

Leiomyosarcoma of the Renal Pelvis: Report of a Case and Review of the Literature

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Abstract

We report a case of leiomyosarcoma of the renal pelvis developed in 56-year-old man whom underwent MDCT scan presented with hematuria and left flank pain. There was a 7.8×7.6 cm heterogeneous enhanced mass filling in the left renal pelvis with necrotic portion and multiple dystrophic calcifications. The tumor measured about 6.8×5.5 cm in size composed of atypical spindle cells arising from the muscularis mucosa of the renal pelvis. The tumor cells were positive for smooth muscle actin, focal positive for desmin and myoglobin but negative for CD 117 (c-KIT), pancytokeratin and CD34. The histopathologic diagnosis was made as a leiomyosarcoma of the renal pelvis.

Keywords: Leiomyosarcoma; Kidney tumor; Renal pelvic tumor; Renal mass

Abbreviation: MDCT: Multi-Detector Computed Tomography; HPF: High-Power Fields; SMA: Smooth Muscle Actin; RCC: Renal Cell Carcinoma; TCC: Transitional Cell Carcinoma

Introduction

Leiomyosarcoma of the renal pelvis is an extremely rare mesenchymal tumor of the kidney. To the best of our knowledge, there is no article that focused on radiologic and pathologic correlation of the renal pelvic leiomyosarcoma.

Primary renal sarcomas are rare tumors presenting for 1%-3% of the malignant renal tumors. Leiomyosarcoma is the most common subtype of the renal sarcomas accounting for 50%-60% of all renal sarcomas [1]. Most renal leiomyosarcoma arising from the renal capsule and from smooth muscle of the renal pelvis is uncommon [2].

Here, we describe a rare case of leiomyosarcoma of the renal pelvis detected on the Multi-Detector Computed Tomography (MDCT) with pathologic correlation and we review the previous literature.

Case Report

A 56-year-old man with hematuria and left flank pain presented to the Inje University Haeundae Paik Hospital. The patient had an open ureterolithotomy for left renal stone 26 years ago and extracorporeal shock wave lithotripsy for same reason 23 years ago. Laboratory data showed no impairment of renal function (blood urea nitrogen and creatinine levels, 10.4 and 1.11 mg/dL, respectively). Routine urinalysis displayed proteinuria (+3) and hematuria (+3). A MDCT scan was performed for further evaluation. MDCT included an unenhanced phase scan, a nephrographic phase scan, a pyelographic phase scan and three-dimensional reformations with coronal and sagittal maximum intensity projections of the kidney.

Nephrographic and pyelographic phase scans that were followed with a 100 sec delay after attenuation of the aorta at the thoracolumbar junction had reached 100 HU and a fixed 12 minute delay, respectively, after the intravenous injection of 110 mL of iobitridol (Xenetics; Guerbet, Paris, France) administered at a rate of 3 mL/s with an automatic injector.

MDCT scan demonstrated a 7×6 cm solid and cystic mass with calcifications filling in the left renal pelvis resulting in hydronephrosis but preserved the reniform. The enhancement pattern is heterogeneous

and gradual; diffuse gradual enhancement and some portions showed relatively low attenuation (Figures 1A-1C). Peripheral to the solid portion of the tumor cystic portions were present, intracystic attenuation showed heterogeneous and high attenuation than that of dilated renal calices (Figure 1D). Multiple calcifications of variable size and shape were at both solid component and cystic component. The tumor located in renal pelvis and inferior portion of the kidney, thinning of renal cortex and focal obliteration of thinned renal cortex with perinephric fat infiltration was noted. There was no mass at contralateral kidney, both ureters and urinary bladder. There was no enlarged lymph node at the regional or distant area. Preoperative differential diagnosis was urothelial cell carcinoma such as transitional cell carcinoma, squamous cell carcinoma or mucinous adenocarcinoma considering prevalence and patient's history.



Figure 1a: 56-year-old man with hematuria and left flank pain. (A) MDCT scan axial image without contrast.

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Figure 1b: Nephrographic phase.



Figure 1c: Pyelographic phase demonstrate a 7×6 cm-sized cystic and solid intraluminal mass in the renal pelvis of the left kidney with calcifications. The enhancement pattern is heterogeneously and gradually mild enhancement with internal low attenuated necrotic portions.



Figure 1d: On coronal reconstructed scan of nephrographic phase, hydrocalicex in upper portion of the left kidney and lower cystic portion showing slightly delayed enhancement at inferior cystic and necrotic portion with preserved reniform (arrows).

