

Mixed epithelial stromal tumor of the kidney

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ABSTRACT

In recent years, a rare distinctive kidney tumor composed of a mixture of stroma and epithelium with solid and cystic architecture has been recognized, which has to be distinguished from other renal neoplasms. The term mixed epithelial and stromal tumor was first introduced by Michal and Syrucek in 1998.^[1] The vast majority of cases show a benign course without tumor recurrence. Here, we present a case of this entity, found incidentally.

Key words: Adult type of mesoblastic nephroma, mixed epithelial stromal tumor

INTRODUCTION

In recent years, a rare distinctive kidney tumor composed of a mixture of stroma and epithelium with solid and cystic architecture has been recognized. These tumors, published earlier under different nomenclatures, such as “adult type of mesoblastic nephroma,” “cystic hamartoma of the pelvis,” “adult type of cystic nephroma,” “leiomyomatous hamartoma,” and “solid and cystic biphasic tumor of the kidney,” represent, in fact, the same neoplasm, with a spectrum of differentiation. The name “mixed epithelial and stromal tumor of the kidney” was first introduced by Michal and Syrucek in 1998. Here, we present a case of this entity, found incidentally.

CLINICAL HISTORY

A 35-year-old female patient presented with intermittent right flank pain for 1 year. She denied any hormonal, drug, or surgical history. Physical examination was unremarkable. Her routine blood

investigations were normal. Routine urine analysis revealed no hematuria. Urine cytology was negative for malignancy. Retrograde Pyelography (RGP) showed an obstruction of the proximal right ureter due to a tumor projecting to the lower-right renal pelvis [Figure 1a]. Abdominal computed tomography revealed a huge cystic heterogenic tumor about 10 cm in largest diameter occupying the entire right renal pelvis causing gross hydronephrosis of the right kidney [Figure 1b]. There was no evidence of lymph node or distant metastases. We planned for surgical exploration with a diagnosis of transitional cell carcinoma of the right kidney.

On surgical exploration there was a grossly hydronephrotic right kidney with a well-circumscribed mass arising from the renal pelvis extending into the ureter. A right radical nephroureterectomy was performed. No postoperative complications were observed.

Pathologic findings

Gross examination of the cut surface of the right kidney revealed a polypoidal growth of 8 × 5.5 cm, which is solid cystic in appearance with the solid area showing papillary excrescences [Figure 2a and b]. The growth is seen arising from the renal pelvis with extension into the upper ureter. Histologically, the tumor was composed of large cysts, microcysts, and tubules. Large cysts lined by columnar and cuboidal epithelium with some are forming papillary tufts. Microcysts and tubules are lined by flattened, cuboidal, or columnar cells. Their cytoplasm ranges from clear to pale eosinophilic. The architecture of the microcysts varied from simple microcysts with abundant stroma between them, to densely packed clusters of microcysts to complex branching channels. The stroma consists of variably cellular proliferation of spindle cells with plump nuclei and abundant cytoplasm. Areas of myxoid stroma and smooth

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muscle cells also identified at places. No dysplasia, increased mitotic activity, or tumor necrosis as signs for malignant transformation were observed. By immunohistochemical investigations, the epithelial cells demonstrated expression for pan-cytokeratin, whereas the stromal cells demonstrated co-expression for mesenchymal marker, such as vimentin. Nuclei of the stromal cells also showed react with antibodies to estrogen and progesterone receptors [Figure 2c–h].

Follow-up

The patient is alive and well after 1 year of primary operation.

DISCUSSION

Mixed epithelial and stromal tumor (MEST) represents a recently described tumor entity of the kidney of unknown etiology.^[2] In the past other synonyms, such as cystic hamartoma of the renal pelvis, adult mesoblastic nephroma,

