

Case Report

Primary renal leiomyosarcoma: CT manifestations and correlation with pathologic findings

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Abstract: Primary renal leiomyosarcoma is an extremely rare tumor that is responsible for 50%-60% of all renal sarcomas. Treatment of this disease has been difficult due to the insufficient data, making the clinical information on primary renal leiomyosarcoma extremely important for understanding and treating this challenging disease. This work presents CT diagnosis of 8 cases of primary renal leiomyosarcoma collected from our clinical practice. Among all 8 cases, 7 lesions were in the periphery of the kidney, including 4 ill-demarcated lesions and 3 well-demarcated lesions. On plain scanning, 6 lesions were homogeneous iso- or hyper-attenuation and 2 lesions were heterogeneous with both iso- and hyper-attenuation. CT imaging showed that the tumors were composed of spindle cells interspersed with connective tissue collagen fibers. This study may help us understand primary renal leiomyosarcoma from a clinical aspect, preparing us clinical data on treating this disease.

Keywords: Primary renal leiomyosarcoma, patients, clinical study, CT, lesion

Introduction

Primary renal leiomyosarcoma is an extremely rare tumor constituting only 0.1% of all invasive renal tumors. Despite its rareness in the clinic, primary renal leiomyosarcoma is the most common sarcoma of the adult kidney, accounting for 50%-60% of all renal sarcomas [1-4]. Primary renal leiomyosarcoma can arise from the renal capsule, the renal parenchyma, smooth muscle fibers of the renal pelvis, or the main renal vein [5-7]. One of the largest clinical reports about primary renal leiomyosarcoma included 27 cases identified for 23 years from 3 large institutions of America [7, 8]. In addition to these clinical case reports with a large number of samples, most clinical studies only involve several case reports of CT and MR diagnosis of primary renal leiomyosarcoma [9-16]. Therefore, this study presents CT manifestations of 8 cases of primary renal leiomyosarcoma in the past 8 years.

Materials and methods

CT images of 8 patients with pathological proved primary renal leiomyosarcoma were clearly

notified and agreed with the study. Informed consent was waived by the Ethical Committees of our hospital since it is a retrospective study. There were 5 men and 3 women, with an average age of 47.3 years old. 6 patients presented with related symptoms or signs, including obvious back pain in 4 patients, acute severe abdominal pain in 1 patient and hematuria in 3 patients. Two patients were asymptomatic. All 8 patients received both plain and dynamic contrast-enhanced CT scanning.

Results

A total of 8 masses were detected in the 8 patients. The tumors ranged in diameter from 5 cm to 14 cm with an average of 7.5 cm on CT images. Of those, 4 masses were located in the right kidney and 4 masses were located in the left kidney. Seven lesions were in the periphery of the kidney, including 4 ill-demarcated lesions and 3 well-demarcated lesions. All eight patients underwent radical nephrectomy. The gross specimens showed that 6 lesions arose from renal parenchyma, 1 lesion from the capsule, and 1 lesion from the pelvis. Of 6 parenchyma in origin masses, 3 masses were well-

Clinical study of renal leiomyosarcoma with CT

Table 1. Clinical information about the 8 patients' cases

No.	Age	Male/Female	Right/Left	Tumor diameter (cm)	Clinical features	Stage (AJCC-2013)	Lesion site	Treatment	Survival (months)
1	37	Female	R	7.5	Severe right back pain	T2aNOMO G2 Stage	Renal parenchyma	Radical nephrectomy	38
2	71	Male	L	5	Asymptomatic	T1bNOMO G3	Renal parenchyma	Radical nephrectomy	19
3	40	Male	Rt	10	Right lower back pain, and blood in urine	T2aNOMO G3 Stage	Capsule	Radical nephrectomy	42
4	58	Female	L	7.5	Left back pain	T2aNOMO G3	Pelvis	Radical nephrectomy	17
5	69	Male	L	13.5	Left back pain and losing weight	T2Bn1M0 G3	Renal parenchyma	Radical nephrectomy	3
6	37	Male	L	5.5	Asymptomatic	T1bNOMO G3	Renal parenchyma	Radical nephrectomy	23
7	42	Male	R	6.5	Right lower back pain, and blood in urine	T1bNOMO G3	Renal parenchyma	Radical nephrectomy	9
8	61	Female	R	6	Right abdomen swelling and pain	T1bNOMO G3	Renal parenchyma	Radical nephrectomy	Still alive after 29 months