Laparoscopic left nephroureterectomy was performed. Grossly the tumor was measured 6.8×5.5 cm identified in lower portion of the kidney (Figure 2A). The cut surface was grayish white solid. Microscopically, the tumor was composed of atypical spindle cells that were arranged as alternating fascicles (Figure 2B). The mass originating from the muscularis mucosa layer of renal pelvis contained dystrophic calcifications and necrosis with invasion of renal parenchyma and perinephric fat tissue (Figures 2C and 2D) with underlying chronic pyelonephritis. The number of mitotic activity was 19 mitoses per 10 High-Power Fields (HPF) on average. Immunohistochemically, the tumor cells were positive for Smooth Muscle Actin (SMA) (Figure 3A), and negative for pancytokeratin thus sarcomatoid carcinoma could be ruled out (Figure 3B). Additional immune histochemistry for vimentin

was positive, desmin and myoglobin were positive and CD 34 and EMA were negative. Based on the histopathology and immunohistochemistry, the mass was diagnosed as leiomyosarcoma arising from the renal pelvis. A pulmonary nodule was newly developed on 2 months follow up chest CT and PET CT.

Discussion

Leiomyosarcoma from the renal pelvis is a rare disease entity. 95% of the malignant renal pelvic masses are urothelial cell origin tumor. Only 1-3% accounts for renal sarcomas, and leiomyosarcoma is the most common sarcoma of the kidney accounting for 50-60% of cases. Renal leiomyosarcoma mostly occur in adults and no male



Figure 2a: Gross specimen of large mass at inferior pole of the kidney with central cystic change.

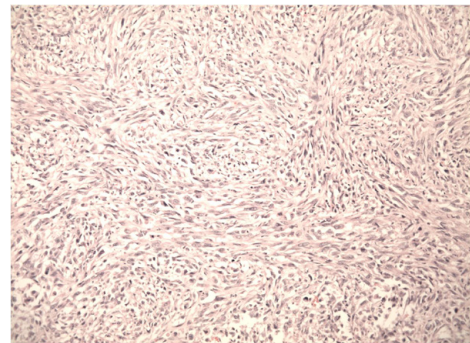


Figure 2b: On histopathology (Hematoxylin and eosin, 200 x), the tumor is composed of atypical bland-ended spindle cells with alternating fascicular pattern.

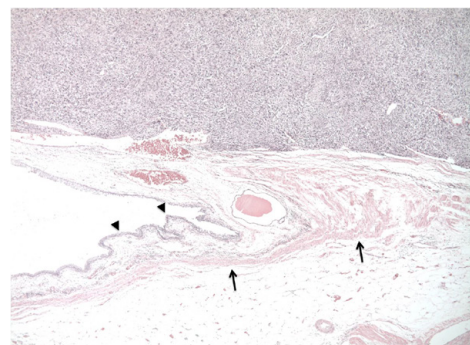


Figure 2c: The arising from muscularis mucosa of the renal pelvis (arrows) and remained normal uroepithelium (arrowheads) (Hematoxylin and eosin, 40 x).

No.	Age (yr)	Gender	Clinical symptom	Size (cm)	Retrograde pyelography or IVP	CT findings	Preoperative diagnosis	Treatment	Follow-up	Reference	Published
1	24	F	hematuria	(-)	Filling defect	(-)	Papillary tumor, radiolucent calculus	Right radical nephroureterectomy	(-)	[4]	1971
2	23	F	LLQ pain	6	Filling defect	(-)	No data	Left nephrectomy	Alive (1yr)	[5]	1972
3	61	F	Right flank pain, hematuria	6	Filling defect, hydronephrosis	(-)	Right renal pelvic tumor	Right radical nephrectomy, subtotal ureterectomy	Expire (2 mo, presumably caused by pancreatic cancer)	[2]	1973
4	65	M	Hematuria, general weakness	3	Filling defect, hydronephrosis	Poorly enhance, filling in renal pelvis	No data	Left nephroureterectomy, bladder cuff excision	Alive (18 mo)	[6]	1997
5	41	M	Left flank pain, hematuria	2	(-)	Solid mass filling renal pelvis	Transitional cell carcinoma	Left nephroureterectomy, bladder cuff excision	Alive (8yr)	[7]	2001
6	54	M	Bowel discomfort	8	(-)	(nonenhanced CT) Solid mass filling renal pelvis	Renal cell carcinoma	Left nephrectomy	Alive (6mo)	[8]	2004
7	44	M	Hematuria	5	(-)	Soft tissue mass occupying the collecting system	Transitional cell carcinoma	Left radical nephroureterectomy	Alive (3yr)	[9]	2006
8	42	F	incidental	5	Bifid left renal pelvis	Heterogeneous enhancement, solid mass	No data	nephrectomy	(-)	[10]	2007
9	60	M	Urinary frequency	10	(-)	Heterogeneous enhancement, solid mass	Renal cell carcinoma	Right nephrectomy	(-)	[11]	2009
10	56	M	Hematuria, left flank pain	7	(-)	Solid mass in renal pelvis, Heterogeneous gradual enhancement, cystic portion, calcifications	Urothelial cell carcinoma	Left nephroureterectomy		Present case	

Table 1: Clinico-radiologic features of 9 reported and present case of leiomyosarcoma arising from renal pelvis.

