



Giant leiomyoma of the renal capsule: CT and MR imaging features with pathologic correlations

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Summary Renal leiomyoma (RL) is a rare benign tumor, usually presenting as a small asymptomatic lesion, frequently found during autopsy with a higher incidence in women (2:1). However, larger RL can become symptomatic and show structural changes (such as hemorrhagic and cystic degeneration), thus posing a diagnostic challenge. A 49-year-old woman with a recent history of weight loss and left flank pain was referred to our institution. An ultrasound exam revealed the presence of a complex mass at the left kidney. Both CT and MRI scans were performed to characterize the renal mass, which appeared well circumscribed with no signs of local invasion. Moreover, CT and MRI provided valuable additional information such as the absence of both macroscopic and microscopic fat and the presence of calcifications, hemorrhagic areas, multiple cysts and delayed enhancement. A left nephrectomy was performed via laparoscopy. Histopathological evaluation confirmed the diagnosis of RL. Imaging techniques can aid in the identification of large RL and guide the differential diagnosis; however, histopathologic analysis is required for a definitive diagnosis.

Keywords Computed Tomography · Kidney neoplasm · Leiomyoma · Diagnostic imaging · Magnetic resonance imaging

Introduction

Renal leiomyoma (RL) is a rare benign neoplasm, accounting for less than 0.5% of all treated renal masses

[1], and is frequently found during autopsies [2]. It may originate from smooth muscle cells located in the renal capsule, pelvis and calices as well as renal vessels [3]. Although RLs are most commonly small sized asymptomatic lesions, clinical manifestations could become apparent in case of larger masses [4] and it has been hypothesized that RL growing in size could lead to intralesional hemorrhagic and cystic degeneration and even malignant transformation [5, 6]. In these cases, the radiological diagnosis is particularly challenging and the differentiation from malignant entities such as leiomyosarcoma [7] and renal cell carcinoma (RCC) is often difficult.

Case report

A 49-year-old woman with a recent history of weight loss and left flank pain was referred to our institution to perform an abdominal ultrasound that revealed the presence of a complex mass at the lower pole of the left kidney. To better evaluate the renal lesion, the patient underwent a CT scan that confirmed the presence of large renal lesion, with smooth and regular margins, presenting heterogeneous intralesional density and generating a mass effect on adjacent structures (Fig. 1). Based on CT findings, a final diagnosis could not be reached, and the possible malignant nature of the lesion could not be ruled out. Therefore, an MRI scan of the abdomen was performed. The MRI showed a heterogeneous mass, with both multiple cystic areas, hemorrhage and solid components, without intracellular fat (Fig. 2). Imaging features on both modalities were highly suggestive for a mass originating from the lower pole of the left kidney. Given the size of the lesion and since malignant entities such as cystic RCC or leiomyosarcoma could not be excluded, a surgical approach was deemed necessary and the patient underwent radical left nephrectomy