Figure 1. A 37 year old woman with sudden severe right back pain. A. Plain CT image shows an ill-demarcated mass (arrow) with hyper-attenuation hemorrhage (arrowhead) in the upper pole of the right kidney. B. Contrast-enhanced CT image shows heterogeneous enhancement (arrow) of the mass, which is less than that of the renal cortex of the right kidney the corticomedullary phase. C. Photomicrograph of the histologic specimen (HE 200×) showed spindle cells with mitotic figures.

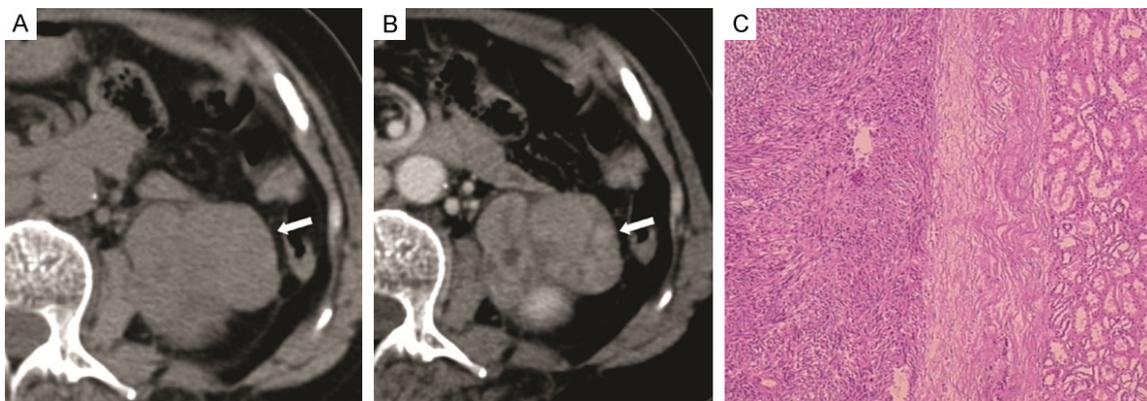


Figure 2. One male patient at his 71 years of age with an asymptomatic symptom was detected by ultrasound at a routine healthcare examination. A. Plain CT image showed a hypertension mass in the inferior pole of the left kidney (arrow). B. CT image at the nephrographic phase shows mild heterogeneous enhancement of the mass (arrow). C. Using histochemical staining, we are able to view the spindle cells with mitotic figures.

circumscribed and 3 masses were ill-defined with surrounding parenchyma. All these information are shown in **Table 1**.

Figure 1 illustrated a representative case of the ill-demarcated lesion. This is the case from a 37 years old female, who experienced severe pain in right back. Plan CT image diagnosis illustrated the ill an ill-demarcated mass as indicated in the arrow in **Figure 1A**. The arrow-head in **Figure 1A** shows hyper attenuation hemorrhage in the upper pole of the right kidney. To further illustrate the pathological information, contrast-enhanced CT was used for diagnosis (**Figure 1B**). Contrast-enhanced CT images illustrated heterogeneous enhancement of the mass, as indicated by the arrow, which is less than that of the renal cortex of the left kidney in the corticomedullary phase (**Figure 1B**). Using histochemical staining, the spindle cells could be viewed with mitotic fig-

ures. Additionally, tumor emboli were also visible in the histologic specimen, as indicated by the arrow in **Figure 1C**.

