Mixed Epithelial Stromal Tumor of the Kidney (MESTK): A Case Report

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Abstract

Mixed epithelial stromal tumor of the kidney (MESTK) is a newly introduced and rare kidney tumor subtype; approximately 100 cases have been reported.

We report a case of MESTK with detailed clinicopathological findings, presented to emergency department with gross hematuria and clot retention managed by radical nephrectomy.

Although MESTK are benign tumors, they cannot be distinguished from other malignant neoplasms until after nephrectomy due to lack of typical radiological features. They can cause serious sequelae; including gross hematuria and serious drop in hemoglobin as the case we present here shows. This is why it is important to early diagnose and treat this condition.

This case highlights the importance of early introduction of imaging in patients presenting with gross hematuria. Failure to diagnose this lesion as the underlying cause of hematuria may lead to recurrence and malignant transformation.

Keywords: Tumors, Kidney, Case Report.

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Introduction

Mixed epithelial stromal tumor of the kidney (MESTK) is a newly introduced and rare kidney tumor subtype, which was included in the 2004 World Health Organization Classification of Tumors.[1]

To our knowledge approximately 100 cases have been reported[2], with most of these reports focusing on the pathological and radiological features of the tumors.

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The lesion has strong female predilection, with a 6:1 predominance of women over men[3],the tumor is typically cystic and solid, which is characterized by a biphasic proliferation of stromal cells with an epithelial component(4,5).

Patients usually present with a palpable abdominal or flank mass, flank pain, and/or hematuria. In a recent study, most of the case were incidental findings. Imaging studies are not diagnostic but reveal a centrally located solid or solid and cystic mass in most cases.(6)

We report a case of MESTK with detailed clinicopathological findings, presented to emergency department with gross hematuria and clot retention.

Case Presentation:

A 38 year old female patient, previously medically healthy nonsmoker, presented to emergency department with severe gross hematuria and clot retention.

On examination, the patient was ill-looking, afebrile, with normal blood pressure. Laboratory findings showed drop in hemoglobin (Hb) from 13mg/dL to 10 mg/dL.

The patient had recurrent attack of gross hematuria over the past 3 months and visited many hospitals and was diagnosed to have left complex renal cyst. At that time she was treated conservatively.

A urinary tract computed tomography with contrast (CT) scan showed parynchymal cystic lesion with enhancing septations at the upper pole of the left kidney 2.7*3.6*3.5 cm in dimensions (Bosniak classification 3). (Fig. 1)

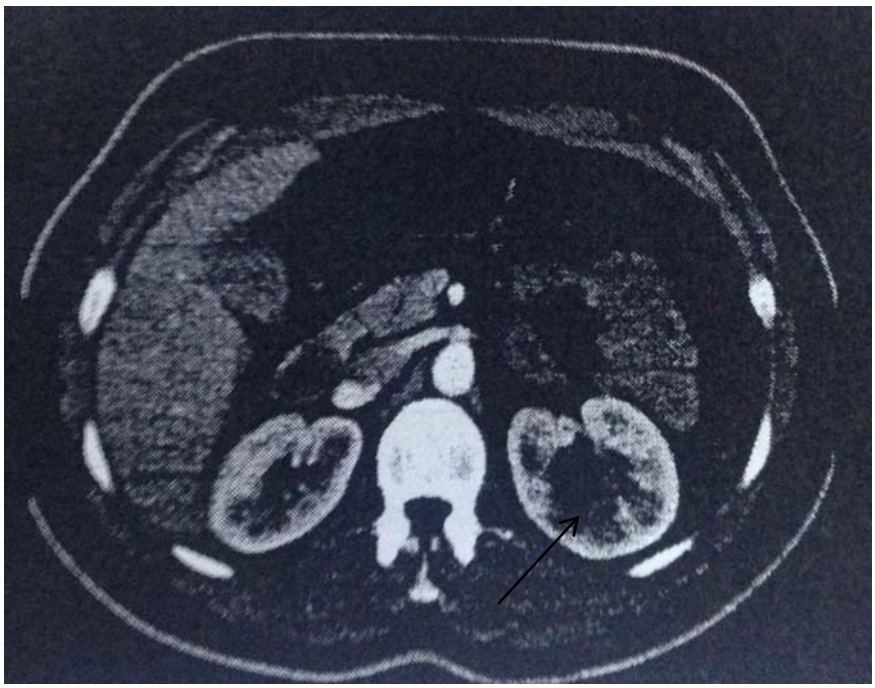

Figure 1: Computed tomography showing parynchymal cystic lesion with enhancing septations (arrow) at the upper pole of the left kidney 2.7*3.6*3.5 cm (Bosniak classification 3)

Patient was admitted as a case of clot retention, for management and further

evaluation. Cystoscopy and clot evacuation carried at that time revealed blood coming out through the left ureteric orifice with no bladder pathology.

