm, and did not contain fat. Radiologically, it corresponded to an indeterminate renal mass, but given that the patient had already been diagnosed with LAM, and that fat was not evident within the mass, it was suggested that it could be consistent with an atypical AML. A percutaneous biopsy performed to rule out...

Fig. 1. Radiological findings in the case, characterized by (A) multiple pulmonary cysts, (B) a right renal mass (yellow arrow), and (C) multiple round cystic lesions in the retroperitoneum (yellow arrows) and pelvis.

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any other type of renal tumor confirmed that it was indeed an AML. The patient remains asymptomatic as regards the renal mass, and is undergoing checkups.

The second case is that of a 38-year-old woman in whom suspected cancerous ovarian cysts were detected during a gynecological examination. On the abdominal CT for staging and diagnosis, multiple round cystic lesions were observed in the retroperitoneum and pelvis (Fig. 1c). Pulmonary cysts were seen in the lung bases. The patient had no respiratory symptoms. Radiologically, the retroperitoneal and pelvic cysts were consistent with lymphangioleiomyomas, and the finding of cysts in the lung parenchyma confirmed the diagnosis of LAM. In this case, the disease presentation was the abdominal findings.

AMLs are benign tumors that are characterized radiologically by the existence of fat; however, this is absent in 5% of tumors, making them more difficult to diagnose. Treatment is conservative unless complicated by bleeding, which may require surgery or embolization. It has been reported that its association with LAM is so strong that, as in this case, when a renal mass is identified in a patient with LAM, an AML should be suspected.  

Lymphangioleiomyomas are cystic dilations of the lymphatic system that are produced by proliferation of the atypical cells of the LAM. Size may vary depending on gravitational factors or diet, the patient may be asymptomatic or report non-specific abdominal discomfort, and the condition can be confused with other processes such as lymphadenopathies or neoplasia, as in the case presented. There is no effective treatment.

In conclusion, we believe that physicians should be aware of the abdominal symptoms associated with LAM, as these may help diagnose the disease and obviate the need for more aggressive tests. Knowledge of the clinical importance of these findings may also avoid unnecessary surgery. We suggest that abdominal CT scans be performed in cases of chest CT consistent with LAM.

Conflict of Interests

The authors declare that they have no conflict of interests.

References


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Stenosing Esophageal Carcinoma Diagnosed by Endobronchial Ultrasound

Carcinoma estenosante esofágico diagnosticado por ecobroncoscopia

To the Editor,

Stenosing tumors of the esophagus occasionally pose a diagnostic challenge to digestive endoscopists due to the difficulty of access through the esophageal lumen. We describe a case of stenosing esophageal carcinoma diagnosed by endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA).

The patient was a 71-year-old man, smoker, with an alcohol intake of 30 g/day, who presented with a 2-month history of dysphagia associated with constitutional syndrome. Physical examination was normal. Laboratory tests were unremarkable, apart from a total bilirubin of 1.3 mg/dl. Computed tomography (CT) detected a 33 mm × 26 mm × 48 mm mass in the upper third of the esophagus (Fig. 1A). Gastroscopy showed stenosis of the cervical esophagus, apparently extrinsic, that prevented advancement of the endoscope. During the transesophageal endoscopic ultrasound (EUS), stenosis of the esophageal lumen made it difficult to introduce the endoscope, and sampling by fine needle aspiration was not optimal. Aspiration performed in the area most proximal to the lesion showed only atypical cells.

Since a diagnosis could not be made through the digestive tract, the possibility of a diagnostic approach using transtracheal ultrasound-guided fine needle aspiration (EBUS-FNA) was studied. Following endoscopic examination, we detected a protrusion of the pars membranacea in the upper third of the trachea. Using ultrasound, a 25 mm × 32 mm retrotracheal mass was identified 2 cm from the vocal cords (Fig. 1B), which was aspirated twice. After histopathological study of the samples, including immunohistochemistry studies (positive for cytokeratin 5 and p63 and negative for cytokeratin 7 and TTF1), esophageal squamous cell carcinoma was diagnosed.

Fig. 1. (A) Chest CT image in which a mass situated in the upper third of the esophagus can be seen (indicated with an arrow). (B) Endoscopic ultrasound image of the esophageal mass needle aspiration.

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