In another representative case, a 71 years old male was diagnosed with a left renal mass by ultrasound imaging in a routine physical examination. Plane CT images indicated a hyper attenuation mass in the inferior pole of the left kidney, as indicated by the arrow in **Figure 2A**. Enhanced contrast CT image at the nephrographic phase shows mild heterogeneous enhancement of the mass (**Figure 2B**, arrow indicated site). Using histochemical staining, the spindle cells could be viewed with mitotic figures (**Figure 2C**).

In the third case, a 40 year old man was found with hematuria. Enhanced CT image at the corticomedullary phase shows a well-demarcated mass (**Figure 3A**, arrow) with heterogeneous

Clinical study of renal leiomyosarcoma with CT



Figure 3. A 40 year old man with right lower back pain, and blood in urine, and was detected with the disease. A. Plain CT image showed the mass in the inferior pole of the right kidney (arrow). B. Enhanced CT image at the cortico-medullary phase shows a well-demarcated mass with heterogeneous enhancement and areas of low density in the sinus of the right kidney. C. Histochemical staining showed the spindle cells with mitotic figures.



Figure 4. A 58 year old female with sudden left back pain. A. Plain CT image shows an ill-demarcated left renal mass (arrow). B. Perinephric high dense hemorrhage is noted (circle). C. Histochemical staining showed irregular cell progression.



Figure 5. A 69 year old man with the symptoms of left back pain and weight loss. A. Plain CT image reveals a well-demarcated mass (arrow) in the inferior pole of the left kidney. B. CT image at the corticomedullary phase shows slight heterogeneous enhancement of the mass. C. Histochemical staining showed irregular cell division and progression.

enhancement and areas of low dense (stars) in the sinus of the right kidney. After intravenous contrast materials and under enhanced CT imaging, the low-density area sinus of the right kidney was visible as well, as indicated by the star labels in **Figure 3A**. Plain CT image also showed an ill-demarcated right renal mass in **Figure 3A** (arrowhead labeled). The patient experienced sudden left back pain, wherein the following diagnosis of hemorrhage was seen in the lesion and spontaneous rupture was noted in one tumor with perinephric high dense hem-

orrhage (**Figure 3B**). Additionally, Perinephric high dense hemorrhage was also visualized as indicated by the arrowhead in **Figure 3B**. The histochemical staining enabled us to view the spindle cells with mitotic figures (**Figure 3C**).

In a female case, a 58 year old woman was found with the disease with left abdomen swelling and pain. Using plain CT, a well-demarcated mass was observed in the inferior pole of the left kidney, as indicated by the arrow in **Figure 4A**. After intravenous contrast materials, the

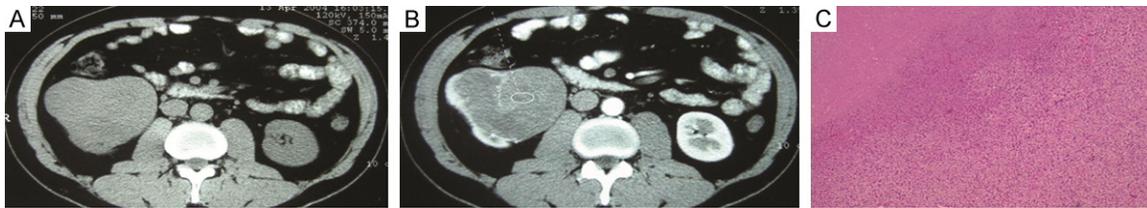


Figure 6. An asymptomatic 37 year old man was detected with the disease in a routine examination. A. Plain CT image illustrated a suspicious demarcated mass in the inferior pole of the left kidney. B. Enhanced CT image at the corticomedullary phase shows an irregular mass in the circled area. C. Histochemical staining showed unusual cell shape with a progressive division.