So the plan was to go for left nephrectomy which was done through left flank incision. And tissue sent to pathology.

The patient's postoperative period was unremarkable; she recovered fully and then was discharged home.

Pathology proved that the mass was a benign mixed epithelial stromal tumor. Sections of the renal tumor represent a solid and cystic tumor with predominance of stromal elements in the form of benign smooth muscles, ovarian like stroma and fibroblasts. Embedded in the stromal elements are many variable sized cysts lined partly by ciliated columnar epithelium and in some areas by hobnail cells. Stroma showed strong positive staining for progesterone receptor (PR) and focally for estrogen receptor (ER). The tumor negative for HMB45. (Fig. 2-A, B, C, D).

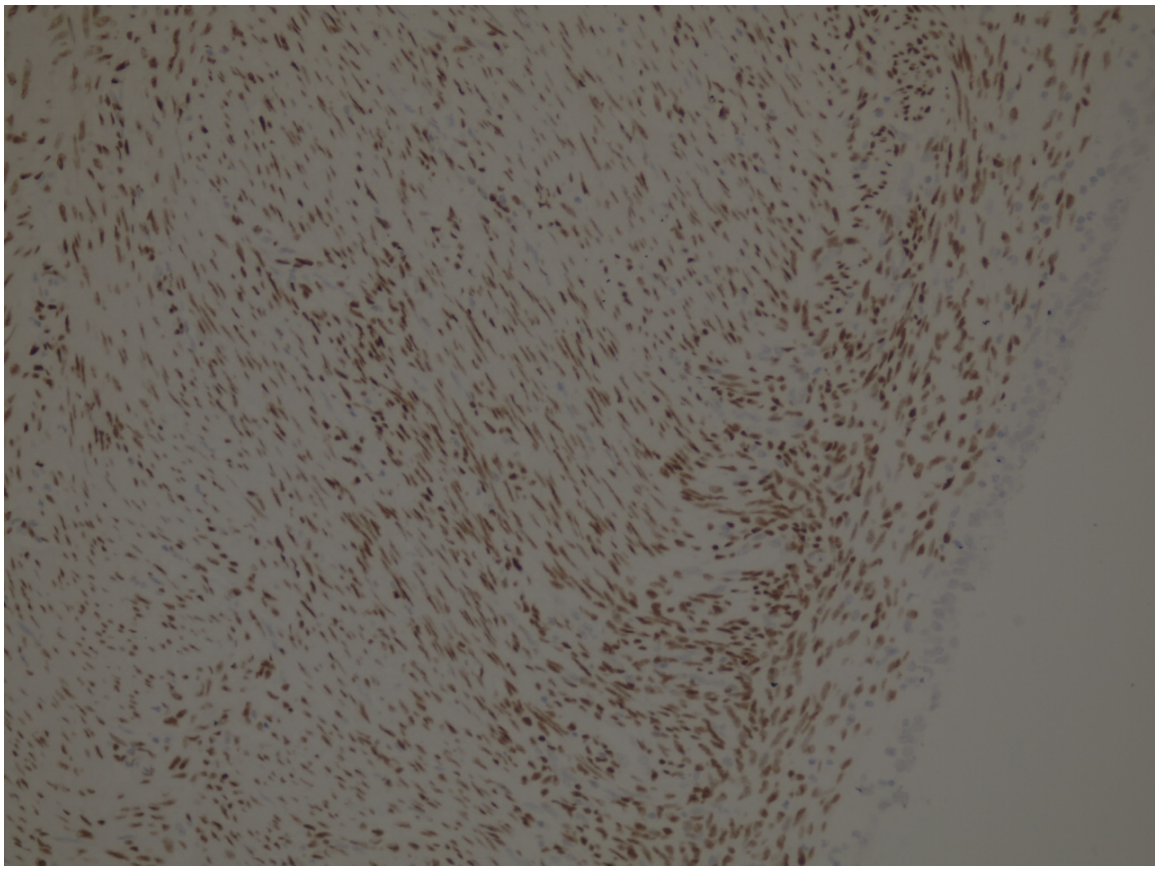

Figure 2: A: ER immunostaining is positive for ovarian like stroma. (200x magnification)

