Mixed epithelial and stromal tumor of the kidney with polypoid component extending into renal pelvis and ureter

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Abstract

Mixed epithelial and stromal tumor (MEST) of the kidney is an unusual benign neoplasm that predominantly occurs in middle-aged females. It typically appears as a well-circumscribed multiseptate mass with solid components on computed tomography (CT) or magnetic resonance image (MRI), reflecting its characteristics of an admixture of stromal proliferation and epithelial cells consisting of multiple cysts. We present a rare case of 61-year-old woman with MEST, which manifested as a multilocular cystic mass with a polypoid component protruding into the renal pelvis and ureter. To our best knowledge, this is the first case of MEST extending into the ureter.

Keywords: Mixed epithelial and stromal tumor, MEST, kidney, ureter, CT, MRI

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Mixed epithelial and stromal tumor (MEST) is a rare, basically benign neoplasm of the kidney that has recently been recognized as a distinctive entity (1). It mostly occurs in middle-aged women and is associated with estrogen exposure (2). The histological characteristic of MEST is an admixture of spindle cells resembling ovarian stroma and epithelial elements (1, 2). Approximately 100 cases have been reported in past literatures; however, most of the reports are focused on their pathologic features (2, 3). Reports of its radiologic characteristics, including computed tomography (CT) and magnetic resonance imaging (MRI), have been still scarce (4–8). We present an unusual case of MEST of the kidney with a polypoid component extending into the renal pelvis and the proximal ureter.

Case report

A 61-year-old postmenopausal woman presented with gross hematuria for two weeks. The physical examination was unremarkable. She had no relevant family history or medical history other than mild obesity. Her routine blood investigation was normal with no elevated serum tumor marker. Ultrasonography revealed the presence of a left renal mass in the interpolar region extending into the renal pelvis. Exfoliative urine cytology was class 3. Excretory phase contrast-enhanced CT (Fig. 1a–c) showed a well-circumscribed multiseptate cystic mass of $44 \times 40 \times 45$ mm in the interpolar region of the left kidney with a polypoid solid component protruding into the renal pelvis and proximal ureter. Calcification was identified within the solid component. Retrograde ureterography (Fig. 1d) showed mild hydronephrosis due to the polypoid part of the tumor in the ureter. Dynamic MRI study (Figs. 2 and 3) demonstrated delayed contrast enhancement of the solid polypoid component and multiple septa.

Since the tumor was consistent with a Bosniak category IV cystic mass, we performed a laparoscopic left nephroureterectomy. Gross examination showed a well-marginated multilocular cystic mass with a polypoid solid component protruding into the renal pelvis and ureter (Fig. 4a). Histopathological analysis revealed an intermixture of epithelial elements consisting of varying sized glands and a proliferation of spindle cells resembling ovarian stroma. The polypoid lesion was superficially covered with normal urothelium. Complex glands with a single layer of flattened, cuboid or hobnail-appearing epithelium and a mild cellular atypia of swollen nuclei were marked in the

