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Leiomyosarcoma of the renal pelvis diagnosed by percutaneous endoscopic resection

Manint Usawachintachit [a,*], Julin Opanuraks [a], Jerasit Surintrspanont [b], Kroonpong Iampenkhae [b], Apirak Santingamkun [a]

[a] Division of Urology, Department of Surgery, Faculty of Medicine, Chulalongkorn University, King Chulalongkorn Memorial Hospital, The Thai Red Cross Society, Bangkok, 10330, Thailand

[b] Department of Pathology, King Chulalongkorn Memorial Hospital, The Thai Red Cross Society, Bangkok, 10330, Thailand

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ABSTRACT

We present a case of leiomyosarcoma arising from the renal pelvis, which is a rare clinical entity. A percutaneous endoscopic resection led to the final histopathological diagnosis. The patient underwent radical nephrectomy and did not receive adjuvant therapy. Based on follow-up CT scans, he remains recurrence-free one year after surgery.

1. Introduction

Primary tumors of the renal pelvis are mostly urothelial in origin. Only a handful of other tumor subtypes, such as leiomyosarcoma, have been reported.[1] Herein, we report a case of renal pelvis leiomyosarcoma and discuss the diagnosis and management.

2. Case presentation

A 28-year-old Thai man initially presented to our hospital with left flank pain and gross hematuria without blood clot. He had a history of left renal stone and underwent three sessions of shockwave lithotripsy with temporary ureteral stent placement elsewhere during the last two years. He never smoked; his past medical history was clear; and the physical examination was normal. Computed tomography urography (CTU) revealed an 8-mm left lower caliceal stone, a 3-mm left distal ureteral stone, and a moderate degree of left hydronephrosis. The contrast-enhanced phases showed no renal mass or filling defect in the left kidney (Fig. 1A). The patient underwent flexible ureterorenoscopy with laser lithotripsy, which successfully eliminated the stones. He recovered well, and the ureteral stent was removed two weeks afterward.

Ten months later, the patient unexpectedly revisited the hospital with left flank pain and gross hematuria. A repeat CTU revealed a 1.9-cm enhancing lesion at the superior aspect of the renal pelvis with focal dilatation of the upper calyces (Fig. 1B). Although not commonly found at his age, a provisional diagnosis of urothelial carcinoma was suspected. Subsequent flexible ureterorenoscopy revealed a submucosal mass fungating from the anterior aspect of the renal pelvis. The overlying mucosa of this mass appeared slightly irregular without the typical papillary configuration of urothelial carcinoma (Fig. 1C). A few cold cup biopsies revealed acute and chronic inflammation without malignancy in the tissue. Urine cytology obtained from his left renal pelvis demonstrated no atypical cell. Afterward, the patient continued to experience intermittent hematuria, and an interval CTU performed four months later demonstrated a significant growth of the tumor to 2.6 cm, which raised a concern of malignancy (Fig. 2A). Following a discussion at the multidisciplinary team meeting, the patient and his family insisted on obtaining a confirmatory histopathological diagnosis before proceeding to radical surgery. Therefore, we decided to perform percutaneous endoscopic resection of the tumor to obtain more tissue. Following a lower pole puncture, a balloon dilator was applied to accommodate a 30-Fr sheath. The endoscopic view revealed a 3-cm sessile tumor protruding from the superomedial aspect of the renal pelvis (Fig. 2B). A 26-Fr resectoscope with monopolar loop cautery was utilized for tissue resection and coagulation. We were able to resect approximately 50% of the total tumor volume.

Histopathology of the resected tissue revealed spindle-shaped tumor

* Corresponding author. Division of Urology, Department of Surgery, Faculty of Medicine, King Chulalongkorn Memorial Hospital, Chulalongkorn University, The Thai Red Cross Society, Bangkok, 10330, Thailand.

E-mail addresses: manint.u@chula.ac.th (M. Usawachintachit), julin.o@chula.ac.th (J. Opanuraks), jerasit.s@chula.ac.th, srjerasit@hotmail.com (J. Surintrspanont), kroonpong@gmail.com (K. Iampenkhae), apirak99@hotmail.com (A. Santingamkun).