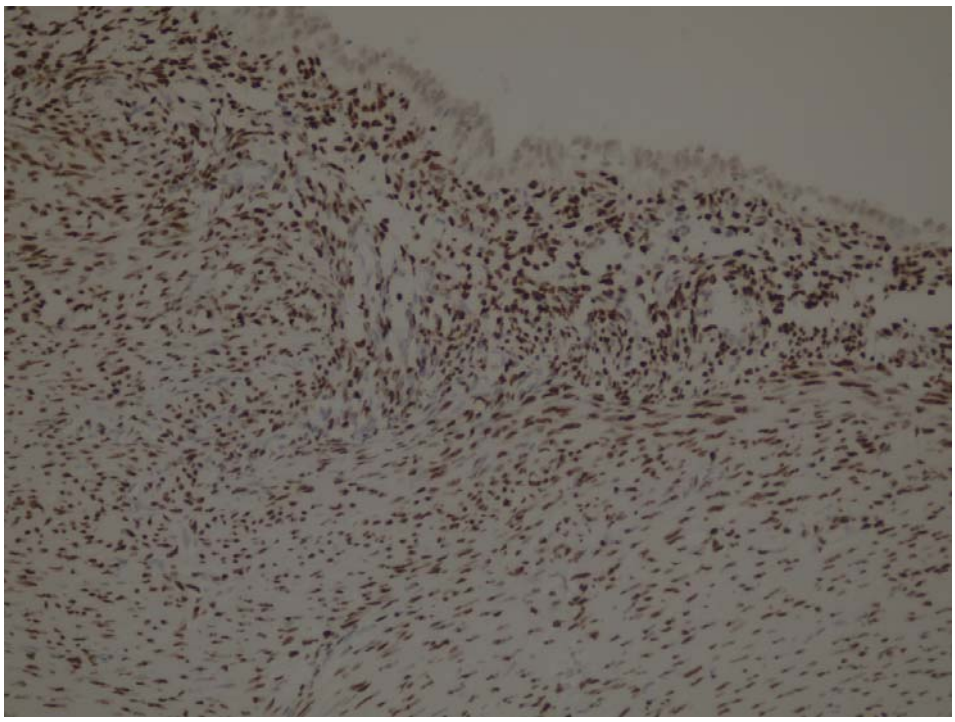

Figure 2. B: PR immunostaining is positive for ovarian like stroma. (200x magnification)

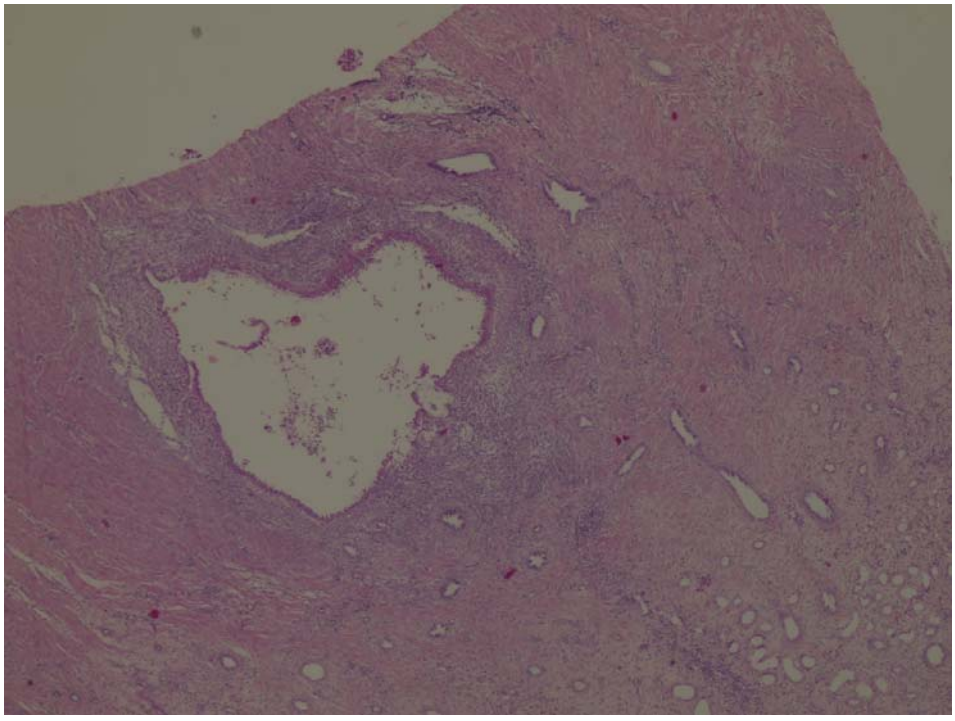

Figure 2.C: Cystic elements lined by ciliated columnar epithelium. (40x magnification)