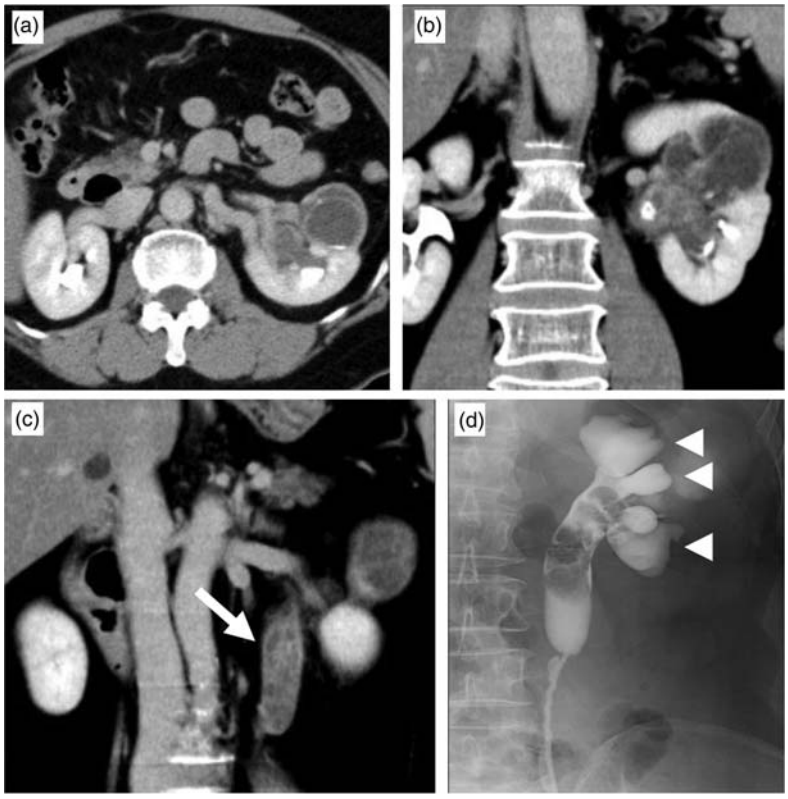

Fig. 1 (a) Axial and (b, c) coronal excretory phase contrast CT showing a multilocular cystic mass with a polypoid part continuously extending into the pelvis and ureter (white arrow); (d) Retrograde ureterography showing a polypoid tumor in the pelvis and proximal ureter and moderate hydronephrosis (white arrow heads)

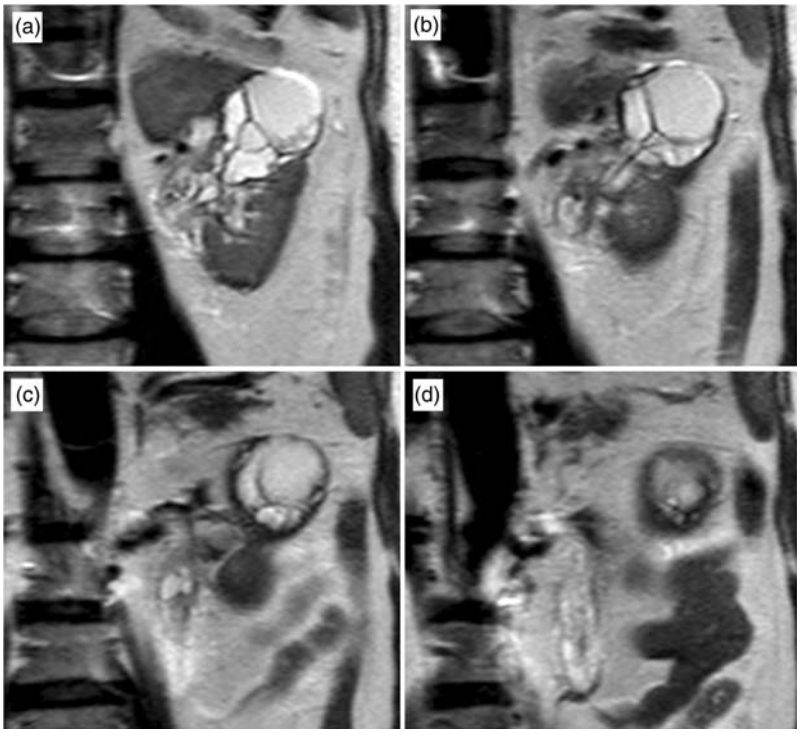

Fig. 2 (a, b, c, d) Continuous slices of coronal T2-weighted MRI showing a well-circumscribed multiseptate cystic mass with a polypoid compartment extending into the renal pelvis and proximal ureter