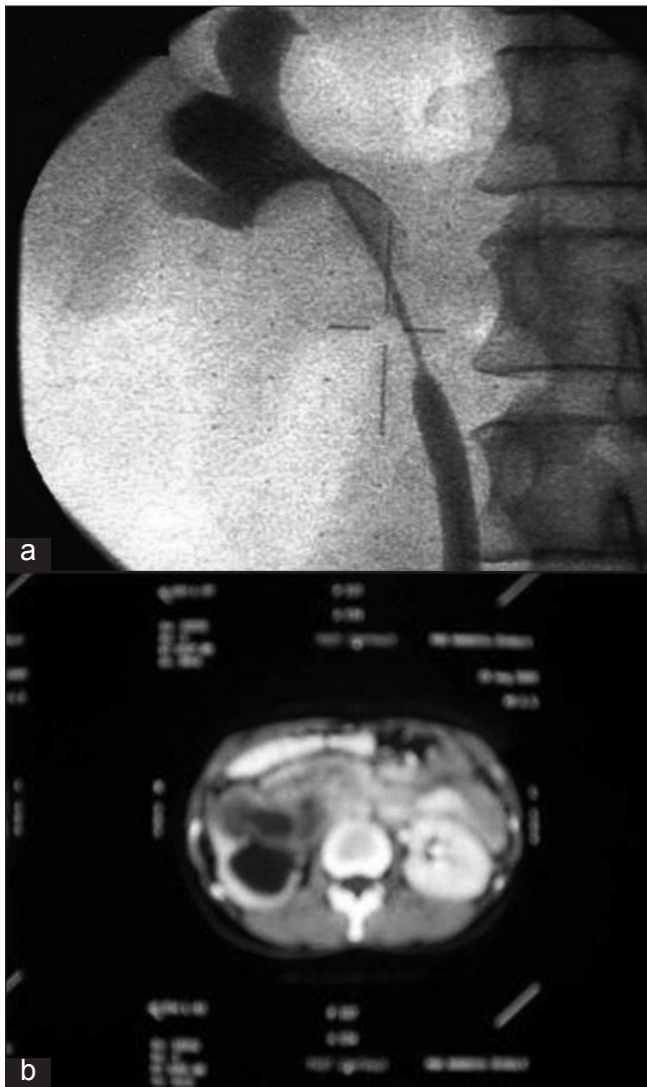


Figure 1: (a) Right Retrograde Pyelogram showing a filling defect in the renal pelvis causing hydronephrosis of the kidney; (b) contrast-enhanced computed tomography showing a large tumor filling the renal pelvis causing hydronephrosis of the right kidney

and cystic nephroma with “cellular” or “ovarian- type” stroma were applied.^[2] About 50 cases have been reported in the literature so far.^[2,3,5]

Typically, the tumor presents in perimenopausal woman as a combined solid and cystic tumor mass. The mean age of clinical presentation is about 45 years.

The patients usually present with nonspecific symptoms, such as flank pain, hematuria, or symptoms primarily suggestive of genitourinary infections. The mean tumor size at primary diagnosis is about 6 cm in diameter.^[2,5] Our patient had a typical clinical presentation as described in the literature. The gross and microscopic features are also similar to those reported earlier.

Nearly all cases described so far demonstrated a benign course without tumor recurrence. However, four cases of aggressive mixed epithelial stromal tumor have been described in the literature so far.^[4] Three patients with MEST with local recurrence of tumor showing a fatal course have been described. In two patients, the recurrent tumor was composed exclusively of malignant transformed stroma. One case displayed malignant transformation of MEST to a sarcomatoid carcinoma with heterologous differentiation. Our patient had no evidence of recurrence or metastases and is on follow-up for 8 months.

The differential diagnoses include renal tumors, which can show at least a partly cystic morphology, such as partially cystic differentiated nephroblastoma, multilocular cystic renal carcinoma, angiomyolipoma with epithelial cysts, or in rare cases synovial sarcoma.^[2,5]

Since some tumors have gross and microscopic features intermediate between cystic nephroma and MEST, it is considered that both tumors might represent different morphological variants of the same tumor entity.^[2,5] Therefore, Turbinder *et al.* (2007) proposed to summarize both tumors under the unifying term “renal epithelial and stromal tumor” (REST).^[2] 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.

CONCLUSIONS

Mixed epithelial and stromal tumor represents a distinctive 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, MEST represents a benign mostly cystic tumor of the kidney, which is predominantly observed in middle-aged, perimenopausal women. Knowledge of this certain but rare tumor entity is important, since in most cases