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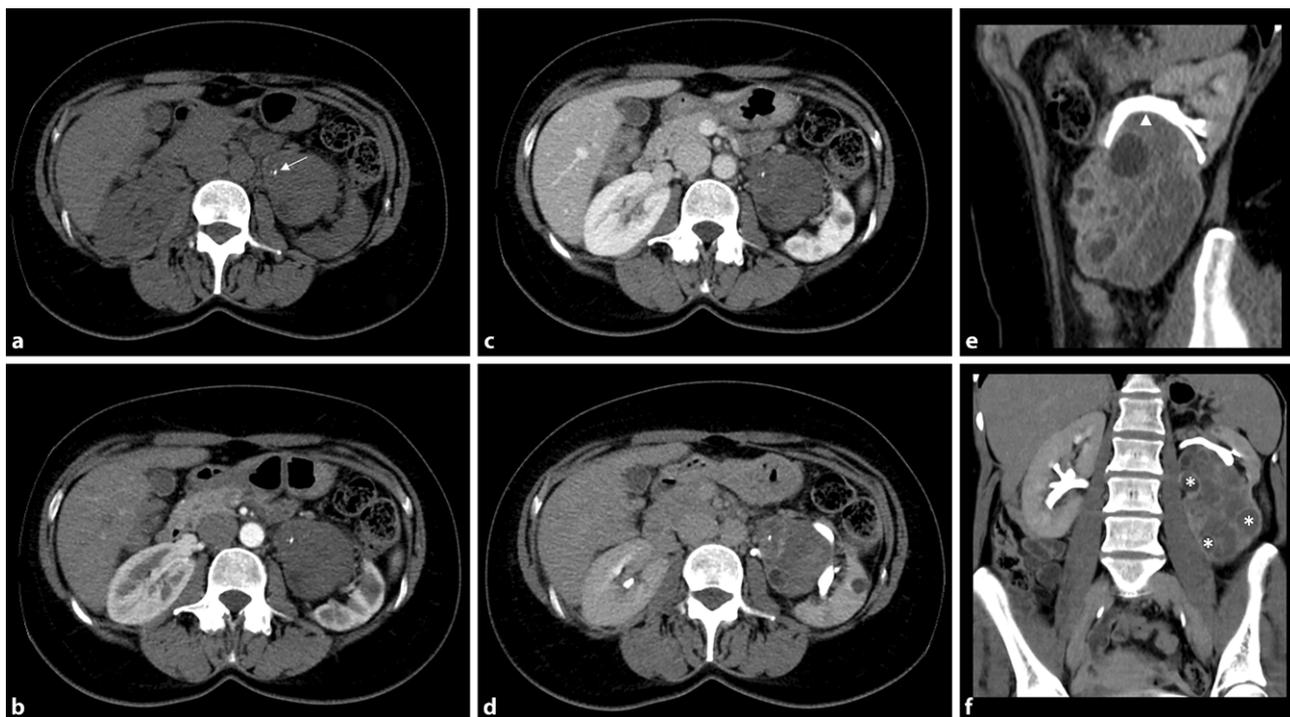


Fig. 1 Computed Tomography of the renal mass: the unenhanced image **a** shows a small calcification inside the lesion (*arrow*) and the absence of macroscopic fat; arterial (**b**), portal (**c**) and delayed (**d**) phase demonstrate a progressive and delayed enhancement of the lesion. Multiplanar reconstruc-

tion images on the sagittal (**e**) and coronal (**f**) planes of the excretory phase allow appreciating the mass effect on the ureter, that appears dislocated cranially without signs of infiltration (*arrowhead*), and the presence of multiple pseudo cystic formations inside the lesion (*asterisks*)

