Case 17186

Embolisation of haemorrhagic lymphangioleiomyomatosis-associated angiomyolipoma
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Section: Interventional radiology
Area of Interest: Interventional vascular Kidney Oncology
Procedure: Embolisation
Imaging Technique: CT
Case Type: Clinical Cases
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Patient: 42 years, female

Clinical History:

A 42-year-old woman with known lymphangioleiomyomatosis (LAM) and renal benign tumours accessed our Emergency Department complaining of a two-week history of continuous left flank pain.

Imaging Findings:

Unenhanced chest and upper abdominal CT scan showed the stability of LAM-associated small thin-walled cysts in the lungs (Fig. 1) and detected multiple bilateral inhomogeneous renal lesions (Fig. 2). The largest lesion (15x10x9 cm) was localized in the left kidney with hypodense fatty areas and hyperdense hemorrhagic areas, without signs of current extravasation. Enhanced upper abdominal CT scan (Figure 3) demonstrated the presence of pseudoaneurysms within the lesion.

The CT findings allowed to reach the diagnosis of hemorrhagic angiomyolipoma (AML) of the left kidney.

A transcatheter selective arterial embolization (SAE) was performed to treat the renal lesion with a recent history of bleeding, avoiding radical nephrectomy. Once a super-selective catheterisation of AML feeding vessels was achieved (Fig. 4), 700-900 µm Embosphere® microspheres were injected and IDC™-18 coils were released in order to assure the occlusion of the hypertrophic capsular arteries. A control selective arteriography (Fig. 5) showed devascularization of almost 90% of the lesion, with a rate of residual renal parenchyma of 30%.

Discussion:

Background

LAM is a rare disease characterized by proliferation of neoplastic pulmonary interstitial smooth cells (LAM cells), resulting in a cystic pneumopathy and recurrent pneumothorax. [1,2] The disease almost exclusively affects women of childbearing age. [1] LAM may occur sporadically but is commonly associated with tuberous sclerosis complex (TSC-LAM). [3]

Clinical perspective
LAM usually presents with obstructive dyspnoea and pneumothorax. [2] Approximately half of patients with sporadic LAM and nearly all ones with TSC-LAM are affected by AMLs, usually of the kidney. [1] When haemorrhagic complication of AML occurs, the associated clinical signs are hematuria, palpable mass and symptoms of retroperitoneal haemorrhage such as flank pain or shock. [4]

**Imaging findings**

On abdominal CT or MRI, AML usually shows hypervascularity, structural inhomogeneity and the presence of fatty areas. The latter are particularly useful to rule out renal cell carcinoma.[4] Abdominal CT provides information on possible bleeding. Once current or recent signs of extravasation are detected at CT scan, a sudden selective arterial embolization should be required in order to perform a nephron-sparing treatment and to avoid radical or partial nephrectomy.[4]

**Therapeutic options**

The treatment of choice for a hemorrhagic AML is transcatheter selective or super-selective arterial embolization (SAE). SAE is less invasive than a surgical intervention and enables targeted treatment of bleeding vessels with a low risk of severe complications.[5] Although technical success has been reported to be achieved in 80 to 90% of AMLs treated with SAE, long-term follow-up has shown tumour recurrence in over 30% of patients, with a higher recurrence rate of 43% in patients with TSC. At 5 years and 10 years from initial SAE, the rate of reembolisation was 29% and 63%, respectively. For this reason, patients with TSC-LAM who underwent SAE for AMLs should be surveilled to detect possible recurrences.[6] Moreover, SAE has been demonstrated to avoid renal surgery in patients with AMLs. The need for surgical operation for AMLs following SAE is 4% at 5 years and 10 years and is associated with the failure of initial embolization.[7]

Alternative treatments for AML are radical or partial nephrectomy and mTOR inhibitors.[4] Arterial embolization for AMLs is also performed for preventing haemorrhages in at-risk lesions. The risk of spontaneous haemorrhage increases significantly to 50-60% in lesions greater than 4 cm or with the presence of aneurysmal blood vessels.[4, 8]

**Take-home message**

Haemorrhagic renal AML must be suspected in patients with LAM presenting with flank pain or shock. A CT abdominal scan should be performed in order to correctly plan the treatment of AMLs.

**Differential Diagnosis List:** LAM and haemorrhagic renal left AML, LAM and bilateral hemorrhagic myelolipoma, LAM and retroperitoneal liposarcoma with haemorrhage, LAM and renal-cell carcinoma

**Final Diagnosis:** LAM and haemorrhagic renal left AML

**References:**


Description: Unenhanced chest CT. Bilateral diffuse LAM. The largest cyst is subpleural, and is located in the apical segment of the right superior lobe (black arrow). Origin: © Diagnostic and Interventional Radiology, Imaging Department, Azienda Ospedaliero Universitaria Pisana, University of Pisa, Pisa, Italy
Description: Unenhanced abdominal CT. Bilateral inhomogeneous renal lesions are demonstrated. The largest lesion (15x10x9 cm) is localized in the left kidney with hypodense areas of fat (red arrow) and hyperdense hemorrhagic areas (black arrow). Origin: © Diagnostic and Interventional Radiology, Imaging Department, Azienda Ospedaliero Universitaria Pisana, University of Pisa, Pisa, Italy
Figure 3

Description: Abdominal CT after iv administration of contrast material, coronal MPR. Hypertrophic and tortuous arteries were demonstrated with pseudoaneurysms (red arrow); no signs of active hemorrhage.

Origin: © Diagnostic and Interventional Radiology, Imaging Department, Azienda Ospedaliero Universitaria Pisana, University of Pisa, Pisa, Italy
Figure 4

Description: Preliminary angiography of left kidney. The image confirms the presence of multiple pseudoaneurysms (black arrows) without signs of current arterial supply of the hemorrhage.

Origin: © Diagnostic and Interventional Radiology, Imaging Department, Azienda Ospedaliero Universitaria Pisana, University of Pisa, Pisa, Italy
Figure 5

Description: Control angiography of left kidney after coil embolization of two collateral branches and two renal artery branches (red arrow). Devascularization of approximately 90% of the lesion and sparing of 30% of renal parenchyma are demonstrated. Origin: © Diagnostic and Interventional Radiology, Imaging Department, Azienda Ospedaliero Universitaria Pisana, University of Pisa, Pisa, Italy