Case Report
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대한영상의학회지

Primary Leiomyoma of Ureter Coexisting with Renal Cell Carcinoma: A Case Report
신세포암과 병발된 요관의 원발성 평활근종: 증례 보고

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Mesenchymal origin of ureter tumors account for less than 3 percent of all primary ureteral tumors. Among mesenchymal tumors, primary leiomyoma of ureter is extremely rare. Here, we present a case of primary leiomyoma of ureter coexisting with renal cell carcinoma. When encountering well-defined homogeneously enhanced mass of ureter on computed tomography, radiologist should keep in mind that ureteral leiomyoma should be considered as differential diagnosis.

Index terms
Leiomyoma
Ureter, Ureter Neoplasm
Computed Tomography

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INTRODUCTION

Leiomyomas are benign tumors of smooth muscle origin. They are characterized by overgrowth of visceral smooth muscle in respiratory, gastrointestinal, and female genital organ with rare development in the urinary tract. Only 13 cases of primary urethral leiomyoma have been reported in the literature (1, 2). Primary ureteral leiomyoma often manifests as a huge mass, causing hydronephrosis. It is difficult to distinguish them from malignant ureteral tumor such as urothelial carcinoma. Therefore, total nephroureterectomy has been performed for most cases due to its differential diagnosis of malignancy. Here, we present a case of a patient with primary leiomyoma of the ureter coexisting with renal cell carcinoma of kidney at the same side.

CASE REPORT

A 50-year-old woman presented with left quadrant lower abdominal pain for one month. The patient did not show other accompanying systemic symptoms. The patient underwent total hysterectomy for the uterine myoma 10 years ago in an outside hospital. Although there was no hematuria, cytological evaluation of urine samples showed a few atypical urothelial cells and non-specific inflammatory cells which were considered to be either reactive urothelial cells or urothelial tumor cell. Dynamic contrast-enhanced computed tomography (CT) was performed for further evaluation of the urinary system. Abdominal CT revealed well-defined small homogeneously enhanced nodular lesion (about 1.4 × 1.1 cm) at the medial aspect of the left distal ureter (Fig. 1A). The periureteral mass was obstructing the left distal ureter accompanied with obstructive hydroureteronephrosis of the left kidney (Fig. 1B). Small round hypervascular mass was seen in the mid-portion of left renal parenchyma. This mass showed hyperenhancement on corticomedullary phase and washout on excretory phase (Fig. 2A, B). It was considered as a renal cell carcinoma. There was no evidence of extracapsular extension or metastases on scanned abdominal CT. Ureteroscopic examination showed luminal narrowing of left distal ureter due to extrinsic pressing into the ureteral lumen. The ureteral surface was covered by normal mucosa. This finding was thought to be a ureteral submucosal tumor rather than a carcinoma of urothelial origin.