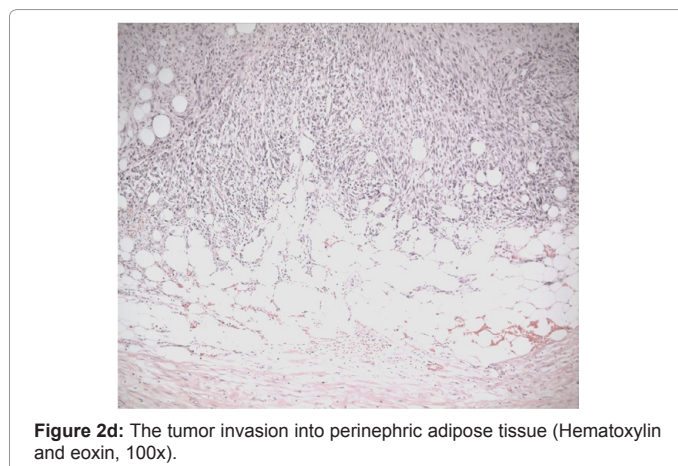


Figure 2d: The tumor invasion into perinephric adipose tissue (Hematoxylin and eosin, 100x).

and female are equally affected. Symptoms are non-specific; include flank pain, palpable mass and hematuria not much different from other renal tumors. Usually metastasize to lung, liver and bone [3]. Leiomyosarcoma may arise from the renal capsule, renal parenchyma, pelvic muscularis, or the main renal vein [3]. Macroscopically, large solid grey-white, soft to firm mass and it may have focally necrotic areas [3]. Renal leiomyosarcoma have no pathognomonic finding that lead to make preoperative diagnosis. In most of the cases, diagnosis of renal leiomyosarcoma would be made after surgical resection and histopathologic examination.

The Table summarizes the clinico-radiological features of the previously reported cases in English literature including the present case. To date, there is no sex predilection, and occurred at wide range of age (23-65 years, mean; 47 years). Renal pelvic leiomyosarcoma ranges in size from 2 to 10 cm with a mean of 5.7 cm. A common characteristic feature is filling defect of renal pelvis on retrograde pyelography or intravenous pyelography (Table 1) and solid mass filling in the renal pelvis on contrast enhanced CT (Table 1). This means intraluminal growth pattern of the tumor. Image findings and histopathologic specimen showed intraluminal growth pattern of the tumor more than outward growth pattern. But the tumor infiltrates to perirenal adipose tissue and renal sinus as well as renal parenchyma. Among 6 cases performed contrast enhanced CT, poor (Table 1) and heterogeneous (Table 1) enhancement patterns were shown after contrast administration. Only one case had reported focal necrosis of the tumor (Table 1) as in our case. Furthermore, none of the case reported intratumoral dystrophic calcifications which were found in our case. Our pathologist suggested for dystrophic calcifications rather than renal calculi. There was no reported lymph node metastasis in all cases.

Renal leiomyosarcoma was known to have poor prognosis, with a 5 year survival rate of 29-36%. However, in reported renal pelvic leiomyosarcoma cases, the follow-up duration was too short and the number of reported cases is small to precisely predict the prognosis.

In 5 cases, preoperative radiologic diagnosis was urothelial cell carcinoma (Table 1) or Renal Cell Carcinoma (RCC) (Table 1). Possible

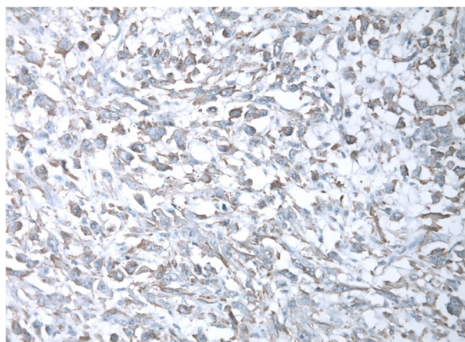


Figure 3a: Immunohistochemically, the tumor cells are positive for smooth muscle actin (400 x).

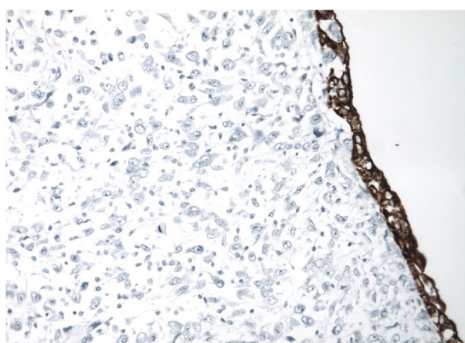


Figure 3b: The tumor cells are negative for pancytokeratin (x 400).

cause of radiologic misdiagnosis is the low prevalence rate of renal pelvic leiomyosarcoma.