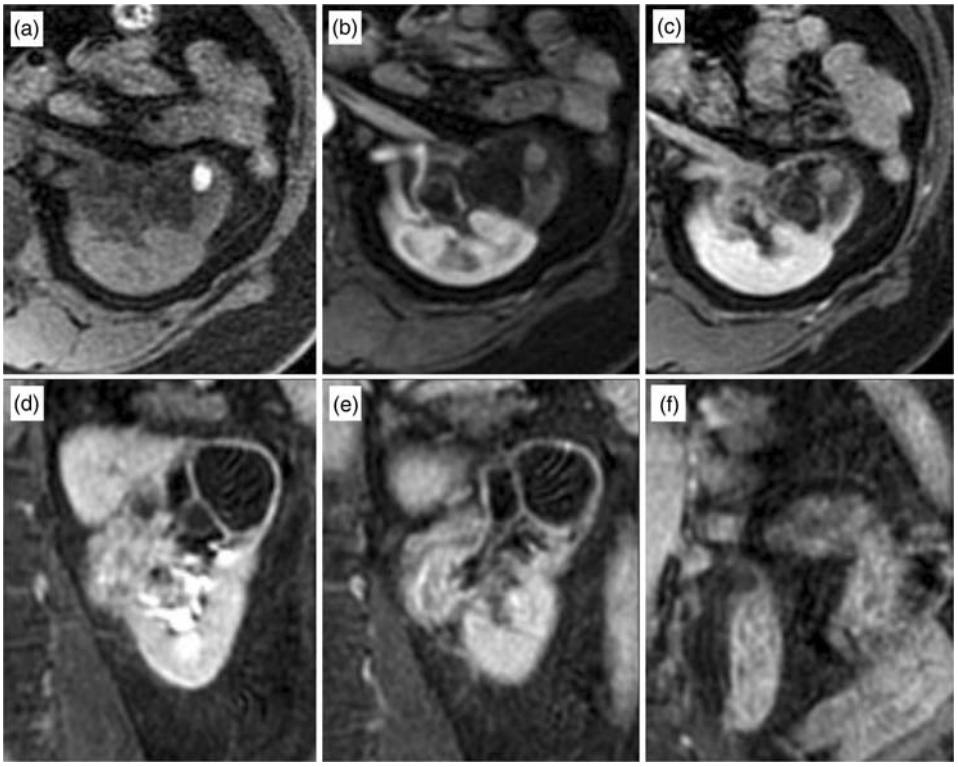

Fig. 3 (a) Axial pre-enhanced, (b) axial arterial phase, (c) axial medullary phase fat-saturated T1-weighted MRI. Dynamic MRI study demonstrates delayed enhancement in the polypoid lesion and septa; (d, e, f) Continuous coronal excretory phase fat-saturated T1-weighted MRI showing marked enhancement in the polypoid lesion