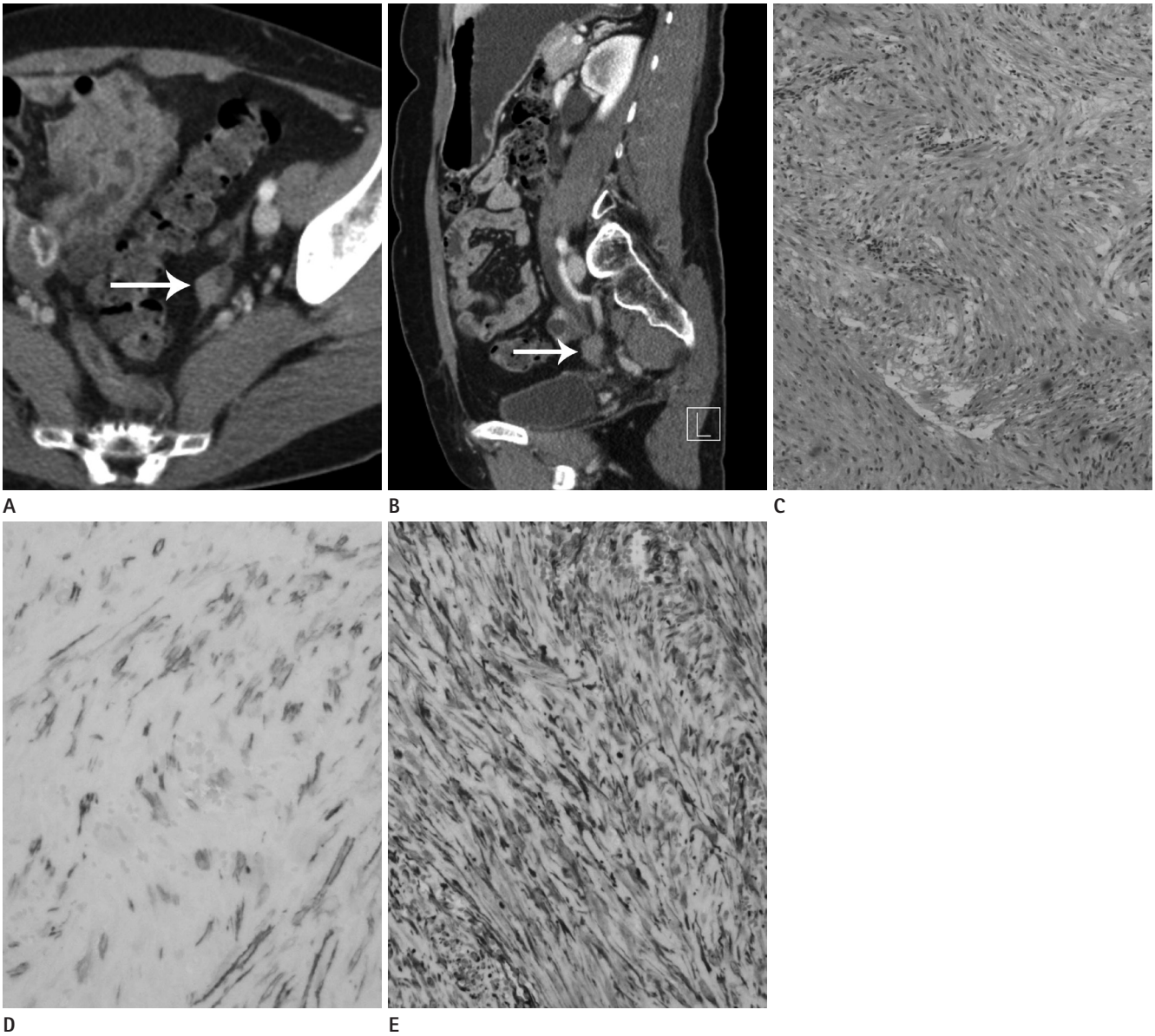
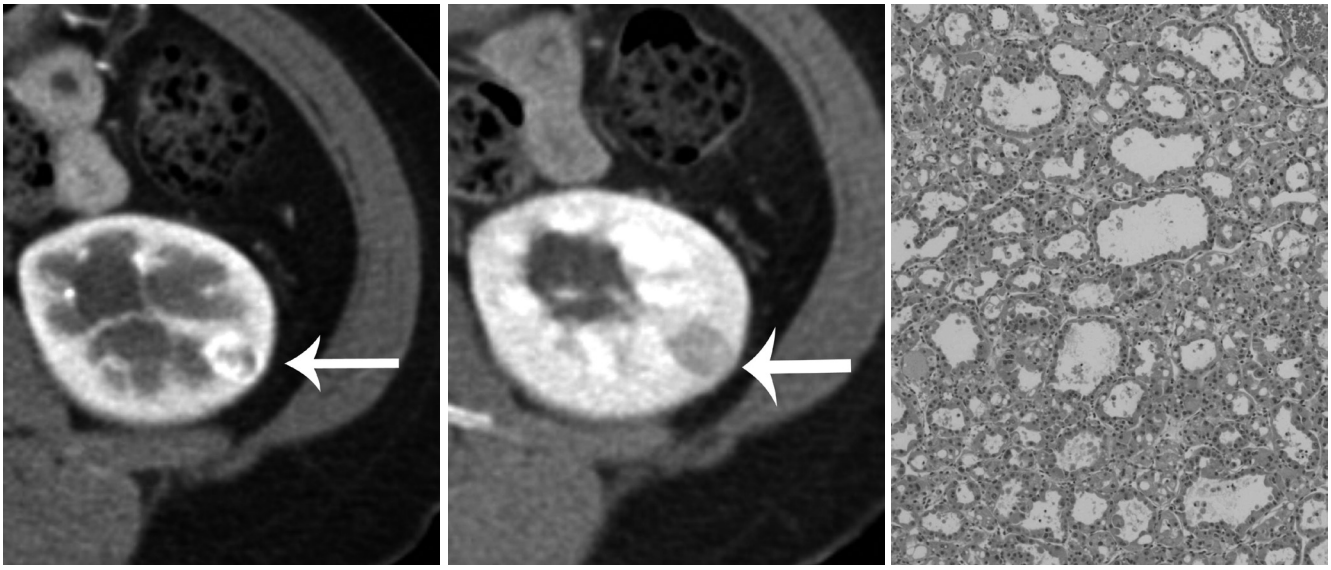


Fig. 1. 50-year-old woman with primary leiomyoma of the ureter.
A. Contrast-enhanced abdomen CT shows well-defined small homogeneously enhancing mass (about 1.4 × 1.1 cm) at the medial aspect of the left distal ureter. The periureteral mass (arrow) was compressing the left distal ureter.
B. Contrast-enhanced abdomen CT shows obstructive hydronephrosis and hydroureter of the left kidney (arrow) and slightly decreased enhancement of renal parenchyma suggesting obstructive uropathy.
C. Microscopic image of the mass shows well-differentiated multiple layers of hypercellular and hyperchromatic interwoven bundles of smooth muscle cells (H&E, × 40).
D, E. On the immunohistochemical staining, the spindle cells were positive for the smooth muscle actin (SMA) **(D)** and vimentin **(E)** (SMA, × 100, vimentin, × 100).