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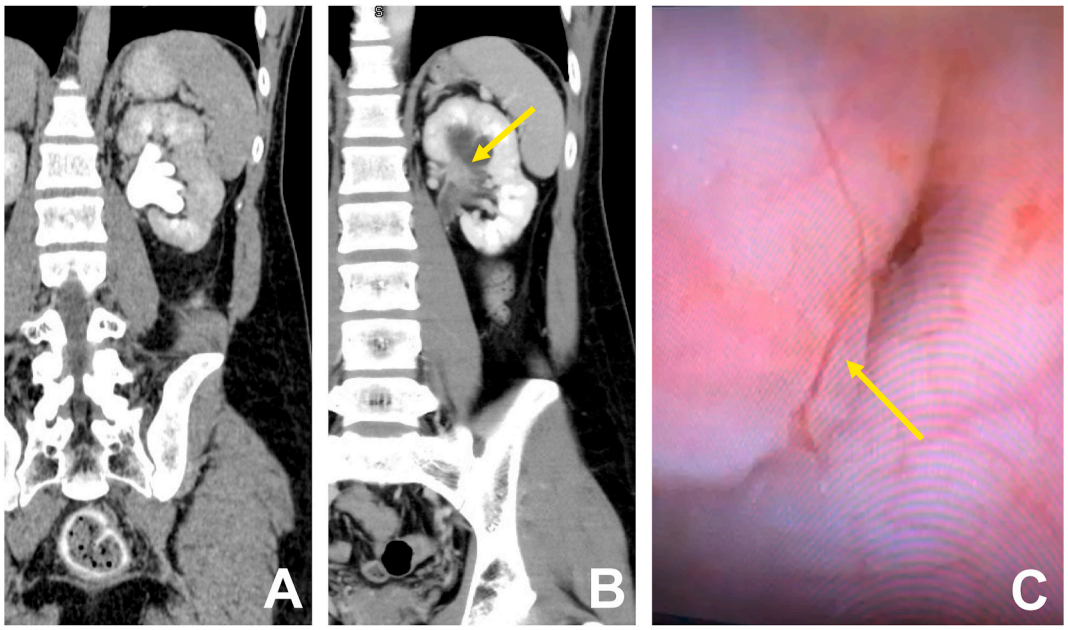

Fig. 1. Initial CT scan; the delayed phase revealed left moderate hydronephrosis without filling defect in the renal pelvis (Fig. 1A). CT scan at the new episode of flank pain and gross hematuria; the venous phase demonstrated a 1.9-cm enhancing mass at the superior aspect of the left renal pelvis (1B). View from flexible ureterorenoscopy showed a submucosal mass corresponding to the CT scan finding (1C).

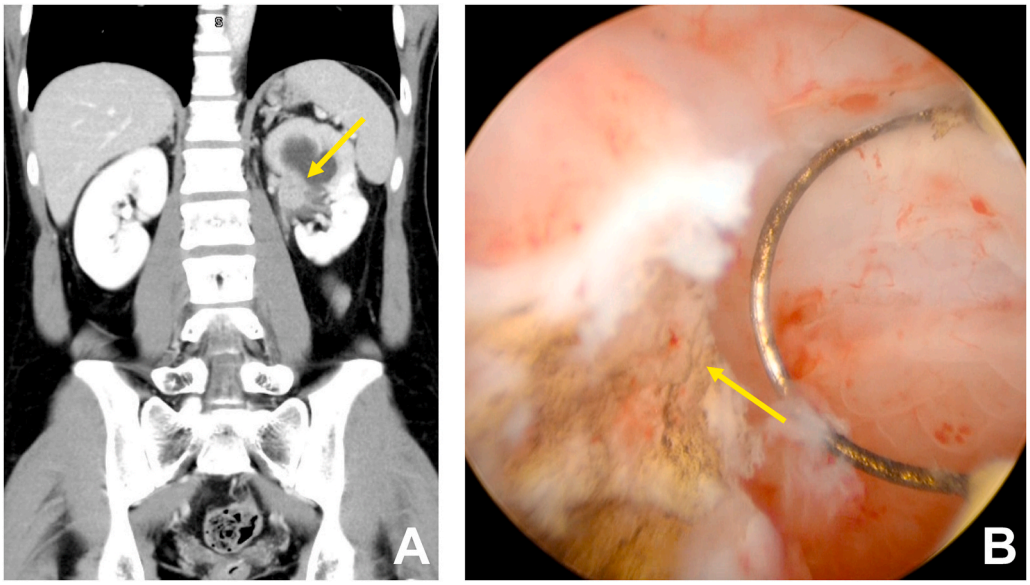

Fig. 2. Interval CT scan; the venous phase showed a significant growth of the mass in the left renal pelvis (2A). Percutaneous endoscopic view demonstrated a cautery loop resection of the renal pelvic mass (2B).

cells arranged in fascicles with moderate nuclear pleomorphism (Fig. 3A). These tumor cells express SMA (Fig. 3B) as well as other smooth muscle markers including MSA, caldesmon, and calponin. Based on these findings, a diagnosis of well-differentiated leiomyosarcoma of the renal pelvis was suggested.