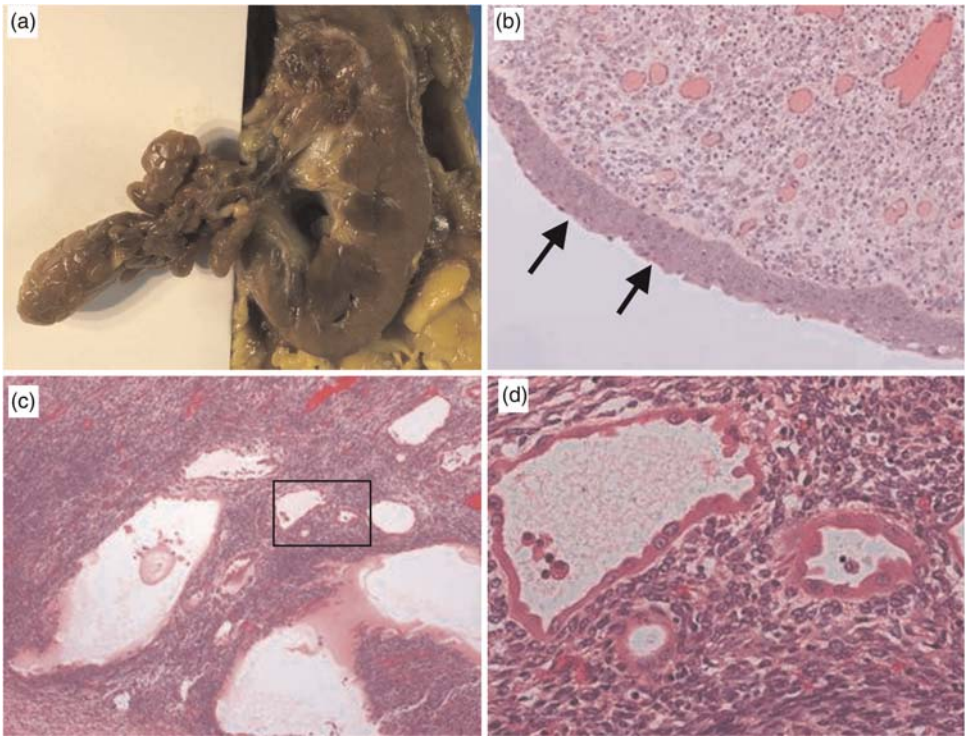

Fig. 4 (a) Gross specimen showing a cystic lesion with septa and a sharply marginated solid polypoid lesion; (b) Photomicrograph of the polypoid lesion showing proliferation of stroma, superficially covered with urothelium (black arrow) (hematoxylin and eosin stain; ×200); (c) Low-power-view and (d) high power-view of the cystic lesion demonstrating an admixture of spindle cells resembling ovarian stroma and epithelial elements consisting of varying sized glands (c, ×100; d, ×400; hematoxylin and eosin stain)

cystic lesion. The tumor was well-marginated but no capsule was found histologically. No malignant cells were found. Immunohistochemically, spindle cells expressed vimentin and alpha smooth muscle actine, and were partially positive for progesterone receptor, bcl-2, CD99, and desmin. Epithelial cells were positive for cytokeratin and epithelial membrane antigen, partially positive for vimentin, bcl-2 and negative for CD99, alpha smooth muscle actine, desmin, or progesterone receptor. The estrogen receptor was negative in both. On the basis of the morphologic and immunohistochemical findings a pathologic diagnosis of MEST was achieved.

The postoperative course of the patient was uneventful. There was no local recurrence or metastasis during 10-month follow-up.

Discussion

MEST of the kidney has been recognized as a distinctive entity since first described in 1998 (1). This rare neoplasm is characterized by a mixture of stromal solid areas and epithelial elements; previously it used to be referred to as a multilocular cyst with ovarian stroma, adult type mesoblastic nephroma, cystic hamartoma of the renal pelvis, or leiomatous renal hamartoma. It predominantly occurs in middle-aged or older women and is strongly associated with estrogen exposure. It is basically a benign neoplasm, but several cases of malignant transformation in the stromal component resembling sarcoma have been reported (9–14). Approximately 100 cases have been reported in past literatures, however, most reports mainly focused on their pathologic features while radiologic reports have been still scarce (4–8).

In a recent series of radiologic case reviews, MEST of the kidney have had a feature of a well-circumscribed multiseptate cystic mass with solid components and thick or thin septa on both the CT and MR imaging (4–8). The septum is more easily detectable and clearer by MRI. A delayed enhancement of solid parts and septa may also be seen as reflecting on the stromal element. A morphologic feature of MEST protruding into renal pelvis have been reported in a few cases (7, 8), however, MEST extending into the proximal ureter is extremely rare.

The differential diagnosis includes adult cystic nephroma (ACN), complex renal cyst, angiomyolipoma with epithelial cysts, and multilocular cystic renal cell carcinoma (RCC). However, it is often difficult to differentiate among them. Although MEST of the kidney and ACN were categorized as separate diseases in the 2004 WHO classification, both tumors have turned out to share many clinical, pathologic, and immunohistochemical features (15–17). The difference is only in their morphologic appearance; lesions with solid areas or cystic septa >5 mm are classified as MEST (17). Very recently, some pathologists tend to regard the two as different parts of the morphologic spectrum of the same disease, and have proposed renal epithelial and stromal tumor (REST) as a unifying term (16). A finding of the pelvic and urinary extension of our case of MEST may be associated with the similarity to ACN, which often protrude into the renal pelvis.

As MEST has characteristics of solid components or thicker septa, almost all MESTs demonstrate Bosniak category III or IV lesions (8). Differentiating MEST from multilocular cystic RCC is of great clinical relevance. According to a recent review, middle-aged women, exogenous estrogen exposure, delayed contrast enhancement, and renal pelvic origin tumor with negative urine cytology may help in the prediction of MEST (8). Regarding mild obesity as high estrogen exposure, all of them apply in our case. Surgical treatment is necessary considering the difficulty in differentiating the above, together with its unknown probability of potential malignant transformation.

In conclusion, we report a case of MEST with a polypoid compartment protruding into the renal pelvis and ureter. To the best of our knowledge, this is the first case of MEST extending into the ureter.

Conflict of interest: None.

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