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Oncology

En-bloc resection of a mixed mesenchymal and stromal tumor from the renal pelvis using laparoscopic pyelotomy – A case report

Ido Orlin*, Yoram Dekel, Boris Friedman, Rani Zreik, Leonid Boyarsky, Gal Rinott Mizrahi, Omer Ephrat, Avi Stein

Department of Urology, Lady Davis Carmel Medical Center, Haifa, Israel

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Introduction

Mixed epithelial and stromal tumor (MEST) of the renal pelvis is a rare, benign adult renal neoplasm with a variable content of epithelial and mesenchymal components. MEST was first described by Pawade et al. as a cystic hemartoma of the renal pelvis.[1] It has also been called adult mesoblastic nephroma, leiomyomatous renal hemartoma, solid and cystic biphasic tumor, solitary multilocular cyst, and adult metanephric stroma tumor.[2]

MEST occurs predominantly in middle-aged women and some correlation with menopausal status and hormonal replacement therapy (HRT) has been hypothesized.[1] Grossly, MEST is described as a typically well-circumscribed, solid and cystic mass, ranging from 2 cm to 24 cm. MEST often protrudes into the renal pelvis and the involvement of the renal hilum with compression of the pelvicalyceal system is common. Gross infiltration of the adjacent renal parenchyma is not usually seen.[1] MEST is classified as a benign tumor with virtually no predilection for metastatic spread or recurrence. However, infrequent malignant cases have been reported[3]

Histologically, MEST is a complex neoplasm, composed of a variety of epithelial elements embedded in a stroma of variable composition. The mesenchymal component varies from regular tubules to complex tubulopapillary structures with or without cystic dilatation. Of note, the stromal elements display variable hypercellularity, sometimes resembling ovarian stroma.[1] Immunohistochemistry staining is often positive for cytokeratins, representing the epithelial component whereas estrogen and progesterone receptors stains are often observed in the majority of the mesenchymal elements of MEST.[2]

MEST may have a diverse radiological appearance, with most presenting as multiseptated cystic renal masses with thick septa and nodular components. Due to the challenge of accurately discriminating MEST from renal cell carcinoma (RCC) based on radiologic imaging, surgical resection is often recommended with nephron sparing surgery (NSS) as the treatment of choice in selected patients.[4]

All reports that we have found state that MEST is excised exclusively by total nephrectomy or NSS. We report the first MEST case removed using laparoscopic pylelotomy with an enbloc excision.

Case presentation

A 47-year-old female presented with right hydronephrosis as an incidental finding when a spine computer tomography (CT) scan was preformed due to chronic lower back pain. She denied flank pain, hematuria or unintentional weight loss. She had no history of smoking nor had she been exposed to chemicals that are known as risk factors for urothelial carcinoma (UC). Her past medical history included hypertension and type 2 diabetes mellitus and she is menopausal for 3 years without Hormone Replacement Therapy (HRT). Physical examination and laboratory evaluation were within normal limits. Urine cytology was negative.

Radioisotope renography with Tc99m-DTPA (diethylenetriaminepentacetate) was performed with equal bilateral renal function with normal excretion.

CT urography (Fig. 1.) revealed a right renal pelvic mass, measuring about 5 cm with mild contrast-enhancement. A filling defect was observed at the pyelographic phase.

Cystoscopy and right side ureteroscopy (URS) demonstrated an intraluminal mass occupying most of the renal pelvis appearing as a

* Corresponding author.
E-mail address: orlinid@gmail.com (I. Orlin).