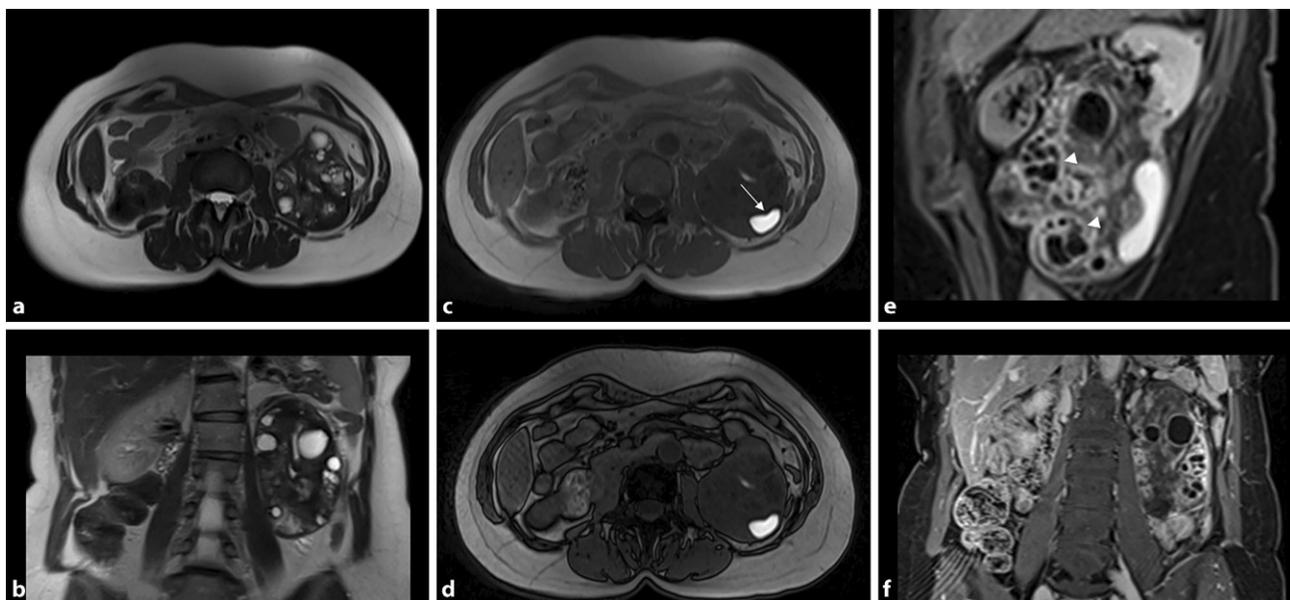
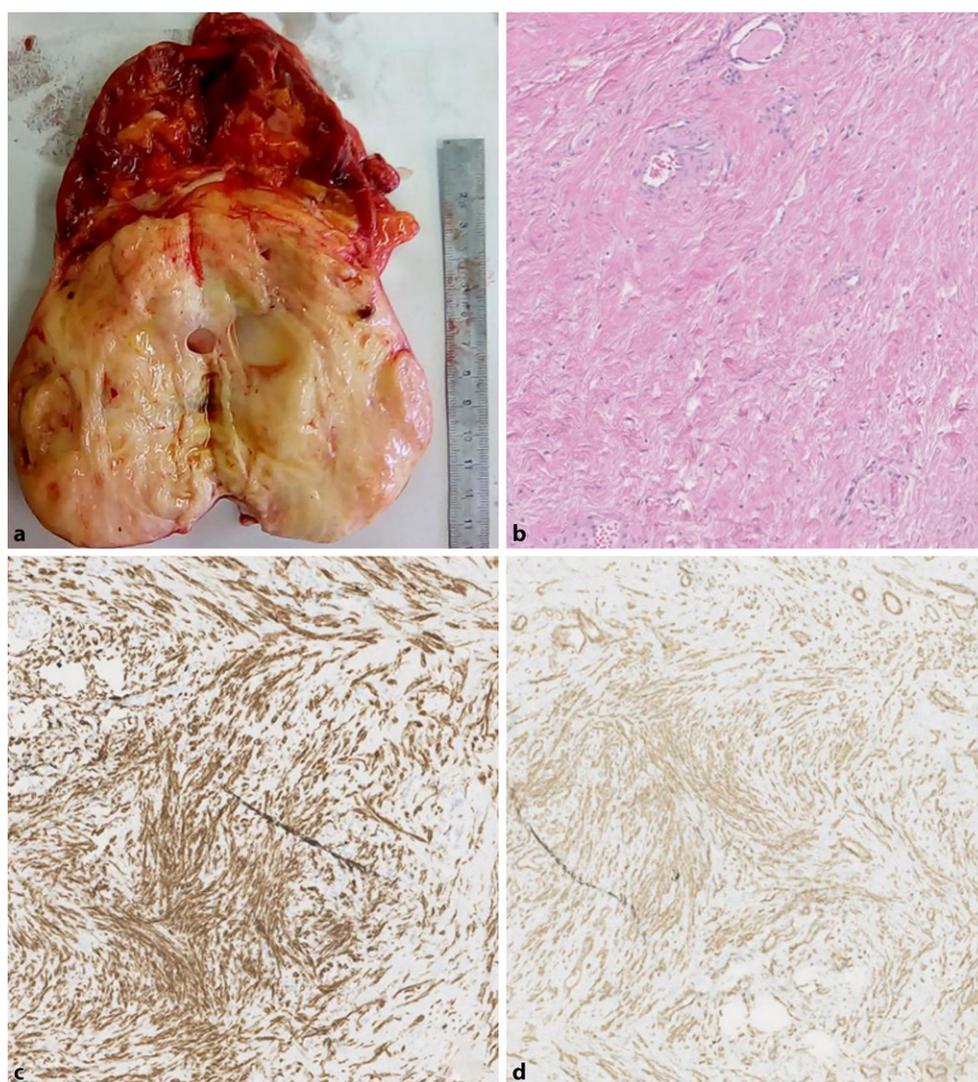


Fig. 2 Magnetic Resonance Imaging of the renal mass: the T2-weighted images on the axial (**a**) and coronal (**b**) planes show a highly heterogeneous renal lesion with cystic degeneration and hypointense tissue. No loss of signal can be appreciated when comparing the T1 in-phase (**c**) and out-of-phase (**d**) images, thus demonstrating the absence of intracellular fat; an

area of hemorrhage is found, markedly hyperintense in this sequence (*arrow*). Post-contrast delayed phase images on the sagittal (**e**) and coronal (**f**) planes show a late and relatively low contrast enhancement, more evident by the cysts (*arrowheads*)

