

Case Report

Enucleation/partial nephrectomy for large mixed epithelial stromal tumor and herniating into the pelvicalyceal system

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Abstract

Objectives: Mixed Epithelial and Stromal Tumor of the kidney is an adult renal neoplasm. It is mostly benign in nature. Typically it is composed of a mixture of epithelial and mesenchymal components. We hereby report on the feasibility of performing partial nephrectomy/enucleation for Huge Mixed Epithelial Stromal Tumor of the kidney without sacrificing the involved renal unit even in the tumors herniating into the collecting system.

Methods: Two female patients on long term hormonal therapy developed large enhancing multiloculated and septated renal masses. Kidney mass size was 18.5 cms in one patient and 11.5 in the second. In one patient, the mass was herniating into the collecting system. Both patients had enucleation/partial nephrectomy.

Results: Enucleation and partial nephrectomy were successfully performed in both patients. In the patient with the mass herniating into the collecting system, the horns of the mass herniating into the collecting system were easily enucleated with repair of the collecting system and salvage of the involved renal unit. Post op pathology revealed MEST in both patients. There were no intraoperative or postoperative complications.

Conclusions: Enucleation and partial nephrectomy for huge MEST is feasible. Mixed Epithelial Stromal Tumor herniating into the pelvicalyceal system may not warrant nephroureterectomy as previously reported.

Key Words: Enucleation, kidney, mixed epithelial stromal tumor, partial nephrectomy

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INTRODUCTION

Mixed epithelial and stromal tumor of the kidney (MEST) is a new entity in renal tumors. It was first diagnosed in 1998 by Michal and Syrucek.^[1] It has a largely female preponderance and usually presents at the 4th decade in women on long-term hormone therapy. MEST of the kidney is largely a benign tumor. However, there are few cases of recurrence following

surgical excision as well as malignant transformation.^[2] Recently, one malignant renal MEST was reported in a male patient on hormonal therapy for prostate cancer.^[3] These tumors have a propensity to occur near the renal pelvis.

Microscopically, these tumors are composed of a mixture of fibrous stroma and epithelial glands and cysts. The stroma can vary from hypocellular, sclerotic fibrous tissue to hypercellular proliferations of spindle cells. Ovarian-type stroma is often present as a component of the fibrous areas. Smooth muscle may also be present within the solid, fibrous component. The glands and cysts are scattered throughout the stroma and may have areas of clustering. The glands and cysts are most often lined by flattened, cuboidal or hobnailed epithelial cells, although columnar, clear, and urothelial-like linings may be encountered as well. The epithelial cells appear bland. Immunohistochemistry shows positive staining for progesterone receptor (PR) and

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estrogen receptor (ER) within the stromal component in the majority of cases. Staining for calretinin, CD10, and inhibin may also be identified within the stromal cells.^[4]

Clinically, these tumors may present as a large renal mass that may compress the pelvicalyceal system. This clinical picture may confuse with conventional renal cell cancer and transitional cell cancer of the collecting system. In the particular cases with MEST compressing the collecting system management was a nephroureterectomy.^[5,6]

In this report, we present two patients with MEST. One had a large MEST and the second had a large MEST compressing the pelvicalyceal system and both were successfully managed with enucleation/partial nephrectomy and without sacrificing the involved renal unit.

CASE REPORTS

Case 1

A 64-years-Caucasian female has a 6 years history of large left “multilocular cystic nephroma.” That has been managed conservatively with yearly renal ultrasound/computed tomography (CT) scan. She has a history of intake of birth control pills for 15 years and she received hormonal replacement therapy for another 15 years following a hysterectomy for endometriosis. She has a history of simple mastectomy for ductal carcinoma *in situ*. In addition, she has a history of hypertension and hyperlipidemia. She is a nonsmoker, never had hematuria and with normal urine analysis. Physical exam revealed an enlarged kidney that was easily palpable through the anterior abdominal wall. After 6 years of follow-up, the left kidney cystic mass enlarged in size from 14 cm to 18.5 cm and started to cause left flank pain and interfere with the patient’s daily activities [Figure 1]. A decision is made to proceed with left partial versus radical nephrectomy.



