Locally Aggressive Mixed Epithelial and Stromal Tumour of Kidney Leading to Nephron Loss

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ABSTRACT

Mixed epithelial and stromal tumour (MEST) is a rare benign renal tumour. It is mainly found in perimenopausal women. We present a case of a 42-year female with no known comorbid who was presented in the outpatient clinic for the right flank pain. Contrast-enhanced CT scan revealed a complex renal cyst with internal septations. Considering a large symptomatic cyst and the presence of internal septations, she was planned for cyst excision. Peroperatively, significant disease progression and loss of renal parenchyma were noted contrary to preoperative scan. Histopathology of the specimen revealed MEST. We demonstrate that MEST of the kidney may have an aggressive local behaviour leading to nephron loss.

Key Words: *Mixed epithelial and stromal tumour, Kidney, Benign, Renal neoplasm.*

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INTRODUCTION

Mixed epithelial and stromal tumour (MEST) is a rare renal neoplasm which accounts for about 0.2% of all renal neoplasms.[1] Michal and Syrucek identified MEST in 1998 which was included in WHO renal tumour classification in 2004 as a separate entity.[2,3]

It is predominantly found in perimenopausal women with male to female ratio of 1:10. Most of these tumours behave in a benign fashion and only a few have been reported as malignant.[4,5] We present a case of a 42-year female with no known comorbid who was presented in the outpatient clinic for right flank pain. She was finally diagnosed on histopathology as MEST of a kidney.

CASE REPORT

A 42-year married lady with no prior known comorbid presented in the outpatient department of urology with the right flank heaviness for four months. She had no lower urinary tract symptoms, normal bowel habits, and normal menstrual cycles. The history was significant for open appendectomy and cesarean section. Her family history was unremarkable for any illness. Her general physical and systemic examinations were also unremarkable.

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The basic haematological and biochemistry workup was normal and serum creatinine was 0.8 mg/dl. Computerised tomography (CT) scan of abdomen and pelvis with contrast revealed a large exophytic, cystic lesion at the upper pole of the right kidney with thick internal septations. No internal calcification or solid component was identified. It measured 80 × 75 mm on axial sections. Imaging features were suggestive of Bosniak type 2F renal cyst (Figure 1).

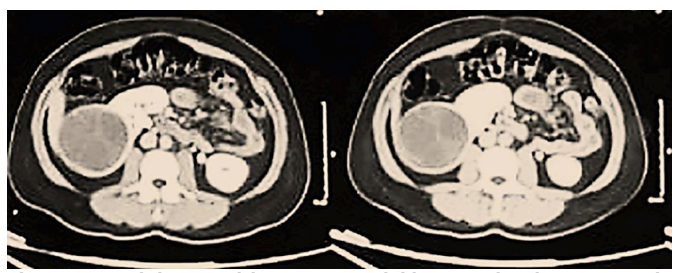

Figure 1: CT abdomen with contrast. Axial images showing upper pole complex renal cyst with internal septations.

Considering the symptoms and large size, she was given the options of follow-up *vs.* cyst excision. She opted for later as she had to visit the outpatient department from a remote area with regular scans on follow-ups. The patient was planned for open cyst excision. Per-operatively, significant progression of cyst size was noted from the time of the last imaging that was done less than four weeks prior to surgery. The cyst was hard in consistency and it had replaced the upper and mid pole and was reaching up to the renal hilum, sparing only a small portion of normal renal parenchyma at the lower pole. Taking the scenario into account, radical nephrectomy was performed. Another important factor which contributed in decision- making was the quantity and quality of residual parenchyma, we had performed partial nephrectomy (Figure 2). The Postoperative course was smooth and she was discharged on the fourth day.

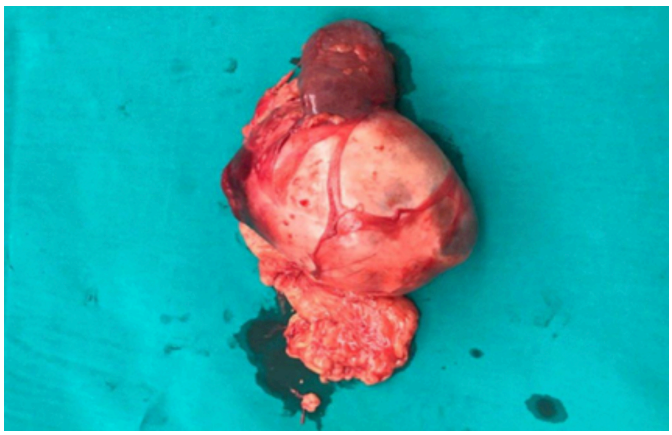

Figure 2: Gross specimen showing large mass replacing upper and mid-pole.

