Case report

Metastasis in mixed epithelial stromal tumour of the kidney: a rare presentation

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SUMMARY

Mixed epithelial stromal tumour of the kidney (MESTK) is a rare genitourinary tract tumour. MESTK is typically seen in perimenopausal women and rarely reported in men and children. MESTK has been included in the WHO renal tumour classification since 2004. Here, we present a case of 50-year-old female patient who underwent left radical nephrectomy for left renal mass. Postoperative positron emission tomography (PET) scan also showed nodal metastasis, for which, she was started on chemotherapy.

BACKGROUND

Mixed epithelial and stromal tumour of the kidney is recently recognised as a distinct neoplasm that should be distinguished from other common renal neoplasm. These tumours are relatively rare and show a female preponderance. Imaging studies are not diagnostic but reveal a solid or solid and cystic component in most cases. Histopathologically, these tumours reveal biphasic growth pattern comprising mesenchymal and epithelial elements with characteristic oestrogen receptor (ER) and progesterone receptor (PR) immune-reactive mesenchyme reminiscent of ovarian stroma. Malignant transformation, recurrence and metastasis are rare; however, recently a few cases of malignant mixed epithelial and stromal tumours have been reported.

CASE PRESENTATION

A 50-year-old female patient presented with complaints of intermittent left sided flank pain of 5-month duration. There were no other associated symptoms. Abdominal examination was unremarkable. Investigations revealed a left renal mass of probable malignant aetiology. Left radical nephrectomy was done. Histopathology revealed, 'mixed epithelial stromal tumour of left kidney' which on immunohistochemistry confirmed the diagnosis of 'mixed epithelial stromal tumour of the kidney' (MESTK). It was decided in the tumour board meeting to evaluate for distant metastasis. Positron emission tomography (PET) scan revealed subcentimetric metastatic lymph nodes. Patient was started on gemcitabine and carboplatin chemotherapy.

INVESTIGATIONS

Blood and urine investigations were normal. On ultrasonographic examination, lobulated mass lesion of size 4.5×3.5 cm arising from the left kidney was located. Right kidney was normal. Abdominal CT (figure 1) revealed a large, well defined, lobulated, heterogeneously enhancing mass lesion measuring 5.1×3.9 cm arising from the anterior aspect of the mid-part of the left kidney. A lobulated component extended into the renal pelvis and up to the pelviureteric junction causing resultant moderate dilatation of the pelvicalyceal system. There was compression and splaying of the superior and middle calyceal system of the left kidney. There was no evidence of abdominal or pelvic lymphadenopathy preoperatively.

Post surgery, specimen on gross examination showed a mass, predominantly cystic with few solid areas measuring 5×4 cm involving upper and mid-pole of left kidney without involvement of renal vessels. Cortical surface showed a bosselated area measuring 3×2.5 cm; cut surface showed papillary projection measuring 3×1 cm (figure 2). Microscopically (figure 3) it showed mixed epithelial and stromal component with low mitotic activity. The epithelial elements revealed tubulopapillary architecture with foci of cystic changes lined by low cuboidal to flat epithelium. Stromal component consisted of ovoid to plump spindle cells. On immunohistochemistry, Paired Box Gene 8 (PAX 8) highlights the epithelial component and stromal cells show diffuse nuclear staining for ER and PR, thus confirming the diagnosis of MESTK.

Postoperative PET scan revealed fluorodeoxyglucose avid (metabolically active) subcentimetric, aortocaval, periportal and precaval lymph nodes

Figure 1 A multicystic mass is seen arising on the anterior aspect of left kidney in the mid-region. Multiple enhancing septae are seen along with few areas along the margins.

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Figure 2 Gross specimen of nephrectomy showing tumour with solid and cystic appearance.

suggestive of metastasis (figure 4). CT/MRI guided biopsy was not feasible.

TREATMENT

Left sided radical nephrectomy was done. Postoperatively, PET scan was done and showed FDG avid (metabolically active) abdominal lymphadenopathy. Adjuvant chemotherapy (gemcitabine+carboplatin) was started in view of immunohistochemistry suggestive of mixed epithelial and stromal tumour with FDG avid (metabolically active) abdominal lymphadenopathy. Intravenous gemcitabine (1200 mg in 500 mL normal saline) and carboplatin (300 mg in 250 mL D5) were administered for eight cycles, along with a proton pump inhibitor and an antiemetic.

OUTCOME AND FOLLOW-UP

Patient is asymptomatic at the end of three cycles of adjuvant chemotherapy. We have planned a reassessment PET scan after eight cycles of chemotherapy.

DISCUSSION

MESTK is a rare kidney tumour described recently of unknown aetiology.¹ This term was first introduced by Michal and Syrucek,² before which these tumours were published with other synonyms, such as 'adult type of mesoblastic nephroma', 'cystic hamartoma of the pelvis', 'adult type of cystic nephroma', 'leiomyomatous hamartoma' and 'solid and cystic biphasic tumour of

Figure 3 Photomicrograph showing epithelial component in form of cysts and surrounding stroma resembling ovarian stroma (H&E, ×400).

Figure 4 PET-CT showing an FDG avid subcentimeter sized lymph node in the left para-aortic region near the operative site. Similar metabolically active lymph nodes were also seen in periportal and precaval regions (not shown).

the kidney'.³ MESTK is typically seen in perimenopausal women with mean age being 45 years^{4,5} and is rarely reported in men and children (6 females to 1 male ratio).^{5,6} MESTK has been included in the WHO renal tumour classification since 2004.

Usually the patient presents with symptoms like a palpable mass, flank pain, haematuria or urinary tract infection. MESTK are benign tumours but are difficult to distinguish from other malignant neoplasms due to lack of typical radiological features. Almost all cases of MESTK are benign. For its diagnosis either CT or MRI is necessary which show multicystic mass with solid components and thick or thin septae.⁷ Although these tumours are more commonly benign, there is a chance of presentation as distant metastasis.⁸ Only a few cases are presented with metastasis in the literature.⁹

Primary treatment of any renal malignancy is radical nephrectomy. In cases of nephron sparing surgery, renal nephrometry score and C-index are considered. Chemotherapy is given when there is any evidence of nodal or distant metastasis.

Learning points

- ▶ In perimenopausal women of middle age presenting with renal mass, mixed epithelial stromal tumour of the kidney should be kept in mind.
- ▶ Although it is a benign tumour, it has potential for malignant transformation.
- ▶ As it has typical histological features, correlation with histopathology and immunohistochemistry is important for further management.
- ▶ These tumours have good prognosis.
- ▶ Evaluation for distant metastasis should be kept in mind.

In our case, a female patient aged 50 years, presenting with symptom of flank pain, with malignant lesion in the kidney on CT, suggestive of the renal mass, on histopathology was diagnosed to be MESTK. As postoperative PET–CT showed FDG avid (metabolically active) abdominal lymph nodes, adjuvant chemotherapy was started.

Gemcitabine and carboplatin was started as, the response rate, time to treatment to failure and overall survival rate have been found comparable to methotrexate, vinblastine, doxorubicin and cisplatin combinations, with low toxicity in a multicentric trial.¹⁰ Carboplatin has emerged as a less nephrotoxic platinum analogue, with less neurotoxicity and preventable haematotoxicity. So, carboplatin was used in this patient.¹¹

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