Figure 7. A 42 year old male had the symptoms of right lower back pain and blood in the urine. He was found with the disease in a routine examination. A. Plain CT image illustrates a well-demarcated mass in the inferior pole of the right kidney (arrowhead). B. Enhanced CT image at the corticomedullary phase shows very irregular mass in the arrow-labeled area. C. Histochemical staining showed unusual aggressive cell division and aggregation in the arrow-labeled area.

tumors showed mild heterogeneous enhancement at the corticomedullary phase, which was hypodense compared to the renal cortex. Under this condition, continuously heterogeneous enhancement in the irregular low-density area was also visible at the excretory phase **Figure 4B** (Circle labeled area). Under H&E staining, images showed tumor spreading in the tissues, where a portion of the tumor cells was dividing (**Figure 4C**). In the 5th case, a 69 year old male was observed with a tumor in the left kidney. Under plain CT scanning, a lesion was observed under the kidney membrane (**Figure 5A**). After intravenous contrast materials, the uneven kidney structure could be observed more clearly (**Figure 5B**). In the H&E staining, tumor cells were aligned in a parallel form (**Figure 5C**).

Using plain CT, enhanced CT and HE staining, we examined another 3 cases of patients. The 6th case was observed in a 37 year old male. This patient was observed with a solid tumor structure in the left kidney and the tissue had an even density and clear structure on the edge (**Figure 6A**). Under enhanced CT image, more tumor structure can be observed. Especially,

the dead vesicles can be observed as well (**Figure 6B**). In the H&E staining, the tumor cells spread with irregular shapes. The division of the nucleus can be observed as well (**Figure 6C**). In the 7th case, a 42-year old male was observed with a high dense tumor structure with clear boundary (**Figure 7A**). Using enhanced CT image, detailed information of the tumor could be observed (**Figure 7B**). In H&E staining, the spread of tumor cells was obvious, where some nucleus division was observable as well (**Figure 7C**). In the 8th patient, tumor tissue was observed on the smooth muscle of the right kidney. The tumor tissue had a structure resembling a tree leaf (**Figure 8A**). In the enhanced CT image, even more tumor structure (**Figure 8B**) could be observed. Similarly, H&E staining revealed an irregular distribution of tumor cells, where the cell shape was not like normal cells and nucleus division was visible as well (**Figure 8C**).

Discussion

Primary leiomyosarcoma of the kidney most likely arises from either intrarenal blood ves-

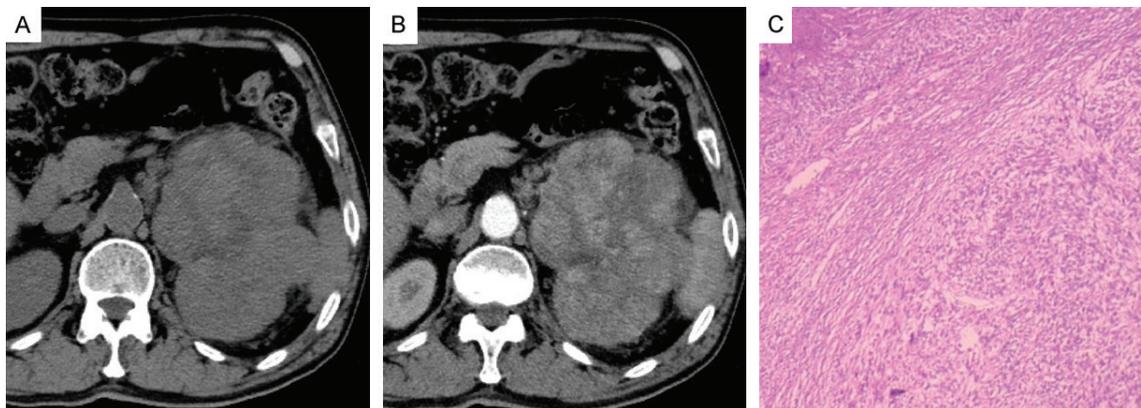


Figure 8. A 61 year old female had right abdomen swelling and pain and was diagnosed with the disease in a routine examination. A. Plain CT image shows a well-demarcated mass in the inferior pole of the right kidney (arrowhead). B. An irregular mass was observed in the arrow-labeled area under enhanced CT image at the corticomedullary phase. C. Histochemical staining showed irregular division and division and shape.