The patient underwent open radical nephrectomy as the definite treatment. On bisection of the kidney, there was a 2-cm, well-demarcated, solid, white/tan mass in the renal pelvis located close to the perihilar soft tissue with marked dilatation of the upper pole calyces. Histopathology demonstrated an infiltrative mass composed of intersecting fascicles of spindle-shaped neoplastic cells with moderate nuclear pleomorphism. Mitotic figures were seen in 2/10 high power fields, but tumor necrosis was not identified. The tumor involved the collecting system with minimal renal parenchymal invasion. All resection margins were not involved, and one reactive perihilar lymph node was found. We were able to confirm the diagnosis of well-differentiated leiomyosarcoma, grade 1, according to the Fédération Nationale des Centres de Lutte Contre Le Cancer (FNCLCC) grading system.[2] Postoperatively, hematuria and flank pain completely resolved, and no adjuvant treatment was provided. The patient has been regularly followed with CT scan, and he remains recurrence-free at one year after surgery.

3. Discussion

Renal leiomyosarcoma is a rare but aggressive tumor, accounting for

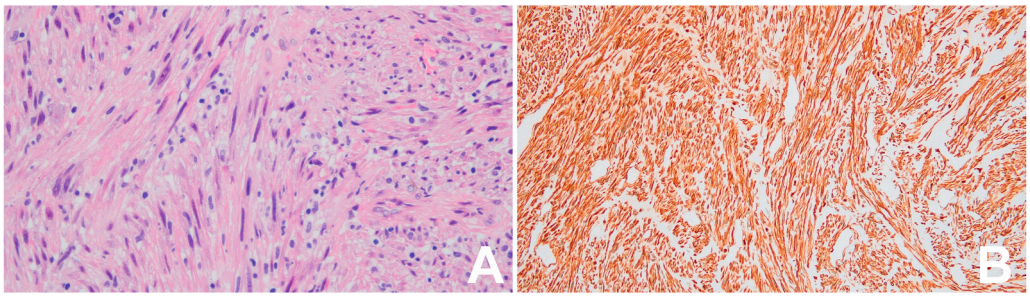

Fig. 3. Histopathological examination revealed spindle-shaped tumor cells arranged in fascicles with moderate nuclear pleomorphism, nuclear hyperchromasia, and a mitotic figure; H&E, original magnification, x400 (3A). Immunohistochemical staining with peroxidase yielded strong and diffuse positivity for SMA; original magnification, x100 (3B).

0.5–1% of all renal malignancies, and the renal pelvis is the least common site.[1] It may arise from the renal pelvic capsule, smooth muscle, or vasculature of the renal pelvis.

Acquiring the diagnosis of renal pelvis leiomyosarcoma preoperatively is challenging. Radiographic findings are nonspecific, and most cases were pathologically diagnosed following nephrectomy or nephroureterectomy. Ureterorenoscopy with biopsy, as in our initial attempt, may fail to obtain an adequate tissue specimen. Chow et al. reported one case correctly diagnosed by ultrasound-guided needle aspiration cytology[3]; however, another group reported a conflicting result. Ranadive et al. found sarcomatoid renal cell carcinoma on the fine needle aspiration cytology, which turned out to be leiomyosarcoma on the final nephrectomy specimen.[4]

In the presented case, we chose to perform percutaneous endoscopic biopsy to obtain more samples for histopathology. Our case is the first published report utilizing an endoscopic approach for tissue diagnosis. Nevertheless, the higher perioperative risks and complications associated with percutaneous endoscopy should be appropriately weighed against the diagnostic benefits. Tumor seeding along the tract has been reported, especially for urothelial carcinoma, and should be thoroughly considered.[5]

Because of the rarity of the disease, the prognosis is unknown, and the standard treatment has not been established. It is uncertain whether radical nephrectomy alone or radical nephroureterectomy is superior. Radical resection with a wide margin, as in other sarcomas, has been generally advised. Some reports recommended adjuvant chemoradiation due to the aggressive nature of the disease. Nevertheless, others reported long survival after surgical treatment alone. Our patient did not receive adjuvant therapy and is disease-free one year after surgery. In fact, the impact of different treatment strategies cannot be evaluated until more cases are reported, and data on long-term prognosis become more abundant.

4. Conclusion

We present a rare case of leiomyosarcoma arising from the renal pelvis in a young male patient. Percutaneous endoscopic biopsy was utilized for histopathological diagnosis, and the patient was subsequently treated by radical nephrectomy. No adjuvant treatment was provided, and the patient remains recurrence-free at one year. Because of the rarity of this disease, the prognosis is unknown, waiting for more reports and data on long-term follow-up.

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Declaration of competing interest

None.

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