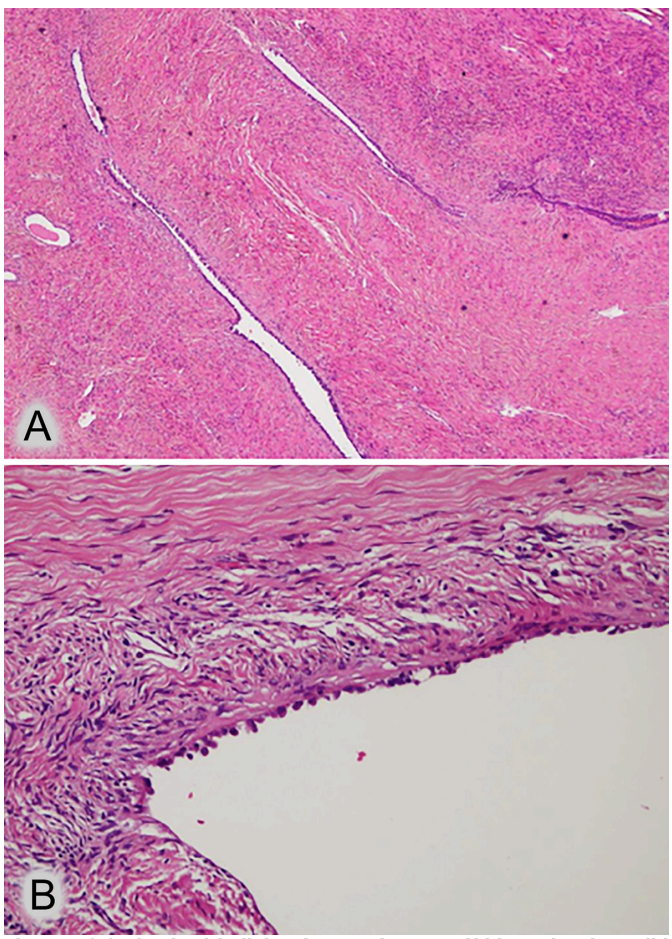

Figure3: (A) Mixed epithelial and stromal tumor of kidney showing solid and cystic components with heavily collagenised stroma and collapsed cystic spaces lined by cuboidal epithelium (H&E, ×10). (B) Mixed epithelial and stromal tumor of kidney showing cystic space lined by cuboidal epithelium showing nuclear "hobnailing". The stroma is ovarian type with relative sub-epithelial condensation (H&E, ×20).

Histopathology of the specimen revealed the unusual finding of MEST. Grossly, the cut surface showed a circumscribed mostly cystic and focally solid tumour in the upper pole and middle part of the kidney measuring 12×8×7 cm. Microscopic examination revealed renal parenchyma mostly replaced by a circumscribed lesion with cystic and solid areas (Figure 3A). The cystic spaces were lined partly by denuded cuboidal cells with bland cytological features and prominent nuclear "hobnailing" (Figure 3B). The underlying stroma was fibromatous, and ovarian type with relative cellular condensation underneath the epithelium. Entrapped within the stroma, were multiple tubules lined by bland columnar cells. These morphological features were consistent with MEST of kidney.

Her immediate course after surgery was uneventful and she was asymptomatic at six-month follow-up. Repeat CT scan at six-month follow-up showed no local recurrence.

DISCUSSION

MEST, formerly called as cystic hamartoma of the renal pelvis or cystic nephroma, was included in the WHO 2004 classification of renal tumours. It is a rare kidney tumour, which contains both epithelial and stromal elements.[2] It is found most commonly in middle-aged peri-menopausal women. There are also some reports of male cases as well.[6,7] Most common presentations of MEST include abdominal mass, flank pain, and haematuria. On CT scan, the tumour appears as solid and cystic mass; however, the final diagnosis is based on histopathology.[8]

Both benign and malignant renal neoplasms can present as complex renal cysts which are usually difficult to differentiate based on imaging. Benign conditions include renal abscess and hydatid cyst in people from endemic areas but these can usually be differentiated based on history and specific CT findings. However, the absence of these symptoms or CT findings does not exclude these diagnoses. More important consideration especially in large (>4cm) complex renal cysts is cystic renal cell carcinoma. A preoperative image-guided biopsy can help in reaching a diagnosis. However, in this type of cyst without a solid component, an image-guided biopsy cannot rule out renal cell carcinoma or other rare conditions like MEST and other multiloculated benign tumours.