sels or muscle fibers of the renal pelvis [4]. Most patients are 40 to 70 years old without gender preponderance [17]. Symptoms, including flank pain, an abdominal mass, and hematuria, are nonspecific. The most common presenting sign is an abdominal mass with or without pain and hematuria similar to renal cell carcinoma. It is very aggressive with the poor clinical outcome. Radical nephrectomy followed by either chemotherapy or radiation is the standard treatment, but the majority of patients develop metastatic disease [18-20]. Therefore, even though it is very rare, radiologists should take it into differential diagnosis when a renal mass is detected. Familiarity with its imaging characteristics may permit radiologists to consider the possibility of renal leiomyosarcoma before the operation, but there were only several case reports about radiological findings of renal leiomyosarcoma and its imaging characteristics have not been well documented. To the best of our knowledge, the present study, which consists of 8 cases, is the largest series to focus on CT findings of renal leiomyosarcoma.

According to its origin, a renal leiomyosarcoma may envelop the kidney, replace large portions of parenchyma with extension into the perinephric and/or renal sinus fat, or originate from pelvic to extend into the collecting system and involve parenchyma [17]. In the present studies, 6 lesions were included originating from parenchyma, 1 capsule in origin lesion and 1 pelvic in origin lesion. The margins of renal leiomyosarcoma may be well- or ill-demarcated. Even though 6 masses were homogeneous and

only 2 masses were heterogeneous at the plain scan, irregular hypoattenuation areas can be seen after intravenous contrast materials in 7 tumors. Another CT characteristic of renal leiomyosarcoma was mild enhancement at the corticomedullary phase and continuous increasing enhancement of solid components at the nephrographic phase, which was revealed in 7 cases of present series. The level of enhancement at the nephrographic phase was lower than that at the corticomedullary phase only in one case. The characteristic of delay enhancement was thought to be due to abundant fibrous tissue [11]. Since renal cell carcinoma is the most common solid fat-free mass, the differential diagnosis of renal cell carcinoma should include any fat-free enhanced renal mass detected by CT or MRI. Both renal cell carcinoma and leiomyosarcoma are aggressive masses with heterogeneous enhancement, unlike delay enhancement of leiomyosarcoma, conventional renal cell carcinoma often shows strong enhancement equal to the renal cortex at the corticomedullary phase and fast wash-out at the excretory phase [19].

Another entity should be taken into differential diagnosis is the benign counterpart of leiomyosarcoma, leiomyoma, which shows delay enhancement also. Leiomyoma often appears as a well-defined mass with homogeneous density and enhancement [21]. For the overlap of manifestations, the final diagnosis depends on histological examinations.

This retrospective study including small samples is the main drawback. Since renal leiomyo-

sarcoma is a rare neoplasm, these cases were collected for more than ten years. Therefore, scanning protocol could not be uniform due to different CT scanners and it is impossible to calculate CT value changes at different phases in all cases for only hard copies could be found in several cases.

Conclusion

In summary, CT diagnoses of 8 cases of primary renal leiomyosarcoma in our clinical practice in the past 8 years were presented in this study. These cases illustrate mild enhancement at the corticomedullary phase, as well as a continuous enhancement at the nephrographic or the excretory phase. The characteristics in these cases could help radiologists to diagnose leiomyosarcoma at the different stages in the clinic.

Disclosure of conflict of interest

None.