Figure 1: Computed tomography scan of left renal multilocular cystic mass

Case 2

A 51-year-old Caucasian female presented to her local Urologist with a history of recurrent urinary tract infections and new onset of left abdominal pain. Her past medical history is significant for intake of birth control pills for 15 years. There is no history of hematuria. She uses to smoke 2 packs of cigarettes every day for the past 30 years. Physical examination was unremarkable. Urine analysis showed trace of leucocytes but otherwise clear. A CT scan was performed and this revealed 11.5 cm left multiloculated cystic renal mass. Of note that the mass on the CT scan looked as if it is compressing or extending into the collecting system giving the impression of possible urothelial carcinoma. Given the symptomatic nature of the cystic disease and the risk that it could be malignant in nature, the patient was advised to proceed with open partial versus radical left nephrectomy ± ureterectomy with bladder cuff excision.

RESULTS

Case 1

The procedure was performed through a transcoastal flank incision with excision of the 11th rib. Following dissection of the mass, despite its huge size a bloodless plane between the base mass and the kidney was identified and this allowed for easy enucleation of the mass with minimal blood loss. No clamping of the renal pedicle was needed. A cavity was left after enucleation that was filled with perinephric fat and the edges were approximated with chromic 2-0 sutures. The patient tolerated the procedure well. The estimated blood loss from the surgery was only 100 ml and the patient was discharged on the post-operative day 6 with no intraoperative or post-operative complications.

Pathology revealed a multi-loculated cystic mass measuring 18.5 cm [Figure 2]. The mass was tan to pink in color and was grossly composed of back-to-back cystic structures, with

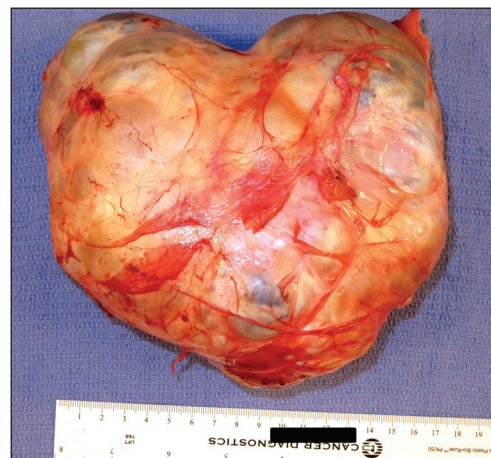


Figure 2: Left renal mass after excision

no solid component noted. Clear, straw-colored fluid was present within the cysts. Microscopically, the majority of the stromal component of the tumor was composed of hypocellular fibrous tissue with variable degrees of sclerosis. Small foci of calcification were present. No ovarian-type stroma was identified.

Case 2

The procedure was performed through a transcostal flank incision with excision of the 12th rib. At the time of exploration, similar to the previous patient, large portion of the mass was easily enucleated with gentle blunt and sharp dissection. A partial nephrectomy was mandated in an area near the renal hilum where the mass was not easily enucleated. The two “horns” herniating into the collecting system and giving the impression of possible urothelial carcinoma were easily enucleated as well with closure of the collecting system [Figure 3]. The partial nephrectomy site was closed in a standard fashion. The patient tolerated the procedure well and the estimated blood loss was 400 ml. The patient was discharged on the post-operative day 3 with no intraoperative or post-operative complications.

Pathology revealed a tan to pink, multi-loculated cystic mass measuring 11.5 cm. The cysts had smooth linings and were filled with straw-colored serous fluid. Areas of ovarian-type stroma were present in multiple foci. Immunohistochemical staining for ER showed patchy positive nuclear staining while staining for PR showed diffuse positive staining in the stromal cells [Figures 4 and 5].

DISCUSSION

MEST of the kidney is a rare renal tumor and more reports are needed to increase the awareness of Urologists about that tumor and to better understand its biological behavior and its optimum management.