Histologically, MEST is a tumour consisting of cysts and tubules embedded in spindle cell stroma. The exact cause is not known, but it has been proposed that both stromal and epithelial components are neoplastic and originate from a single cell. Microscopically, the stroma may look like ovarian stroma and surrounds clusters of tubules or cystically distended glands with variable lining. Immunohistochemically, the epithelial components are generally positive for epithelial membrane antigen and cytokeratin.[9]

MEST mostly behaves in a benign fashion. Recently, few cases have been described in the literature with malignant behaviour. Most cases were in female patients except for a few in males.[10]

This patient was a middle-aged woman who presented with right flank heaviness. MEST should be considered in the differential diagnosis of complex renal cystic lesions, especially in peri-menopausal women.

PATIENT'S CONSENT:
The consent was obtained during the follow-up of the case from the patient in her native language after adequate information was provided by one of the authors of this case Report.

The patient read the informed consent document in detail, and all of the questions were answered to their satisfaction. The patient signed the informed consent document, and a copy of the signed document was provided to her.

COMPETING INTEREST:
The authors declared no competing interest.

AUTHORS' CONTRIBUTION:
MS, IKJ, AR: Contributed to literature search and writing of manuscript case.
ZU: Provided histopathology slide images and description.
FA: Provided patient data including patient consent.
All the authors have approved the final version of the manuscript to be published.

REFERENCES

1. Horsanali MO, Yilmaz Y, Ozer K, Kose O, Cakalagaoglu F, Etit D. Mixed epithelial and stromal tumor of the kidney: Two case reports of a rare benign kidney tumor. *Clin Genitourin Cancer* 2017; 15(1):e55-9. doi: 10.1016/j.clgc.2016.10.009.
2. Arriola AG, Taylor BL, Ma S, Malkowicz SB, Lal P. Malignant mixed epithelial and stromal tumor of the kidney with 2 simultaneous renal carcinomas in a male patient: Case report and review of the literature. *International J Surgical Pathology* 2018; 26(1):56-63. doi: 10.1177/1066896917720032.
3. Gokden N, Dawson K, Lindberg M. Malignant rhabdoid tumor arising in a mixed epithelial, stromal tumor of kidney: report of a male case, review of the literature. *Pathology Research Practice* 2020; 216(10):153151. doi: 10.1016/j.prp.2020.153151.
4. Mohanty SK, Parwani AV. Mixed epithelial and stromal tumors of the kidney: An overview. *Arch Pathol Lab Med* 2009; 133(9):1483-6. doi: 10.5858/133.9.1483.
5. Jung SJ, Shen SS, Tran T, Jun SY, Truong L, Ayala AG, Ro JY. Mixed epithelial and stromal tumor of kidney with malignant transformation: Report of two cases and review of literature. *Human Pathology* 2008; 39(3):463-8. doi: 10.1016/j.humpath.2007.08.008.
6. Michal M, Hes O, Bisceglia M, Simpson RH, Spagnolo DV, Parma A, *et al*. Mixed epithelial and stromal tumors of the kidney. A report of 22 cases. *Virchows Archiv* 2004; 445(4):359-67. doi: 10.1007/s00428-004-1060
7. Adsay NV, Eble JN, Srigley JR, Jones EC, Grignon DJ. Mixed epithelial and stromal tumor of the kidney. *American J Surgical Pathol* 2000; 24(7):958-70. doi: 10.1097/00000478-200007000-00007.
8. Wang CJ, Lin YW, Xiang H, Fang DB, Jiang P, Shen BH. Mixed epithelial and stromal tumor of the kidney: Report of eight cases and literature review. *World J Surgical Oncol* 2013; 11(1):207. doi: 10.1186/1477-7819-11-207.
9. Kum JB, Grignon DJ, Wang M, Zhou M, Montironi R, Shen SS, *et al*. Mixed epithelial and stromal tumors of the kidney: Evidence for a single cell of origin with capacity for epithelial and stromal differentiation. *Am J Surgical Pathol* 2011; 35(8):1114-22. doi: 10.1097/PAS.0b013e3182233fb6.
10. Montironi R, Cheng L, Lopez-Beltran A, Scarpelli M: Editorial comment from Dr Montironi *et al*. to malignant mixed epithelial and stromal tumor of the kidney: Report of the first male case. *Int J Urol* 2013; 20(4):451-2. doi: 10.1111/j.1442-2042.2012.03164.x.