The majority malignant tumor arising from renal pelvis is urothelial cell carcinoma including Transitional Cell Carcinoma (TCC), squamous cell carcinoma and mucinous adenocarcinoma. Among them TCC is the most common urothelial neoplasm hematuria as common clinical presentation and well known as multiplicity. TCC is papillary intraluminal mass, spreads centrifugally and dilates collecting system [12]. Thus, hydrocalices, amputated calices or phantom calices are shown on excretory urography. On CT scan after contrast injection the mass typically enhances, but lesser degree than normal renal parenchyma [12]. TCC could be noncalcified and calcified. Squamous cell carcinomas and mucinous adenocarcinomas are strongly related with renal calculi and hard to distinguish from TCC [13].

RCCs projecting into renal sinus also should be considered, which also can lead to hydronephrosis or caliceal displacement. However, well enhancement on post contrast CT images will be shown unlike TCCs. In histopathological examination the most important disease to differentiate from leiomyosarcoma is sarcomatoid RCC. Negative for Pancytokeratin immunochemical stain can rule out sarcomatoid RCC.

In conclusion, we report a case of leiomyosarcoma arising from renal pelvis, rarely reported malignant mesenchymal neoplasm of the kidney. CT scan of this rare tumor showed heterogeneously enhancing large mass filling in the renal pelvis with cystic change and calcifications. Leiomyosarcomas have histological characterization

of alternating fascicular haphazard pattern, bland spindle cells and nuclear atypia, pleomorphism, and necrosis. They confirmed by immunohistochemistry, the tumor cells of leiomyosarcoma are positive for SMA and desmin and the tumor cells are negative for pancytokeratin, CD 117 (c-KIT) and CD34. These histological and immunohistochemical features are helpful in diagnosing this uncommon disease. Our report may help increase the awareness of this rare disease entity and take account of differential diagnosis for renal pelvic mass.

Authors Contribution

EY Jo: Clinical study, manuscript editing; JH Yoon: Literature search, manuscript editing; W Kim: Pathologic editing.

References

1. Katabathina VS, Vikram R, Nagar AM, Tamboli P, Menias CO, et al. (2010) Mesenchymal neoplasms of the kidney in adults: imaging spectrum with radiologic-pathologic correlation. *Radiographics* 30: 1525-1540.
2. Tolia BM, Hajdu SI, Whitmore WF Jr (1973) Leiomyosarcoma of the renal pelvis. *J Urol* 109: 974-976.
3. Eble JN, Sauter G, Epstein JI, Sesterhenn IA (2004) Pathology and genetics of tumours of the urinary system and male genital organs. World Health Organization Classification of Tumours, IARC Press, Lyon, France.
4. Litzky GM, Seidel RF, O'Brien JE (1971) Leiomyoma of the renal pelvis. *J Urol* 105: 171-173.
5. Loomis RC (1972) Primary leiomyosarcoma of the kidney: report of a case and review of the literature. *J Urol* 107: 557-560.
6. Chen JH, Lee SK (1997) Renal leiomyosarcoma mimicking transitional cell carcinoma. *AJR Am J Roentgenol* 169: 312-313.
7. Moudouni SM, En-Nia I, Rioux-Leclercq N, Guille F, Lobel B (2001) Leiomyosarcoma of the renal pelvis. *Scand J Urol Nephrol* 35: 425-427.
8. Minami H, Ueki O, Tanaka T, Nishida H, Hashimoto T, et al. (2004) Case of leiomyosarcoma of the renal pelvis. *Int J Urol* 11: 122-124.
9. Kartsanis G, Douros K, Zolota V, Perimenis P (2006) Case report: leiomyosarcoma of the renal pelvis. *Int Urol Nephrol* 38: 211-213.
10. Chung YG, Kang SC, Yoon SM, Han JY, Seong DH (2007) Leiomyosarcoma arising from the blind end of a bifid renal pelvis. *Yonsei Med J* 48: 557-560.
11. Dhamne SA, Gadgil NM, Padmanabhan A (2009) Leiomyosarcoma of the renal pelvis. *Indian J Pathol Microbiol* 52: 549-551.
12. Dyer R, DiSantis DJ, McClennan BL (2008) Simplified imaging approach for evaluation of the solid renal mass in adults. *Radiology* 247: 331-343.
13. Rha SE, Byun JY, Jung SE, Oh SN, Choi YJ, et al. (2004) The renal sinus: pathologic spectrum and multimodality imaging approach. *Radiographics* 24 Suppl 1: S117-S131.