An Unusual Presentation of Mixed Epithelial and Stromal Tumor of the Kidney

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ABSTRACT

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A 45 year old male reported with flank mass which on evaluation was found to be gross hydronephrosis containing multiple nodular lesions. Nephroureterectomy was done with a diagnosis of urothelial cancer in a hydronephrotic kidney. Various pathological tests confirmed MEST. Mixed epithelial and stromal tumor (MEST) of kidney often arises centrally in the kidney and grows as an expansile mass. Most of the tumors are solid and many have cystic areas. MEST presenting as multiple polypoidal lesions in a grossly dilated kidney due to pelvi ureteric junction obstruction (PUJO) has not been reported so far.

Keywords: Mixed epithelial stromal tumor, Pelviureteric junction obstruction and kidney

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INTRODUCTION

Mixed epithelial and stromal tumor (MEST) of kidney, which is predominantly seen in females, is a rare tumor and is usually detected incidentally or as a flank mass. Hematuria and urinary tract infections are also common presenting features. It often arises centrally in the kidney and grows as an expansile mass, frequently herniating into the renal pelvis. Most of the tumors are solid and many have cystic areas. Because of the cystic nature, they are often mistaken for cystic nephroma. MEST presenting as multiple polypoidal lesions in a grossly dilated kidney due to PUJO has not been reported so far. To the best of our knowledge, this is the first such case to be reported.

CASE REPORT

A forty five year old man presented with painful swelling on the left side of abdomen of 5 months duration. Abdominal examination showed a large mass of about 15 x 10 cm. Ultrasonography (US) of the abdomen showed grossly dilated kidney with a 6X5 cms mixed echoic soft tissue lesion within the dilated pelvi calyceal system **(Figure 1A)**. Contrast enhanced CT (CECT) abdomen showed grossly dilated left kidney a with a mildly enhancing nodular lesion within the pelvicaliceal system. Urine cytology was negative. Considering a diagnosis of urothelial carcinoma with congenital PUJ obstruction, nephroureterectomy was done in the standard method. Gross examination showed **(Figure 1B)** markedly dilated pelvi calyceal system filled with hemorrhagic fluid, corticomedullary thinning and multiple polypoidal growths. Cut surface of all were greyish white with hemorrhagic necrosis and cystic changes. Microscopically, the polypoidal lesion was lined by cuboidal epithelium and was composed of biphasic components

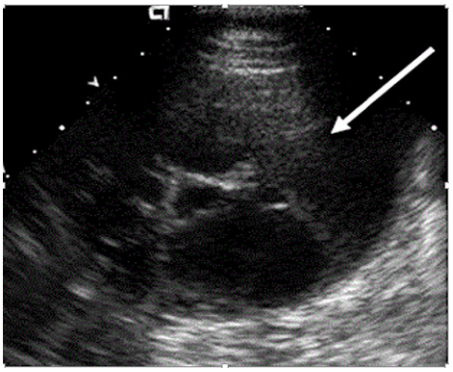

Figure 1A. **Grossly dilated PCS with a mixed echoic lesion within it**

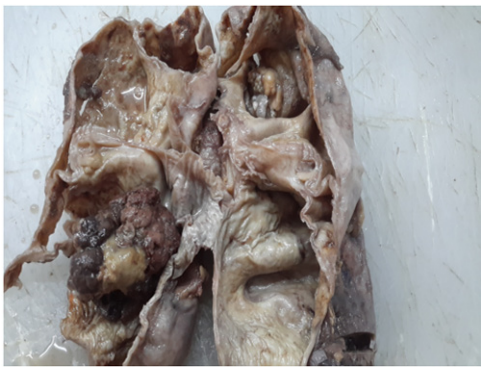

Figure 1B. **Grossly dilated PCS with multiple polypoidal growth**

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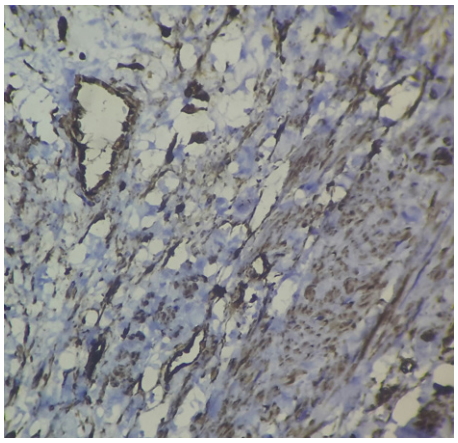

Figure 2A. **Positive for SMA**

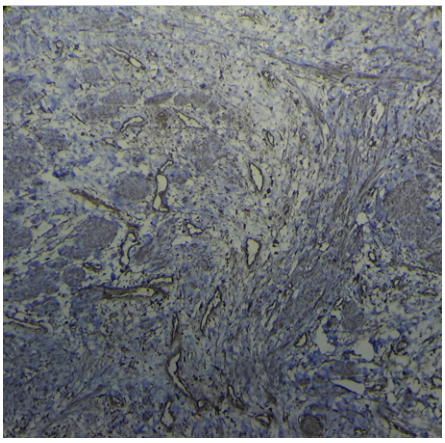

Figure 2B. **Positive for desmin**

including multiple branching tubules embedded in edematous myxoid spindle cell stroma. Section from ureter and renal vessels showed no pathology. Immunohistochemical examination revealed that the stromal cells were positive for smooth muscle actin **(Figure 2A)**, desmin **(Figure 2B)** and negative for estrogen and progesterone receptor. These findings confirmed the diagnosis of MEST. Post operative period was uneventful and at 6 months followup the patient was asymptomatic.

DISCUSSION

Mixed epithelial stromal tumor of kidney is a rare tumor that is more common in women and most are benign. In males, those who have received estrogen therapy may develop this tumor. This suggests a hormonal etiology for this tumor. Patients present with renal mass, flank pain, hematuria and urinary tract infection or are incidentally diagnosed. These tumors are also called as cystic hamartoma of the renal pelvis, adult mesoblastic nephroma, leiomyomatous renal hamartoma and mesoblastic nephroma. Malignant transformation, recurrence and metastases are rare.

Grossly, MEST Kidney is a well-circumscribed tumor with cystic and solid components of variable proportions. However, in our patient the presentation was unique in that there was grossly dilated kidney due to PUJ Obstruction and there were multiple polypoidal tumors in the pelvicaliceal system. Ramya et al described a case of MEST in a kidney which on US showed features of hydronephrosis as well.[1] However, post operatively it was seen that the cystic component of the tumor was mistaken for hydronephrosis on US. Sountoulides et al described a case of MEST simulating upper tract urothelial cancer.[2] The IVU and CT urogram demonstrated filling defect in the lower calyx in this patient. In our case, the estrogen receptors(ER) and progesterone receptors (PR) were negative on IHC. Turbiner et al in their analysis of 34 cases reported that ER and PR will be expressed in 62% and 85% of benign MEST kidney respectively.[3] MESTs of the kidney with negative ER and PR have been described by Zou et al and Nakagawa et al.[4,5] Both had malignant MEST. Menindez et al and Wang et al have reported a case each of MEST with negative PR and ER respectively.[6,7]

END NOTE

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Conflict of Interest: None declared

Editor's Remarks: This is an unusual case presenting as a cystic mass. The awareness needs to be present to suspect such cases.

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