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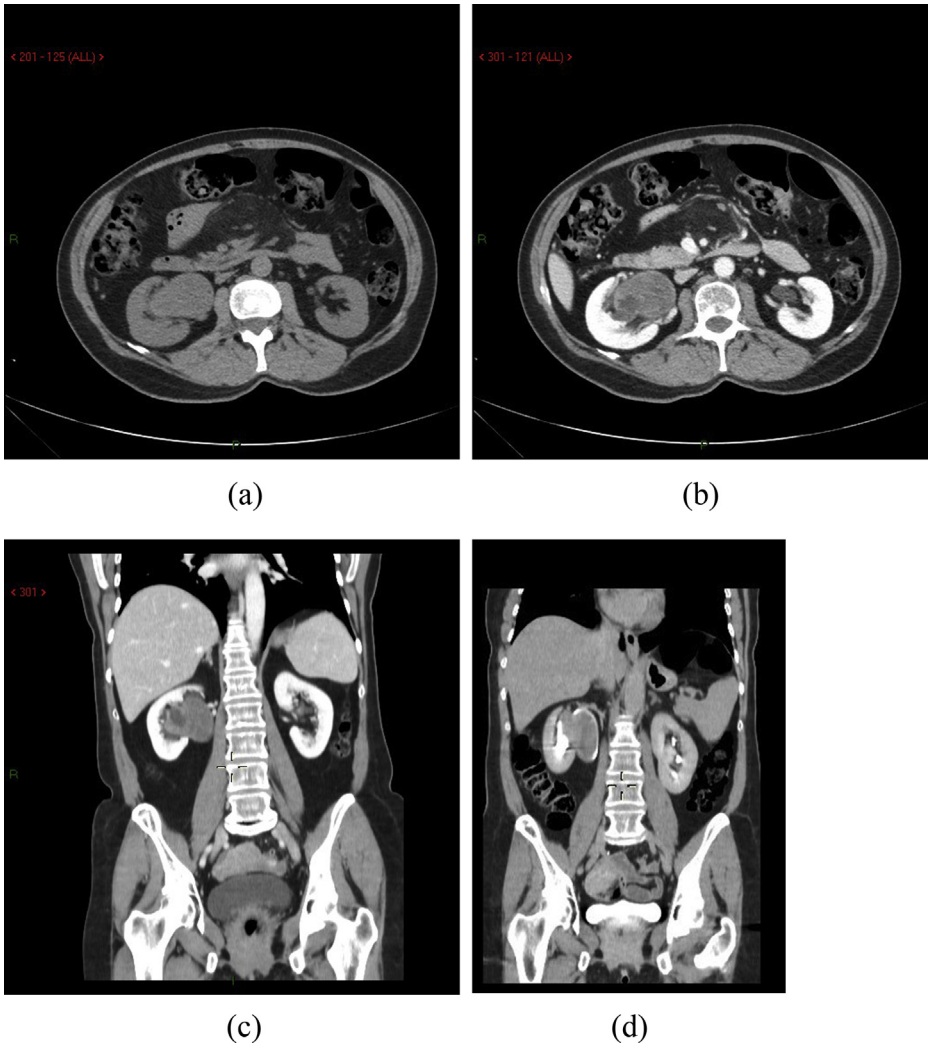

Fig. 1. Computer tomography urography (CTU). A- Non-contrast cross section, B- Nehprographic phase showing slightly-enhanced right pelvic mass, C- Nephrographic phase coronal section, D- Coronal section of the pyelographic phase showing right pelvic filling defect.

smooth walled mass with a well demarcated border. Upon retrograde pyelography, a clearly visible filling defect was seen in the right renal pelvis. The lesion was biopsied. Cytoscopy was unremarkable. Selective cytology from the right ureter was negative for malignant cells.

Histologic examination of the specimen showed a spindle and elongated non-atypical cells partly organized in bundles and situated beneath the urothelium. Mitotic activity was absent. Immunohistochemistry for vimentin, desmin, estrogen receptors (ER) and progesterone receptors (PR) was positive. keratin, keratin 7, keratin 907, p-63, s-100, PAX-8, HMB-45, CD34, LCA immunostains were negative. Diagnosis of a benign mesenchymal lesion suggestive of leiomyoma was made.

The patient continued to be asymptomatic throughout the diagnostic evaluation without impairment of renal function or symptoms of urinary tract infection.