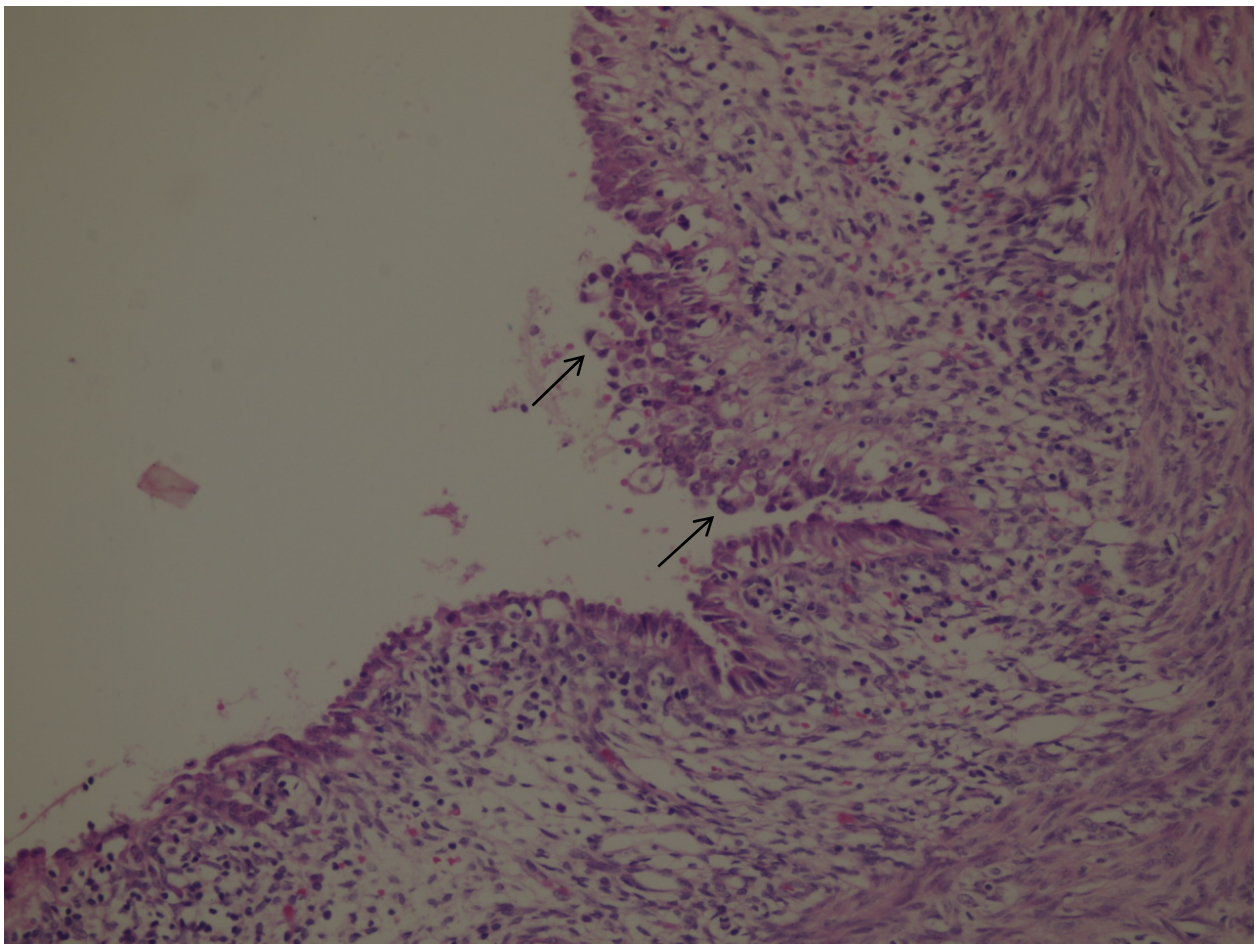

Figure 2. D: Cystic elements lined by ciliated columnar epithelium featuring hobnailing of the nuclei (arrows). (200x magnification)

Follow up:

Over a follow-up, of 3 years, the patient had no urinary symptoms, doing well and CT scans were normal.

