CASE OF THE MONTH

Acute abdominal pain and shortness of breath in a female patient

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Case presentation

A 36-year-old female patient presented to the local hospital with swelling and pain in her abdomen, fever and shortness of breath for 1 week. The patient was a smoker and had been diagnosed with asthma by her general practitioner several years previously, but had no other medical history of note. On clinical examination, she was found to have a large right-sided abdominal mass, which was thought at the time to indicate hepatomegaly, and expiratory wheeze with bilateral fine crepitations. She had an oxygen saturation of 92% in air. The patient had a chest radiograph, the results of which

were unremarkable, and an abdominal radiograph, which is shown in Figure 1. An ultrasound was arranged to evaluate the palpable abdominal mass; this demonstrated a large highly echogenic mass in the right upper quadrant. The patient underwent CT examination of the chest, abdomen and pelvis (Figures 2 and 3). In order to refine the diagnosis, a CT brain scan was performed, which had a normal result.

What are the relevant imaging findings seen in Figures 1–3?

What differential diagnosis was considered prior to the CT brain scan?

What is the likely final diagnosis?

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Figure 1. Abdominal radiograph demonstrating (a) the outline of a large soft tissue mass in the right upper quadrant, displacing bowel loops.

Figure 2. Coronal image from a portal venous phase CT of the abdomen and pelvis demonstrating (a) a large heterogeneous mass containing fat, soft tissue and blood vessels; the mass arises from the upper pole of the right kidney.

Figure 3. Axial image from a high-resolution CT of the chest (lung windows) demonstrating (a) multiple uniform thin-walled cysts in a symmetrical distribution throughout the lungs and (b) a large low-density right pleural effusion suggestive of a chylothorax.

Findings

Figure 1 shows a plain abdominal radiograph demonstrating the outline of a large soft-tissue mass in the right upper quadrant, displacing bowel loops. Figure 2 shows the coronal reformat of the portal venous phase CT demonstrating a large heterogeneous mass containing fat, soft tissue and blood vessels; the mass arises from the upper pole of the right kidney. High-density areas within the mass are suggestive of haemorrhage, and surrounding free fluid is present. Figure 3 shows a high-resolution CT (HRCT) scan of the chest demonstrating multiple uniform thin-walled cysts in a symmetrical distribution throughout the lungs and a large low-density right pleural effusion suggestive of a chylothorax (average -20 HU).

A CT scan of the brain was performed to differentiate between tuberous sclerosis complex (TSC) and lymphangioleiomyomatosis (LAM), both of which are associated with the above appearances. The brain imaging was normal; however, in TSC, findings include cortical tubers and subependymal nodules. The findings of multiple thin-walled lung cysts, chylothorax and a large solitary angiomyolipoma of the kidney in a pre-menopausal female patient with no evidence of TSC are in keeping with a diagnosis of LAM.

Discussion

LAM is a rare disorder characterised by the proliferation of abnormal smooth muscle cells in the lungs and lymphatics. It affects almost exclusively pre-menopausal women, and presentation usually includes shortness of breath or pneumothorax [1, 2]. Chest radiograph usually demonstrates a reticular appearance throughout the lungs. These reticular lines are defined on HRCT as cyst walls, with thin-walled cysts distributed

in a predominantly uniform pattern throughout the lungs. There may be a pleural effusion of a chylous nature owing to lymphatic involvement. Other chest manifestations include thoracic duct dilatation and, rarely, non-calcified lung nodules [1]. LAM is associated with abdominal findings including angiomyolipoma (AML), lymphangioleiomyomas, lymphadenopathy and chylous ascites. AML are hamartomas which can arise from hepatic or renal tissue. These lesions are heterogeneous on CT (containing fat, smooth muscle and blood vessels) and fat content is virtually diagnostic [1, 3]. AML can occur sporadically, but occur in up to 80% of cases of TSC and 50% of cases of LAM [3]. AML may be asymptomatic or present with abdominal pain, and can haemorrhage spontaneously. The treatment of choice is embolisation, which has durable results [4].

TSC is an autosomal dominant triad of mental retardation, epilepsy and adenoma sebaceum. AML are common in TSC and may be multiple and bilateral. Up to one-third of women with TSC may demonstrate lung cysts similar to those found in LAM; however, patients with LAM do not have the typical skin and central nervous system findings of those with TSC [1, 2, 5]. Lung nodules are more likely to be seen in TSC than in LAM sufferers [3].

Langerhans cell histiocytosis is associated with cigarette smoking, and features diffuse pulmonary cysts that are more marked in the upper lobes, with relative basal sparing. Unlike in LAM, the cysts are often bizarre and thick walled, and nodules are common and may cavitate [1].

Lymphangioleiomyomatosis is a progressive disease leading to chronic respiratory failure, and effective treatment options are limited. Patients may require lung transplantation, and treatment decisions (such as pleurodesis) should be taken carefully so as not to compromise suitability for transplant [6]. It is important for the radiologist to be aware of the associations of AML with LAM and TSC in order to carry out appropriate imaging investigations.

References

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