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References

- [1] Cocuzza M, Arap S, Lucon AM and Saldanha LB. Renal leiomyosarcoma treated with partial nephrectomy. *Clinics (Sao Paulo)* 2005; 60: 345-346.
- [2] Grignon DJ, Ayala AG, Ro JY, el-Naggar A and Papadopoulos NJ. Primary sarcomas of the kidney. A clinicopathologic and DNA flow cytometric study of 17 cases. *Cancer* 1990; 65: 1611-1618.
- [3] Kendal WS. The comparative survival of renal leiomyosarcoma. *Can J Urol* 2007; 14: 3435-3442.
- [4] Moudouni SM, En-Nia I, Rioux-Leclercq N, Guille F and Lobel B. Leiomyosarcoma of the renal pelvis. *Scand J Urol Nephrol* 2001; 35: 425-427.
- [5] Kaphan E, Eusebio A, Witjas T, Donnet A, Vacher-Coponat H, Figarella-Branger D and Cherif AA. Primary leiomyosarcoma of the cavernous sinus associated with Epstein-Barr virus in a kidney graft. *Rev Neurol* 2003; 159: 1055-1059.
- [6] El Otmany A, Bouklata S, Chami I, Souadka A, Jahid A and Belabbas M. Primary leiomyosarcoma of the kidney. A case report. *Prog Urol* 1999; 9: 310-312.
- [7] Akkaya B, Tuncer SS, Gurer IE and Baykara M. Primary leiomyosarcoma of the kidney: three cases. *Virchows Archiv* 2012; 461: S303-S303.
- [8] Miller JS, Zhou M, Brimo F, Guo CC and Epstein JI. Primary leiomyosarcoma of the kidney: a clinicopathologic study of 27 cases. *Modern Pathology* 2010; 23: 207a-207a.
- [9] Van Rooij WJ, Martens F, Verbeeten B Jr and Dijkstra J. CT and MR imaging of leiomyosarcoma of the inferior vena-cava. *J Comput Assist Tomogr* 1988; 12: 415-419.
- [10] Rakowsky E, Barzilay J, Schujman E and Servadio C. Leiomyosarcoma of kidney. *Urology* 1987; 29: 68-70.
- [11] Ochiai K, Onitsuka H, Honda H, Kawamoto K, Uozumi J, Kumazawa J and Masuda K. Leiomyosarcoma of the kidney: CT and MR appearance. *J Comput Assist Tomogr* 1993; 17: 656-658.
- [12] Martin J, Garcia M, Duran A, Forcada P and Marco V. Renal vein leiomyosarcoma: a case-report and literature-review. *Urol Radiol* 1989; 11: 25-29.
- [13] Lipton M, Sprayregen S, Kutcher R and Frost A. Venous invasion in renal vein leiomyosarcoma: case-report and review of the literature. *Abdom Imaging* 1995; 20: 64-67.
- [14] Kaushik S and Neifeld JP. Leiomyosarcoma of the renal vein: imaging and surgical reconstruction. *AJR Am J Roentgenol* 2002; 179: 276-277.
- [15] Farah MC, Shirkhoda A, Ellwood RA, Bernacki E and Farah J. Leiomyosarcoma of the renal vein: radiologic pathologic correlation. *Clin Imaging* 1989; 13: 323-326.
- [16] Chung YG, Kang SC, Yoon SM, Han JY and Seong DH. Leiomyosarcoma arising from the blind end of a bifid renal pelvis. *Yonsei Med J* 2007; 48: 557-560.
- [17] Katabathina VS, Vikram R, Nagar AM, Tamboli P, Menias CO and Prasad SR. Mesenchymal neoplasms of the kidney in adults: imaging spectrum with radiologic-pathologic correlation. *Radiographics* 2010; 30: 1525-40.
- [18] Miller JS, Zhou M, Brimo F, Guo CC and Epstein JI. Primary leiomyosarcoma of the kidney: a clinicopathologic study of 27 cases. *Am J Surg Pathol* 2010; 34: 238-242.
- [19] Kim JK, Kim TK, Ahn HJ, Kim CS, Kim KR and Cho KS. Differentiation of subtypes of renal cell carcinoma on helical CT scans. *AJR Am J Roentgenol* 2002; 178: 1499-506.
- [20] Deyrup AT, Montgomery E and Fisher C. Leiomyosarcoma of the kidney: a clinicopathologic study. *Laboratory Investigation* 2003; 83: 147a.
- [21] Cong Z, Gong J and Wang Y. Radiological findings of renal leiomyomas: a report of five surgically confirmed cases. *Abdom Imaging* 2011; 36: 604-608.