We performed a laparoscopic pyelotomy of the right renal pelvis with an en-bloc resection of the mass (Fig. 3.). The renal pelvis was opened and the large mass was completely resected using scissors. Closure of ureteropelvic junction (UPJ) and the proximal ureter were primarily sutured. Before complete closure, a double J stent was placed which was removed 6 weeks post-operatively without complication.

The final pathologic examination of the extracted specimen confirmed the diagnosis of mixed epithelial and stromal tumor (MEST) of the renal pelvis with negative margins (Fig. 2.).

Discussion

Benign tumors of the kidney are rare neoplasms. According to the medical literature they can only be diagnosed after radical nephrectomy, nephrouretrectomy or partial nephrectomy. This is due to a strong radiologic indication of malignant tumors of the urological system. In fact, on imaging, almost 70% of MEST and other benign renal neoplasms have the imaging characteristics of Bosiniak category IV or solid lesions and therefore definitive surgical indication is required.[5] Diagnosis of MEST should be kept in mind in cases of middle aged women with cystic renal tumors arising from the renal pelvis with negative cytology. Ureteroscopy should be considered if the lesion in question is accessible for biopsy to confirm the benignity of the mass. Our decision to collect tissue diagnosis from the mass endoscopically led to a preoperative diagnosis of leiomyoma which would have been completely resected in the same way. The complete excision of the mass using minimally invasive techniques with minimal damage to normal

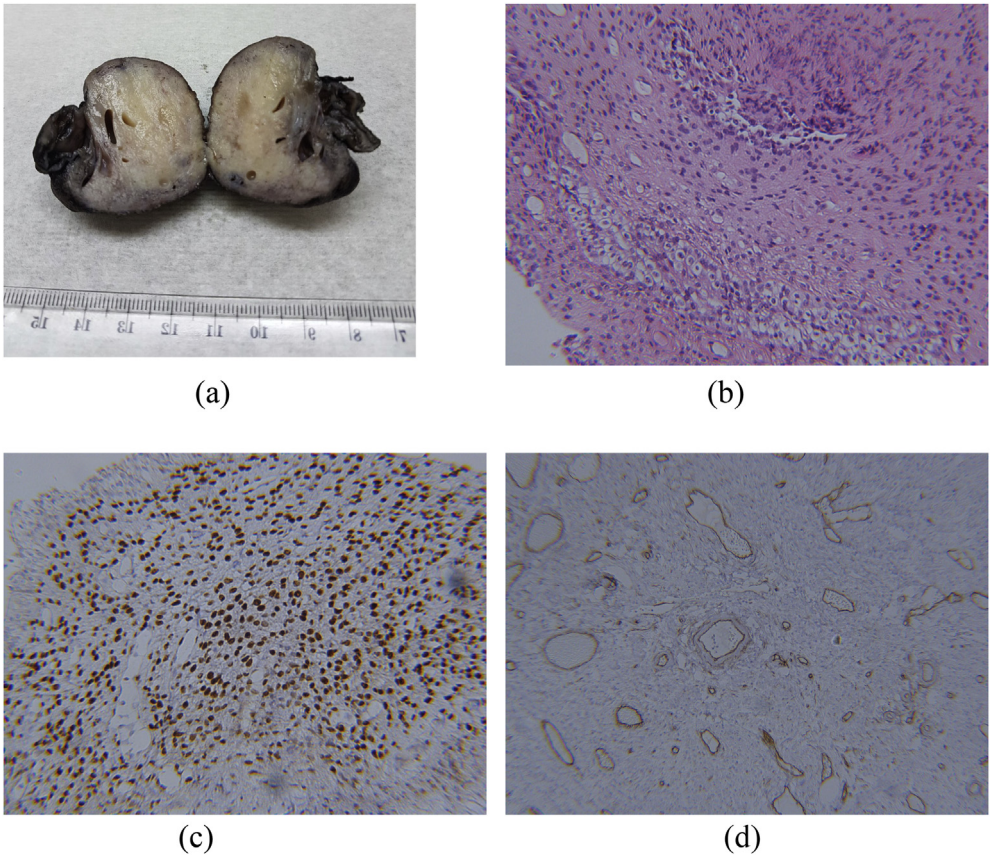

Fig. 2. Gross Pathology and Histopathology; A. Cross section of the excised specimen B. Hematoxylin and Eosin (H&E) staining C. Immunohistochemistry positive for estrogen stain D. Immunohistochemistry staining negative for CD-34.