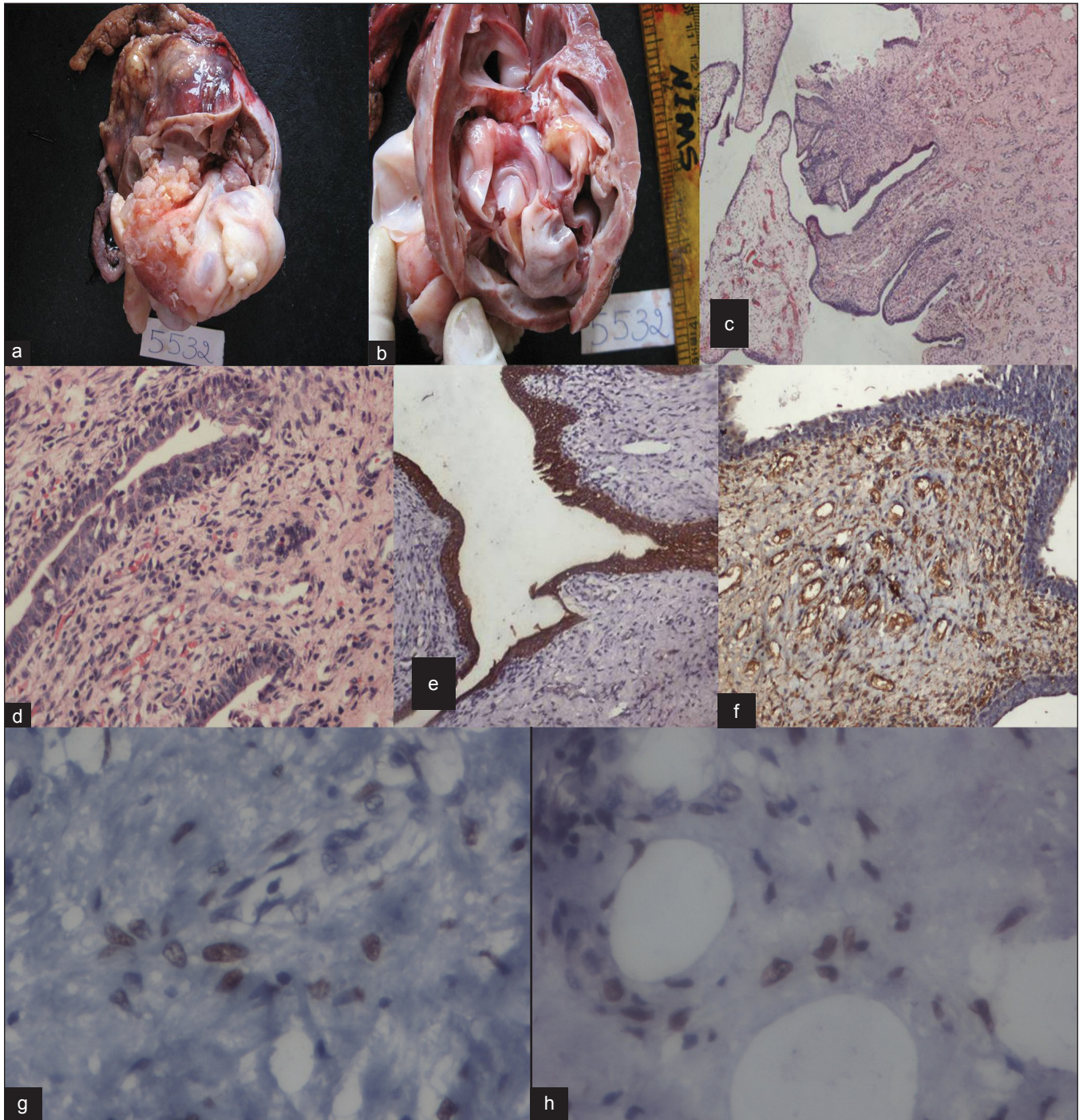


Figure 2: (a) Gross photograph of the tumor showing predominantly solid area with papillary excrescences; and (b) cystic areas; (c) low magnification photograph of the tumor showing both epithelial and mesenchymal components (HandE, ×40); (d) higher magnification shows cuboidal cell lining of epithelium with underlying ovarian like stroma (HandE, ×400); (e) intense reactivity for pancytokeratin in epithelial lining of tumor cells (HRP Polymer Cytokeratin, ×100); (f) vimentin reactivity in ovarian-like mesenchymal stroma (HRP Polymer Vimentin, ×100); (g) Estrogen Receptor shows nuclear positivity in spindle cells (IHC, ×400); (h) Progesterone Receptor shows nuclear positivity in spindle cells (IHC, ×400)

conservative surgery with preservation of kidney function is the therapy of choice.

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