In the largest reported series by Michal *et al.* on MEST, one third of 22 female patients had MEST bulging into the collecting system. In that same report, a comparison regarding the histopathological features of MEST in females was made to MEST diagnosed in 2 male patients in their data base. In the male patients, the cyst walls tend to be thinner and completely devoid of ovarian type stroma.^[7]

Radiologically, these tumors present as large multiloculated cystic masses with internal septa and solid components. Delayed contrast enhancement may be present as well.^[8]

In one of the two cases, we present (case 2), it was very difficult to pre-operatively determine if the mass was compressing or actually invading the collecting system. We did not perform a retrograde study on that patient since the mass was predominantly parenchymal and the smooth well-circumscribed



Figure 3: Left renal mass with 2 horns herniating into the collecting system

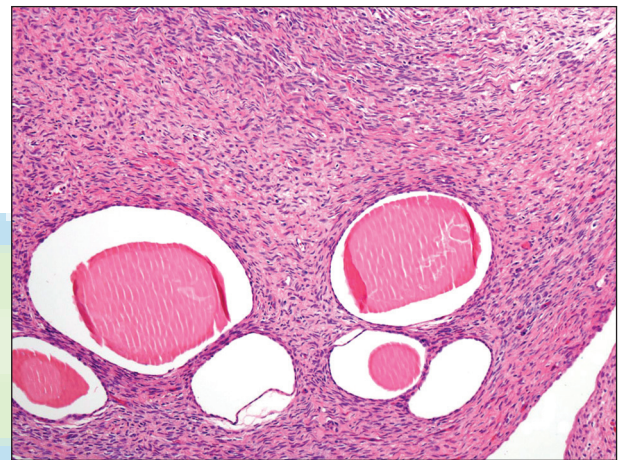


Figure 4: Microscopic picture for mixed epithelial and stromal tumor

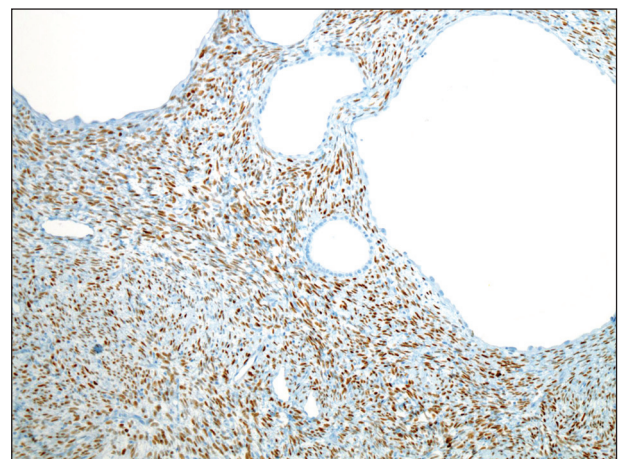


Figure 5: Immunohistochemical staining positive for progesterone and estrogen receptors

wall was not favoring a transitional cell cancer. Similarly, the absence of hematuria despite the large size of the mass was not favoring Transitional Cell Carcinoma of the upper urinary tract. In previously reported MEST cases with similar radiologic

findings, retrograde imaging findings led to immediate nephroureterectomy for a presumed diagnosis of urothelial carcinoma of the upper urinary tract. However, post-operative findings revealed these tumors were MEST herniating into the collecting system.^[5,6] Consequently, we recommend keeping a high index of suspicion for these tumors since retrograde studies are not diagnostic for these tumors and may actually lead to a radical surgery with the loss of the involved renal unit for these largely benign tumors. Interestingly was the ease of finger enucleation of the cyst herniating into the collecting system. We stress in these situations that if excessive bleeding is encountered a radical nephrectomy must be performed.

In our case, presentations despite the huge size of MEST we were able to enucleate them and with acceptable blood loss and with preservation of kidneys. We have noted in our two patients and upon review of the reported cases in the literature that these tumors tend to be exophytic. The largely benign nature of these lesions may encourage a nephron sparing approach as well. Consequently, we recommend an attempt to enucleate or perform partial nephrectomy for MEST even if they were large. We also recommend to attempt to enucleate the portion of the tumor that may give the impression of urothelial carcinoma of the renal pelvis before embarking into nephroureterectomy.

CONCLUSIONS

MESTs of the kidney is common in women of child bearing

age in particular those on hormonal therapy. Urologists should attain a high index of suspicion with this particular patient population. Our report demonstrates the feasibility of open enucleation and partial nephrectomy for large MEST. The picture of MEST compressing the collecting system may not mean that a nephroureterectomy is an absolute necessity.

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