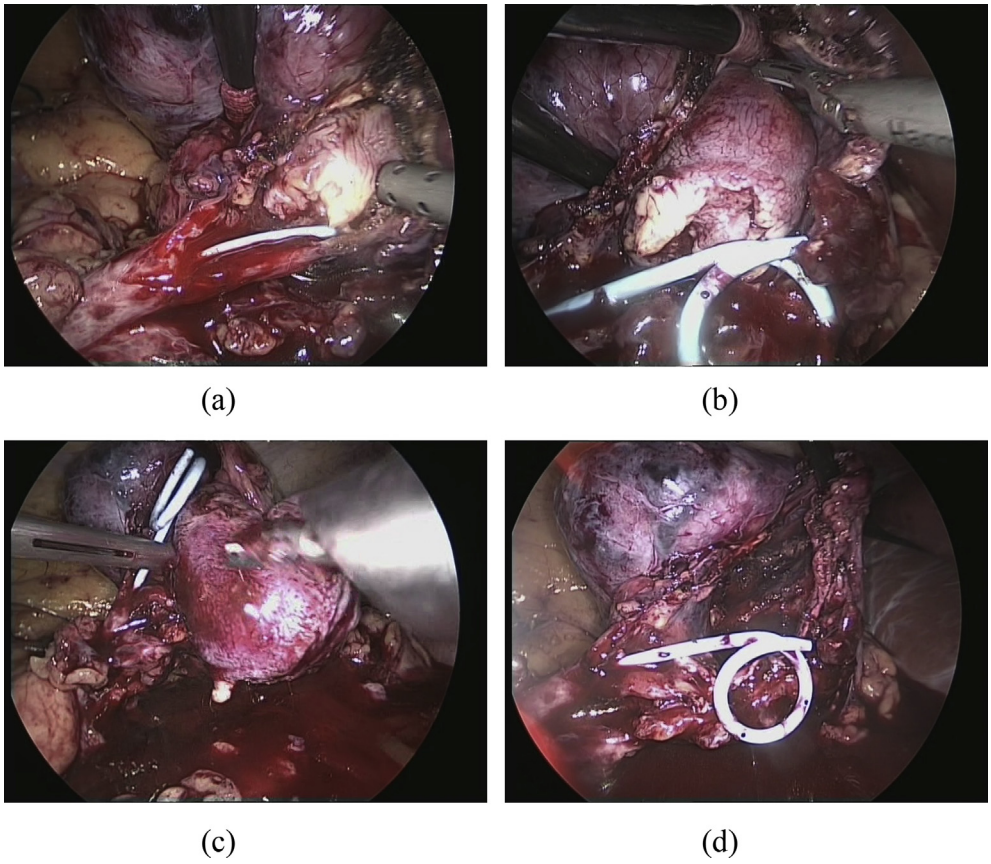

Fig. 3. Intra-operative images A. Unraveling the mass from the renal pelvis B. Complete dissection around the mass within the renal pelvis C. En-bloc excision of the mass D. Empty Renal Pelvis.

renal parenchyma should be carefully considered after examining tumor location in respect to vital renal structures and the surgeon's expertise. Open or laparoscopically pyelotomy with en bloc resection are feasible options in selected cases.

Conclusion

MEST is a rare, benign neoplasm of the renal parenchyma and pelvis. Complete excision of renal pelvic masses through pyelotomy using minimally invasive techniques with minimal removal of renal parenchyma is feasible in selected cases where the index of suspicion for malignancy is minimal.

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