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Pathology

Mixed Epithelial Stromal Tumour Kidney: Report of Two Cases, Benign and with Malignant Transformation

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Abstract **Case Report**

Mixed Epithelial Stromal Tumor kidney (MESTK), is a rare renal tumor, with stromal as well as epithelial components. It is predominantly benign with rare malignant transformation as morphology resembling that of a stromal sarcoma, carcinosarcoma, rhabdomyosarcoma, chondrosarcoma, undifferentiated sarcoma, or malignant tumour with rhabdoid features. Here we present 2 cases of MEST of kidney both are middle aged females with history of episodes of hematuria. Radiology of both showed heterogenous lesion. Histopathological and IHC findings favoured a diagnosis of first case as MEST kidney benign entity and second case as MEST with malignant transformation.

Keywords: Mixed Epithelial Stromal Tumor kidney.

INTRODUCTION

Mixed epithelial and stromal tumor Kidney (MESTK) is a complex neoplasm composed of a mixture of stromal and epithelial elements which occurs predominantly in middle-aged peri-menopausal women and older women. Mixed epithelial and stromal tumour (MEST) develops from Müllerian-like stromal cells and accounts for 0.2% of all renal cancers. The tumor was first identified by Michal and Syrucek in 1998 and included in the WHO renal tumour classification as a separate entity in 2004 [1]. Most of these tumours behaved in a benign fashion and only 13 cases with malignant transformation were reported: 11 cases in women and two cases in men [2]. Current evidence suggests that the most common type of malignant MEST is undifferentiated sarcoma. In this paper, we report detailed clinicopathological findings and clinical outcomes of 2 cases of MEST exhibiting focal positive immunoexpression of estrogen and progesterone receptors and one with malignant transformation.

CASE REPORT

Case 1- 38 year old female with history of episodes of hematuria for 6 months. Her physical examination and blood investigations were within normal limits. CECT abdomen showed heterogeneously enhancing renal lesion involving upper calyx, measuring 13x6x5.5cm extending into pelvis upto pelviureteric junction with possibility of neoplastic etiology. So, she underwent right radical nephrectomy.

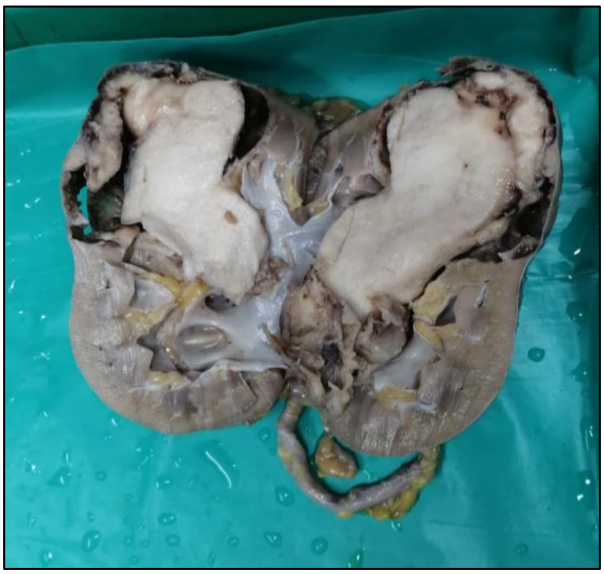

Fig. 1: Nephrectomy specimen with upper pole showing lesion

Right nephrectomy specimen showed a solid mass, measuring 11x6x5cm, involving upper pole and pelvis cut section tan coloured, with focal cystic areas.

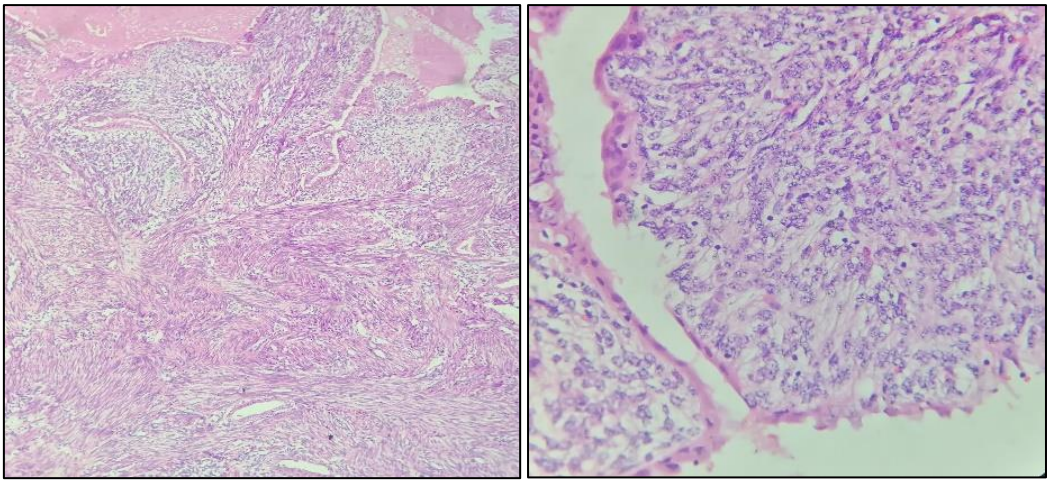

Fig. 2 & 3: H&E sections showing ovarian like stroma and hobnailing

Histopathological evaluation showed a neoplasm composed of variable sized cysts and glands separated by stroma. Cysts and glands lined by columnar to cuboidal cells showing focal stratification and hob nailing. Stroma is cellular composed of spindle cells arranged in fascicles and diffusely.

Immunohistochemistry showed strong positivity SMA, Desmin CD10 and focal positivity for ER, PR – in Mesenchymal cells. Epithelial cells showed focal positivity for Calretinin, WT1, Inhibin was negative and Ki67-5%. The diagnosis of MESTK was then established.

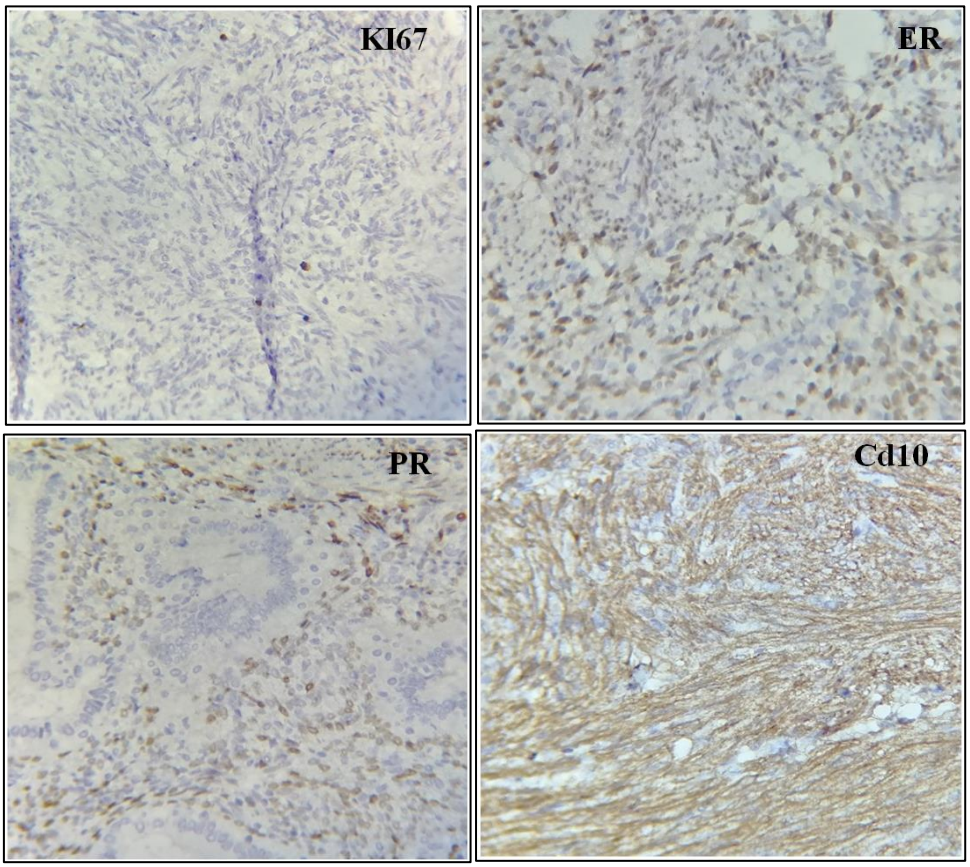

Fig. 4, 5, 6 & 7: Immunohistochemistry showing low ki67, ER, PR- positivity, CD 10 positivity

Case 2– 32 year old female, with history of haematuria for one week. Lab investigations were within normal limits. CECT abdomen showed a heterogenous lesion measuring 9.9x7.9x7.1cm in

interpolar region of left kidney extending to lower and upper pole with possibilities of angiomyolipoma and cystic nephroma were considered as differentials.

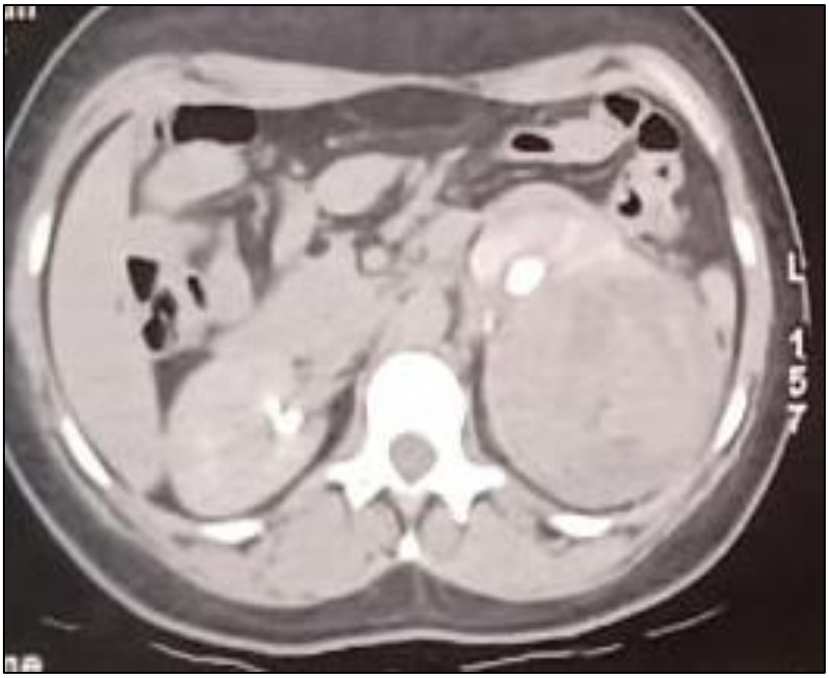

Fig. 8: CECT showing left renal mass

Left radical nephrectomy specimen showed a mass measuring 10x7.5x5.5cm, cut section yellowish to grey white fleshy with lobulations, areas of haemorrhage and necrosis, with periphery showing areas of cystic degeneration.

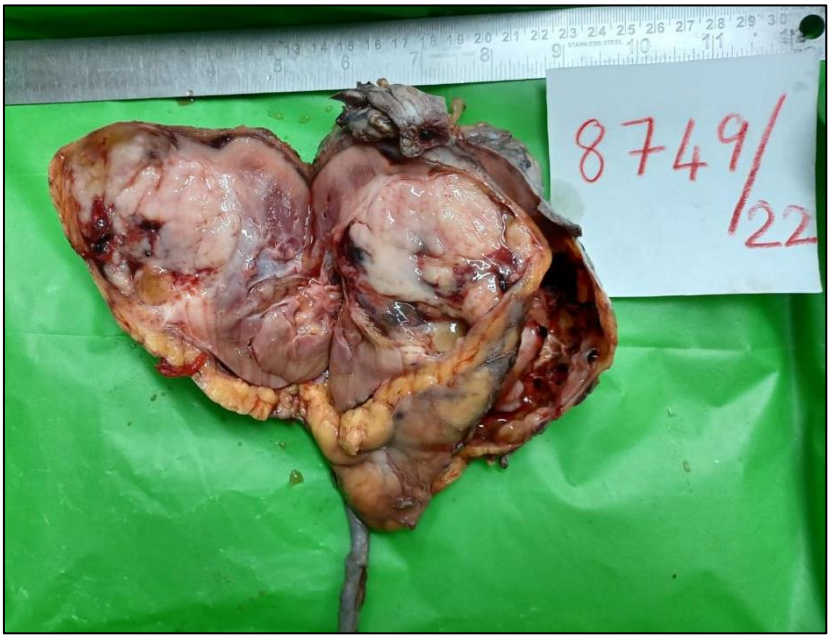

Fig. 9: Left radical neprectomy showing lesion

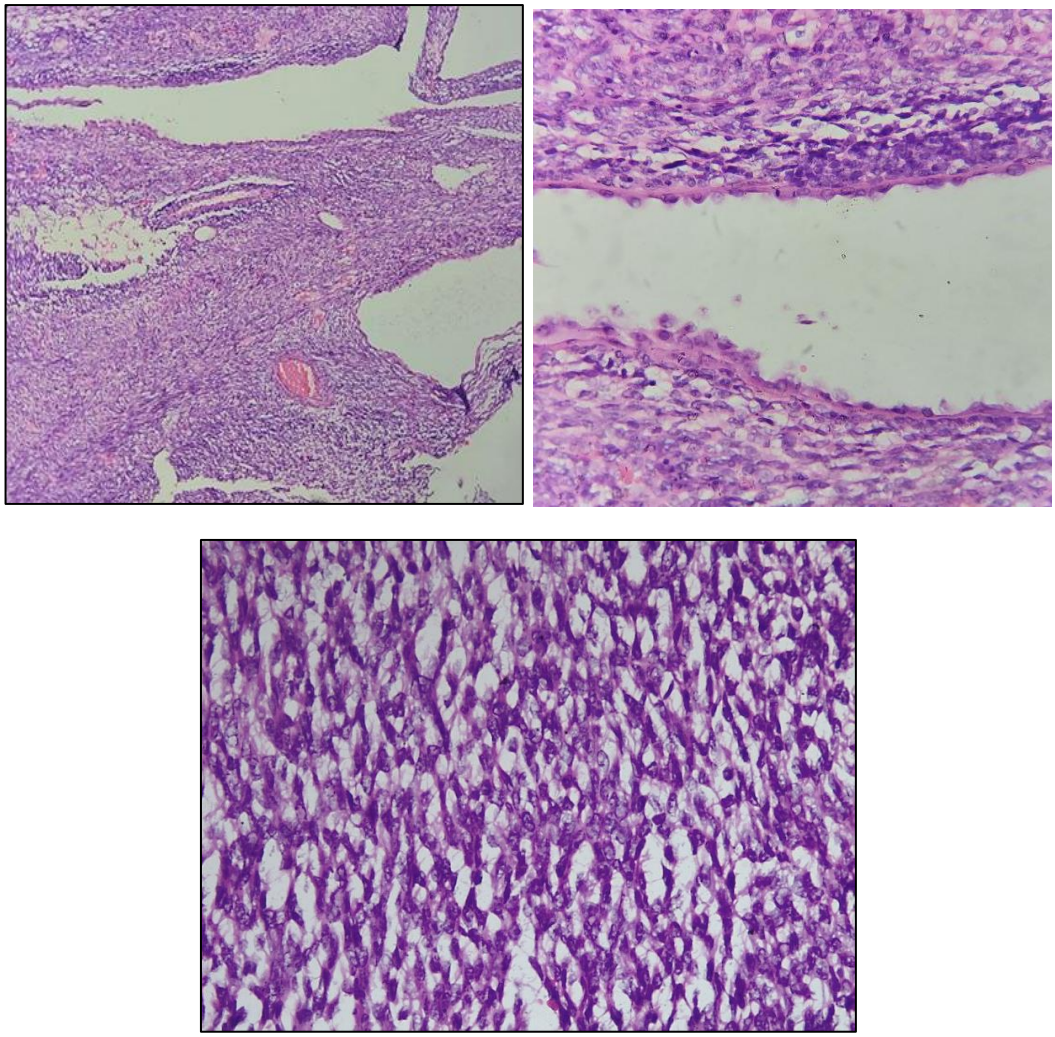

Fig. 10, 11 & 12: H & E sections showing ovarian like stroma, hobnailing and sarcomatous area

Histopathological evaluation showed a circumscribed neoplasm with solid areas and cystic spaces. Solid area showed spindle cells with sheet/herring bone pattern and increased mitotic activity 6-8/hpf. Collapsed cystic area showed epithelial lining with low mitotic activity and hob nailing of nuclei and ovarian stroma like component in cyst wall.

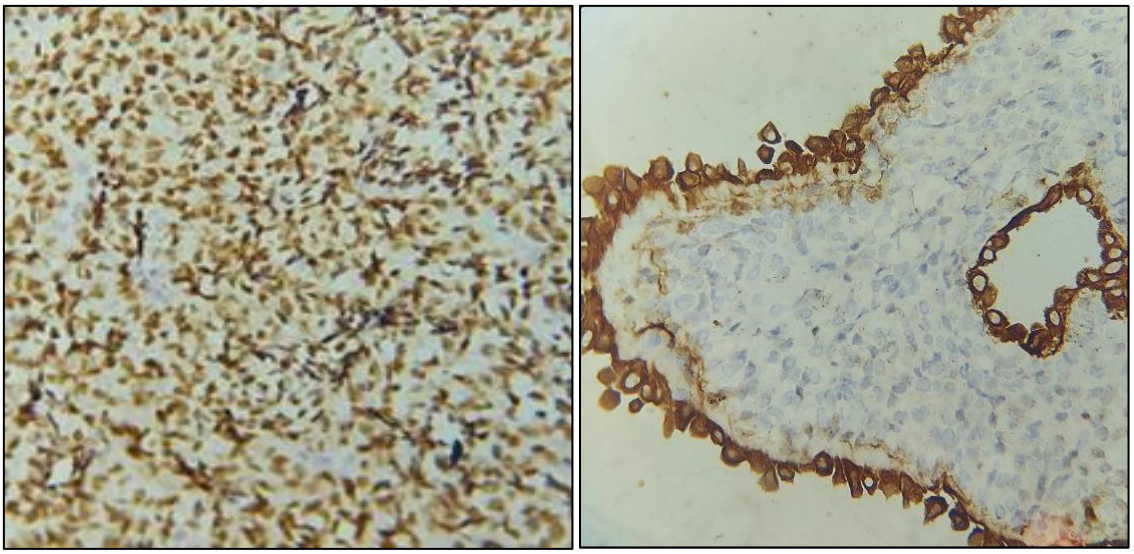

Fig. 13 &14: Immunohistochemistry show TLE1 positivity and CK postivity in epithelial component

Immunohistochemistry showed CK positivity in epithelial component and diffuse nuclear positivity of TLE1 in stromal component. ER is negative and Ki67 is >50%.

DISCUSSION

MEST comprise a rare and distinctive kidney tumour composed of both epithelium and stroma with solid and cystic architecture that is more common in women than in men (6:1, respectively). In general, patients are adults, with a mean age of 45 years with rare male or paediatric cases [3]. Most of the female patients were middle-aged peri- menopausal to older women, this implies that a disturbed hormonal environment contributes to the pathogenesis of MESTK; in our case patients had regular menstrual cycles and no history of hormonal therapy. The most common clinical presentations of MESTK include palpable abdominal mass, flank pain, and hematuria. In our case also both patients had multiple episodes of hematuria.

Radiologically, MESTK appears as well-circumscribed multi-septate cystic mass with thick or thin septa and solid components on both CT and MRI scans, so mimic complex renal cyst or cystic nephroma. In our cases also similar radiological findings were observed.

Generally, MESTK is a centrally located, circumscribed, solid-cystic kidney lesion that frequently extends into the renal pelvis. A partial or complete capsule is often present. The tumor infrequently shows an infiltrative border. Our cases also showed solid and cystic circumscribed lesions extending to renal pelvis.

Histologically, MESTK is a dimorphic tumor composed of cysts and tubules embedded in the spindle cell stroma. Both components of tumor stromal and epithelial are neoplastic and probably arise from a common cell of origin. Epithelial component is composed of clusters of tubules or cystically dilated glands with cuboidal/columnar lining with some showing hobnailing of nuclei [4]. Our cases also showed cystic spaces lined by cuboidal epithelium with hobnailing and low mitotic activity. The stromal component may resemble ovarian stroma, both morphologically and immunohistochemically. Our cases also showed similar ovarian like stroma with low mitotic activity but one with malignant transformation showed spindle cells with sheeting/herring bone pattern and increased mitotic activity 6-8/HPF.

Immunohistochemically, the epithelial components are usually positive for epithelial membrane antigen and cytokeratin. Spindle cells usually show diffusely and strongly positive immunostaining with desmin, smooth muscle actin, and vimentin. In addition, there is a high frequency of estrogen and progesterone receptor present in the nuclei of the spindle cells in benign cases [5]. Although the expression of focal progesterone receptors has been described in malignant MEST, all cases of malignant MEST were negative for estrogen receptors, which supports the idea that estrogen hormonal milieu has no impact on the pathogenesis of malignancy. In our report one case showed ER, PR, CD10 positivity in stromal cells and the one with sarcomatous differentiation showed CK positivity in epithelial component and diffuse nuclear positivity for TLE1 in stromal component, ER was negative and Ki67 was >50%.

To facilitate the diagnosis of malignant MEST, Jung et al. proposed diagnostic criteria to be fulfilled: (a) the epicentre of the tumour should be in the kidney; (b) clear-cut evidence of benign epithelial and stromal components with tubules or cysts lined by bland epithelial cells and a spindle-cell stroma resembling that of an ovarian-type stroma; (c) morphologically malignant components should be intimately associated with benign counterparts; and (d) primary renal sarcoma or metastases should be ruled out.

The differential diagnosis includes adult nephroblastoma, cystic nephroma, leiomyosarcoma, synovial sarcoma, and sarcomatoid renal cell carcinoma. The lack of cellular anaplasia in first case rules out the sarcomatous tumors but second case needed immunohistochemistry for differentiation. The absence of blastema excludes nephroblastoma, and cystic nephroma typically shows a thin and fibrous stroma [6].

MEST have been treated with surgical excision, and a few cases of recurrence, malignant transformation, and metastases development have been reported. There is limited data about systemic treatment but some malignant MESTs seemed to be chemosensitive and responded to doxorubicin and ifosfamide [6].

CONCLUSION

MESTK is a rare clinical entity. It is generally considered to be a benign tumor with good prognosis, but there is malignant potential. MESTK should be considered as a possible diagnosis in cases of cystic renal mass, especially in peri-menopausal women or those who have received hormonal therapy [7]. In benign cases surgical excision are curative but malignant cases show aggressive course of disease so treated with radical nephrectomy with extended lymphadenectomy and some cases seemed to be chemosensitive.

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