Discussion:

Mixed epithelial stromal tumor represents a recently described tumor entity of the kidney of unknown etiology. Typically the tumor presents in perimenopsal women as a combined solid and cystic tumor mass. The mean age of clinical presentation is about 45 years[7].

Patients usually present with nonspecific symptoms, such as flank pain, hematuria, or symptoms primarily suggestive of urinary tract infection. The average tumor size is 6 cm in diameter with a range from 2-24 cm. The most popular postulation for its origin is based on hormonal disturbances, which is typically seen in perimenoposal women or which may be caused by hormonal replacement therapy or oral contraceptive pills. Thus hormonal imbalance might induce the proliferation of a misplaced immature or fatal mesenchyme, which harbours the capacity for a dual epithelial and mesynchymal differentiation. This theory is supported by the observation of the expression for the estrogen and

progesterone receptor within the stromal cells by nearly all cases of MESTK.

In addition, this hypothesis is also supported by a study showing that most of the affected women had a long history of treatment with estrogen. And the only male patient reported so far had a history of prostate cancer for which he was treated with hormonal therapy for 7 years.[7-10]

Most cases described so far showed a benign nature without tumor recurrence. However a case of malignant transformation to a sarcomatoid carcinoma and several cases of local recurrence of a malignant stromal component with a dismal clinical course have been described.[7,8]

MESTK demonstrates confusing morphological overlap to cystic nephroma, both types show some similarities including sex predominance, age distribution and morphological appearance of the epithelial and stromal component, as well as immunohistochemical staining. Therefore, there are some variations with a higher prevalence of stromal to epithelial ratio, prominent ovarian stroma, smaller cysts more common in MESTK. In contrast, larger cysts with only thin septa (lesser than 5 mm) and in addition a lower stromal to epithelial ratio are typically observed in cystic nephroma. However, the presence of ovarian like stroma and mullerian related immunohistochemical markers raise the possibility that these tumors might originate from mullerian remnants displaced during embryogenesis.[11]

Since some tumors have gross and microscopic features intermediate between cystic nephroma and MESTK, it is considered that both tumors might represent different morphological variants of the same tumor entity. Therefore, Turbiner et al. (2007) proposed to summarize both tumors under the unifying term "Renal epithelial and stromal tumor (REST)". However, this proposal is primarily based on the observation of an ovarian-like stroma in both tumor entities. But since this kind of stroma is also observed in obstructed kidneys, even without neoplasia, some authors consider the stromal differentiation as a form of reactive metaplasia. However, since there are no specific molecular markers for the differentiation of MEST from cystic nephroma and there are no further clues to the origin of both tumor entities, a definitive classification remains outstanding.[12]

MESTK represents a rare benign tumor of the kidney that should be distinguished from other cystic renal neoplasms. Prognosis of this tumor is favorable in nearly all cases published so far. Only rare cases of malignant transformation have been published. In summary, MESTK is a benign, mostly cystic tumor predominantly found in middle aged premenopausal women. Knowledge of this certain but rare tumor entity is important, since in most cases conservative surgery with preservation of kidney function is the therapy of choice.

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الورم الكلوي اللحمي الظهاري المختلط

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الملخص

الأورام الظهارية اللحمية المختلطة هي أورام نادرة تم إدراجها حديثا ضمن تصنيف منظمة الصحة العالمية للأورام 2004.
معظم الحالات التي تم تقريرها والتي على حد علمنا لا تتجاوز المئة حالة كانت حميدة على الرغم من وجود بعض الحالات الخبيثة.
يصعب تشخيص مثل هذه الحالات بالفحص السريري والصور الشعاعية الملونة او التلفزيونية ولكن التشخيص النهائي يكون بواسطة استئصال الورم ودراسة الخزعة مخبريا.
وحيث أن عددا قليلا من الحالات تم تقريره فسوف نقوم بوصف هذه الحالة وذكر الأعراض التي عانت منها المريضة وطرق تشخيصها وعلاجها لإتاحة الفرصة لمقارنتها بحالات اخرى مشابهة.

الكلمات الدالة: الأورام، الكلية، دراسة حالة.