Fig. 3 Gross features of the renal specimen are shown (a). A proliferation of spindle cells was evident in the hematoxylin and eosin stain, 86× magnification (b); in particular, on the top of the picture, renal tubules were entrapped by neoplastic fascicles. Clear reactivity to desmin (c) and smooth muscle-actin (d) was detected (50× magnification)



via laparoscopy. The specimen measured 16 × 7 × 4 cm; grossly, a white–yellowish colored mass was found, extending from the renal pelvis to the lower pole. It was well-capsulated, and on cut-section it appeared fasciculated (Fig. 3a). Microscopically, the neoplasm was characterized by low cellularity, and was composed by spindle cells, with elongated nuclei and a fascicular arrangement with interspersed amount of stroma similar to uterine leiomyoma (Fig. 3b). Some renal tubules were entrapped and vessels with thick walls could be observed. Necrosis and mitosis were absent. Vessels of the hilum and ureter were disease-free. The specimen was sampled entirely. No adipocytic component was detected within the lesion. Immunohistochemically the tumor showed a strong positivity for desmin (Fig. 3c) and smooth muscle-actin (Fig. 3d) and negativity for CD34, HMB45, S-100 protein and MART-1, excluding the possibility of angiomyolipoma and suggesting the diagnosis of renal leiomyoma. The patient did not suffer from any complications after surgery and is currently in good health.

Discussion

Leiomyomas are benign tumors deriving from smooth muscle cell, representing common uterine lesions [3]. They rarely occur in kidneys, with a higher incidence in women (2:1) and during the fourth decade of life [8]. While RLs are more frequently discovered incidentally, they can also become symptomatic when reaching significant volumes. It has been reported that larger RLs can lose their typical homogeneous appearance due to degenerative phenomena. When this occurs, the differential diagnosis becomes challenging and generally includes both benign lesions, such as angiomyolipoma (AMLs) and oncocytoma, and malignant ones, such as leiomyosarcoma and RCC [9]. Although we were not able to reach a pre-operative diagnosis, both the CT and the MRI scan provided useful information. First, they allowed confirmation of renal origin of the mass. Indeed, the lower pole of the left kidney was completely affected, while the kidney did not appear dislocated and the lesion showed regular margins and no evidence of local invasion, el-

ements disfavoring the hypothesis of a retroperitoneal infiltrating mass. Additionally, demonstrating the absence of a macroscopic fat component and the presence of calcifications and hemorrhagic areas, diagnostic imaging allowed ruling out both AMLs and its less common fat-poor variant with a reasonable degree of certainty [10]. Typical imaging findings of a large oncocytoma, like the presence of a central scar and homogenous early enhancement, were not present; however, these signs are variable and the hypothesis of oncocytoma could not be dismissed [11]. Clear cell RCCs typically appear hyperintense in T2-weighted images and large RCCs usually show areas of necrosis; however, these findings were not considered sufficient to exclude papillary RCC and other less frequent RCC variants from the differential diagnosis [12]. Furthermore, the abovementioned absence of radiological signs of local invasion and the regular and well-defined margins are uncommon features for leiomyosarcomas of such dimensions [6]. Another rare entity, the mixed epithelial and stromal tumor (MEST) of the kidney, was considered in the differential diagnosis; MEST typically occur in perimenopausal women and present as a well-circumscribed, multicystic and solid mass with delayed enhancement [13]. Finally, rare kidney sarcomas such as the dedifferentiated liposarcoma or the undifferentiated pleomorphic sarcoma might have been included in the differential diagnosis, although very rare and usually showing fat components [14, 15].

In conclusion, large RLs can cause mass effect, become symptomatic, show complex structural changes and possibly degenerate into their malignant counterpart. In these cases, reaching a radiologic diagnosis is extremely challenging and histopathologic examination is required. Nevertheless, this pathologic entity should always be considered in the differential diagnosis of renal masses at imaging.

Compliance with ethical guidelines

Conflict of interest A. Stanzione, M. Santangelo, F. De Rosa, A. Ponsiglione, G. Peluso, L. Insabato, and M. Imbriaco declare that they have no competing interests.

Ethical standards For this article no studies with human participants or animals were performed by any of the authors. All studies performed were in accordance with the ethical standards indicated in each case. For images or other information within the manuscript which identify patients, consent was obtained from them and/or their legal guardians.

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