A case of pulmonary lymphangioleiomyomatosis and renal angiomyolipoma
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Case Type: Clinical Cases
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Patient: 32 years, female

Clinical History:
A patient with a history of recurrent pneumothoraces underwent chest X-ray and high-resolution chest and abdominal CT scanning, which revealed multiple, bilateral pulmonary cystic lesions, right pneumothorax and a left renal mass.

Imaging Findings:
The patient with a history of recurrent pneumothoraces underwent chest X-ray and high-resolution chest and abdominal CT scanning, which revealed multiple, bilateral pulmonary cystic lesions, right pneumothorax and a left renal mass. AP chest X-ray revealed irregular cavities and evidence of prior lung resection (Fig. 1). A lobulated right anterior pneumothorax was also visible inferiorly on the lateral view (not shown).

A non-contrast high-resolution chest (Fig. 2) and upper abdominal (Fig. 3) CT study with 5mm sections demonstrated multiple thin walled cysts diffusely involving both lungs. The largest complex cyst abnormality was in the right upper lobe, near the apex, measuring 4.2cm x 4.2cm. There was a small right-sided pneumothorax (Fig. 2b). Post-surgical changes were evident at the right apex. Posterior calcified opacities conforming to the pleural contour, were also seen, and probably related to prior pneumothoraces or prior pleurodesis.

CT of the upper abdomen revealed distortion of the left renal contour due to the presence of a mass containing fat and a calcification. There was no evidence of mediastinal or hilar lymphadenopathy.

Discussion:
Lymphangioleiomyomatosis (LAM) is a rare, idiopathic hamartomatosis characterized by smooth muscle cell hyperplasia along the terminal bronchioles in the lung, the lymphatic and blood vessels of the thorax and the retroperitoneum. The pathogenesis of LAM is still unclear. Pulmonary LAM can occur as part of the tuberous sclerosis complex. Some investigators claim that isolated pulmonary LAM and LAM associated with renal angiomyolipomas are a rough form of tuberous sclerosis. Pulmonary involvement in tuberous sclerosis is very uncommon, occurring in only 0.1%–1% of cases. The prevalence of LAM in women with asymptomatic tuberous sclerosis may be as high as 34%. LAM has been rarely described in males with tuberous sclerosis. The majority of patients are women. Patients present with pulmonary symptoms and signs such as increasing shortness of breath, cough, pneumothorax, chylous pleural effusion and haemoptysis. Some patients have abdominal pain or a painless abdominal mass, either in isolation or coincident with chest symptoms. The classic triad of chest radiographic findings includes a reticular interstitial pattern, chylous pleural effusion and recurrent pneumothoraces. The
interstitial changes seen on chest radiography represent the superimposition of the thin-walled cysts uniformly distributed throughout the lungs that are a hallmark of LAM on computed tomography (CT). Some authors have described the radiological pattern as a honeycomb appearance of small cysts, while others have described larger cysts. It has been suggested that the size of the cysts tends to be larger in more severe disease.

The four major abdominopelvic abnormalities seen in patients with LAM are renal angiomyolipoma (AML), as in this case; lymphadenopathy; lymphangioma; and chylous ascites.

Renal AML is the most common tumour associated with LAM. AMLs are benign, non-capsulated masses containing varying amounts of abnormal blood vessels, smooth muscle and mature fat. It is frequently associated with tuberous sclerosis. The key to diagnosis of an AML is the demonstration of fat. Because of its sensitivity in detection of small foci of fat, CT is the optimal imaging modality. Careful focal sampling of the low-attenuating regions within the mass must be performed, since the masses are frequently heterogeneous. A major but uncommon complication of renal AML is haemorrhage, which may manifest as flank pain or shock.

The recommended management of renal AML is based on tumour size and symptoms. Patients with asymptomatic lesions smaller than 4.0 cm in diameter may be followed up with annual US or CT and those with lesions larger than 4.0 cm in diameter by semiannual US or CT. Only patients with progressive symptoms should undergo nephrectomy or preferably a partial nephrectomy or embolisation.

LAM is believed to be hormonally mediated and therapies have been directed towards reducing circulating oestrogen levels. Lung transplantation has recently become an option for patients with end-stage LAM, but it has been associated with increased morbidity owing to unique complications related to the underlying disease, such as intraoperative bleeding due to pleural adhesions, native lung pneumothorax, chylous effusion, and ascites.

Chest and upper abdominal CT scanning is highly capable, as shown in the present case, of achieving an accurate evaluation of both pulmonary LAM and renal angiomyolipoma.

**Differential Diagnosis List:** Pulmonary lymphangioleiomyomatosis and renal angiomyolipoma

**Final Diagnosis:** Pulmonary lymphangioleiomyomatosis and renal angiomyolipoma

**References:**


Description: AP chest X-ray revealed irregular cavities and evidence of prior lung resection with minor scarring. Origin:
Figure 2

**Description:** Multiple thin walled cysts randomly involving both lungs are evident. The largest complex cystic abnormality is in the right upper lobe, near the apex, measuring 4.2cm x 4.2cm. **Origin:**

**Description:** There is a small right-sided pneumothorax. **Origin:**
Description: Unenhanced upper abdominal CT revealed distortion of the left renal contour due to the presence of a mass containing fat and a calcification. There was no evidence of mediastinal or hilar lymphadenopathy. Origin: