Giant Renal Angiomyolipoma Associated With Pulmonary Lymphangioleiomyomatosis: A Case Report

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Citation

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Abstract

Renal angiomyolipomas are benign tumours known to appear sporadically and in association with genetic syndromes, including lymphangioleiomyomatosis. Treatment of large angiomyolipomas is usually indicated because of an increased risk of spontaneous haemorrhage.
We report in a patient with radiographic criteria of pulmonary lymphangioleiomyomatosis, a successful trans-arterial embolization of a giant angiomyolipoma with active anemization due to retroperitoneal bleeding and deferred surgical treatment due to persistent symptoms.

KEY OF DEFINITIONS FOR ABBREVIATIONS

AML: Angiomyolipoma.
LAM: Lynphangioleiomyomatosis.
TSC: Tuberous sclerosis complex.

INTRODUCTION

Angiomyolipoma (AML) is an uncommon benign tumour composed of fat tissue, thick-walled vessels and smooth muscle elements in varying proportions ([1], [2]). Lynphangioleiomyomatosis (LAM) is a rare disorder, characterized by a non-neoplastic proliferation of abnormal smooth muscle cells in the lung parenchyma and in the lymphatic system ([3]). Its association with renal AMLs has been reported previously ([1], [4]).

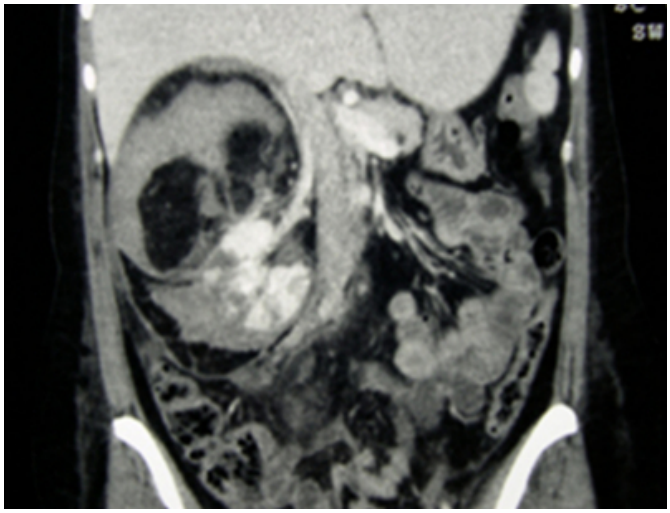
We report a case of exitous embolization in a patient with LAM and Wunderlich syndrome due to a giant renal AML, and deferred surgical treatment due to persistent symptoms.

CASE REPORT

A 38-year-old woman came to the emergency department due to the sudden onset of right flank pain. She present rapid drop of haemoglobin (from 11 to 7,3 g/dl) and haematocrit (from 33,7 to 22,7%), and hemodynamic instability. Computer tomography of the abdomen showed a large mass, suggestive of renal AML, with active extravasation of contrast media to a large retroperitoneal haematoma (figure 1.).

Figure 1

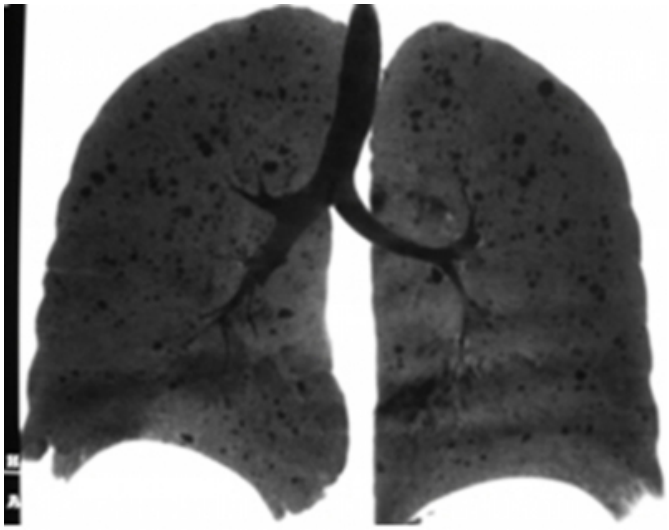
Figure 1: Abdomen CT showed a large mass, in the right kidney, suggestive of AML, with active extravasation of contrast media to a large retroperitoneal haematoma.



On thorax CT, multiple well-defined thin-walled pulmonary cysts, distributed diffusely throughout both lungs (figure 2.).

Figure 2

Figure 2: Multiple well-defined thin-walled pulmonary cysts, distributed diffusely throughout both lungs. Suggesting the radiological diagnosis of pulmonary LAM.

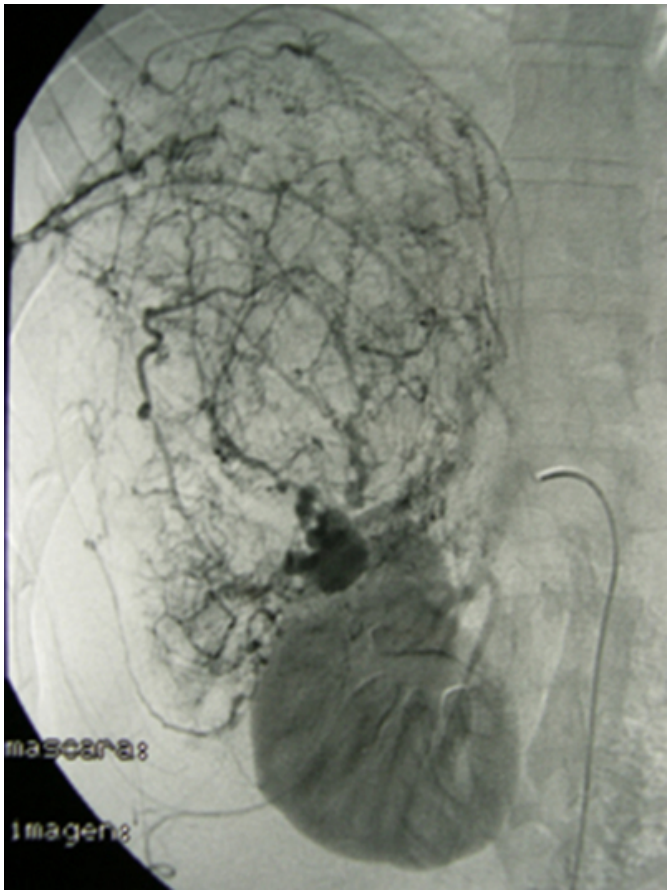


Suggesting radiological diagnosis of pulmonary LAM, she had no pulmonary symptoms, and no other stigmata of tuberous sclerosis complex (TSC).

An urgent arteriography was performed, with successful selective angioinfartion of the dilated renal arterial branches feeding the tumour, with microcoils and microspheres (figure 3.)

Figure 3

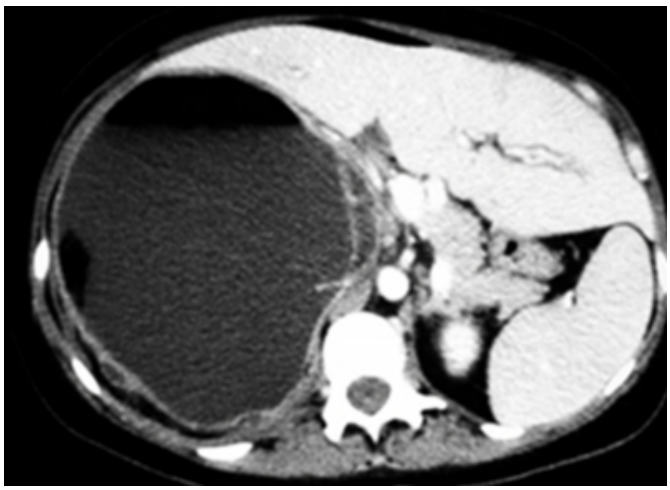
Figure 3: Arteriography showing successful selective angioinfartion of the dilated renal arterial branches feeding the large tumour, with microcoils and microspheres.



Three moths later, a control CT showed persistent renal mass with sings of necrosis and liquefaction (figure 4).

Figure 4

Figure 4: Control CT showed persistent renal mass with sings of necrosis and liquefaction.



The patient referred abdominal pain, nausea and fever, so a

radical nephrectomy was performed without complications.

COMMENT

AMLs appears as part of the TSC (60-80% have AMLs), or as sporadic occurrence in healthy individuals (70% of all AMLs occur sporadically in the general population) ([6,7,8,9]). More than 50% of AMLs are found incidentally ([9]). CT scan has been the most reliable modality for the diagnosis ([10]).

AMLs have a propensity to bleed, with size, multifocality, and vascular abnormalities being the main risk factor ([8, 9, 11]). Massive retroperitoneal haemorrhage from AML, has been found in up to 10% of patients and represents the most significant and feared complication ([2, 8]). Common signs or symptoms include flank pain, haematuria, palpable abdominal mass, and hypovolemic shock, which can be fatal ([2, 8]). Selective renal artery embolotherapy, has become increasingly popular as a treatment option in the management of renal AMLs ([6, 7, 11]). Its common, after embolization the development of moderate flank pain and low-grade fever ([6, 7]), our patient had these symptoms for almost one week. If the AML is bleeding in the emergent setting with a life-threatening haemorrhage may require total nephrectomy if explored. In such circumstances, selective embolization can temporize and, in many cases, provide definitive treatment.

LAM is a rare lung disease characterized by an abnormal proliferation of smooth muscle-like cells that over time can grow to obstruct airways, lymphatics and blood vessels ([12]). Commonly manifests as exertional dyspnea and recurrent pneumothorax ([3]). It often leads to irreversible, progressive deterioration of pulmonary function. Abdominal manifestations of LAM are common, the most frequent is renal AML seen in between 32-60% ([4]). Radiographic imaging is indicated in patients with LAM to screen for renal AML ([5]).

CONCLUSION

To our knowledge trans-arterial embolization of renal AML is a safe and effective procedure, controlling haemorrhage, but in large masses can not prevent surgical treatment in most patients.

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