The patient underwent nephroureterectomy to remove the left renal parenchymal mass and the left ureteral mass. Intraoperative inspection of the left ureteral mass revealed the medial aspect of the left iliac vessels. The hard mass was tightly adherent to the surrounding soft tissue. Histopathologic examination of the left renal parenchymal mass was consistent with clear cell renal cell carcinoma (Fig. 2C). Histopathologic findings for the left ureteral mass showed interlacing bundles of eosinophilic spindle cells (Fig. 1C). On immunohistochemical staining, the spindle cells were positive for smooth muscle actin and vimentin (Fig. 1D, E). The left ureteral mass was diagnosed as leiomyoma arised from ureter.



A B C

Fig. 2. Coexistent renal cell carcinoma in the mid-portion of the left kidney.
A, B. Contrast-enhanced abdomen CT with axial scan shows 1.4 × 1.3 cm sized hypervascular mass (arrow) in the mid-portion of left renal parenchyma. This mass showed hyperenhancement on corticomedullary phase **(A)** and washout of enhancement on excretory phase **(B)**. It was considered as a renal cell carcinoma.
C. Micrograph shows epithelial cells with clear cytoplasm and a distinct cell membrane, separated by a delicate branching network of vascular tissue (H&E, × 100). It is typical histologic findings of renal clear-cell carcinoma.

DISCUSSION

Primary tumor of the ureter is relatively rare. Among ureteral tumors, malignant tumor such as transitional cell carcinoma is the most commonly observed type. Benign ureteral tumor of mesodermal origin including leiomyoma is extremely rare compared to urothelial tumor (3). Leiomyoma of the ureter accounts for less than 3% of all primary tumors of the ureter. The exact mechanism of leiomyoma of the ureter is unclear. However, ureteral obstruction due to ureterolithiasis, inflammation, chronic irritation, and trauma are suspected factors (4, 5). Among them, history of ureterolithiasis was the most common suspected causes. However, in our case, the patient had no history of ureterolithiasis.

Hereditary leiomyomatosis and renal cell cancer (HLRCC) is characterized by cutaneous leiomyomata, uterine leiomyoma, and/or a single renal tumor (6). HLRCC is known to be caused by a mutation in fumarate hydratase gene, leading to an accumulation of fumarate. With some incomplete genetic penetration, uterine leiomyoma and renal cell carcinoma could be observed clinically. Although we did not perform genetic evaluation, some genetic defect might be considered as a predisposing factor in the patient's uterine leiomyoma, ureteral leiomyoma, and renal cell carcinoma.

It is very difficult to diagnose leiomyoma of the ureter. Various tests such as excretory urography, retrograde urography, magnetic resonance image (MRI) or CT, and urine cytology could be helpful. Treatment of ureteral leiomyoma includes surgical excision or ablation. Our patient underwent a nephroureterectomy to exclude primary ureteral malignancy and to remove renal cell carcinoma of the left kidney.

To the best of our knowledge, only thirteen cases about leiomyoma of the ureter have been reported in literature. Including our case, patient's age showed distribution from 24 to 60 years old except for one infant. Lesions in eight cases were located at the left side, with six at the right side. Eight cases involved men whereas six involved women. There was no significant difference in the location of the lesion, sex, or site (5). Surgery was performed in 13 out of 14 cases. Surgeries were performed for most cases without differential diagnosis of benign or malignant tumor. Endoscopic biopsy was performed for only one case (7).

The radiological feature of leiomyoma on the CT is the well-defined round enhanced mass (7). Except this feature, no other characteristic findings may be encountered. Radiological features of leiomyoma of the ureter have not reported yet. In this

case, the well-defined homogeneously enhanced mass was observed on CT findings. However, it was difficult to diagnose the leiomyoma of the ureter initially due to its low prevalence rate in the ureter.

The most commonly encountered mass on the ureter is the urothelial cell carcinoma. It is difficult to differentiate urothelial cell carcinoma from leiomyoma based on image findings. However, urothelial cell carcinoma seems to have strong ureter wall enhancement and infiltrative features around periureteral soft tissue and ureter wall, whereas leiomyoma of the ureter seems to have well-defined homogeneously enhanced mass rather than strong enhancement of ureter wall or infiltrative feature (8).

Leiomyomas of genitourinary tract are rare. Ureter is an uncommon site for them. Ureteral leiomyoma often causes hydronephrosis, making diagnosis difficult (5). When encountering well-defined homogeneously enhanced mass of the ureter on CT, radiologist should keep in mind that rare ureteral leiomyoma should be considered as a differential diagnosis.

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신세포암과 병발된 요관의 원발성 평활근종: 증례 보고

백승환 · 김희진 · 한현영

요관에 발생하는 중배엽 기원의 종양은 전체 원발성 요관 종양의 3% 미만의 매우 드문 종양이다. 또한 그 중에서 요관 기원의 원발성 평활근종은 극히 드문 것으로 알려져 있다. 저자들은 신세포암과 동반된 요관의 원발성 평활근종 1예를 경험하고 이를 보고한다. 복부 전산화단층촬영에서 요관의 균질한 조영증강을 보이며 경계가 좋은 종양이 있다면 감별진단으로 요관의 평활근종을 고려해야겠다.

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