CASE REPORT

Mixed epithelial and stromal tumour of the kidney masquerading as hydatid cyst: a rare case report from the developing world

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Abstract

Mixed epithelial and stromal tumour (MEST) is a rare entity with an incidence of 0.2% among renal cancers. It has strong predilection towards females with a 1:6 male to female ratio, the tumour is cystic with some solid component with biphasic proliferation of stromal and epithelial cells.

The case of a 37-years-old female is presented with right lumbar pain since 3 months. The family history was unremarkable. The routine workup revealed mild neutrophilia and borderline Echinococcus antibody titres. Ultrasound revealed a complex cystic lesion with a solid component in the right kidney. CT scan with contrast confirmed a multiloculated mixed density lesion with daughter cysts arising from the middle lobe of the right kidney. Initial diagnosis of renal hydatid cyst was established and she underwent partial nephrectomy with excision of the cystic mass. Surprisingly the histopathology revealed mixed epithelial and stromal tumour.

Keywords: Hydatid cyst, Mixed epithelial and stromal tumor, Echinococcus, Partial nephrectomy.

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Introduction

Mixed epithelial and stromal tumour of the kidney (MEST) is a very rare entity with a prevalence of 0.2% of all renal tumours. It usually occurs in females with the ratio being 1:6 male to female[1]. The tumour is composed of stromal cells with an epithelial component[2]. Mostly it is an incidental finding but sometimes patients may present with a flank mass, pain, and haematuria. Although not pathognomonic, but common radiological finding is a solid and cystic mass[3]. We report a case of MESTK mimicking hydatid cyst disease of the kidney which was treated by nephron sparing surgery.

Case History

A 37 year old female with no known comorbids presented to the uro-oncology clinic of Sindh Institute of Urology and Transplantation, Karachi in June 2021 with right lumbar pain since 3 months. There were no other associated symptoms like fever or haematuria. Her family history and physical examination was unremarkable. Routine laboratory investigations were performed. The complete haemogram revealed mild neutrophilia whereas the liver and renal function tests were normal. The immunological examination showed borderline positive Echinococcus antibody titres, with no evidence of hydatiduria. Ultrasound scan of the abdomen and pelvis revealed a complex cystic lesion arising from mid- pole of the right kidney with solid component and minimal internal vascularity. Rest of the viscera were unremarkable. (Figure1)

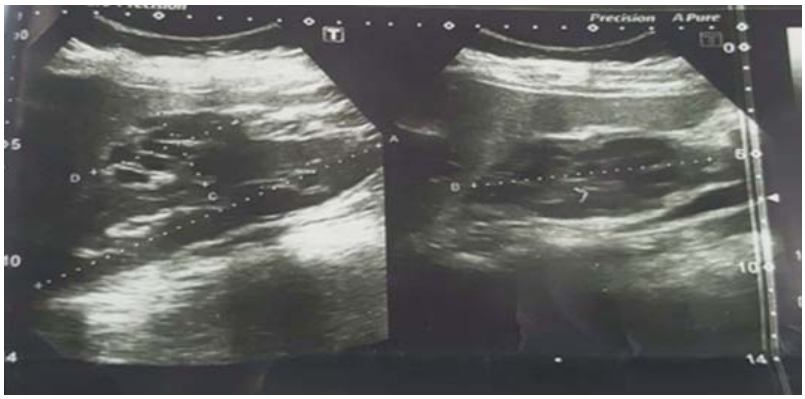

Figure-1: Greyscale Ultrasound showing cystic lesion in right kidney.

CT Scan of the abdomen with contrast revealed normal left kidney with the right kidney showing a mixed density cystic lesion involving the midpole with contrast enhancement.

A selected coronal section image of the right kidney showed a mixed density mass arising from the mid-pole extending into the upper pole with endophytic as well as

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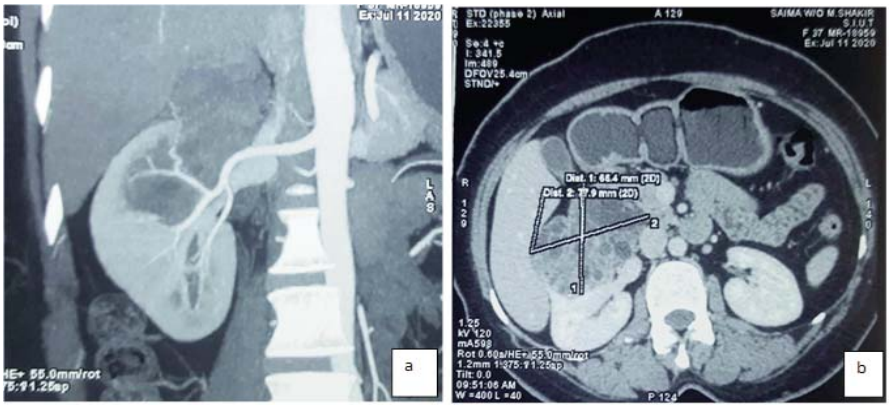

Figure-2: Contrast CT scan. (a): Coronal section of CT scan showing mixed density space occupying lesion with minimal enhancement at mid and lower pole of right kidney. (b): Axial section showing mixed density exophytic lesion with endophytic component in right kidney.

exophytic component (Figure-2 a). The selected axial section image demonstrated a contrast enhancing mixed density mass with multiple cysts (Figure-2 b).

Nephron sparing surgery was performed via the lumbar approach. The right kidney was mobilised to take arterial control (Figure-3 a). A large multilocular cystic lesion arising from the middle portion of the kidney in front of the hilum extending from upper pole to lower pole was identified (Figure-3 b and c). Right partial Nephrectomy including excision of the cystic mass was done after inserting a double J (DJ) stent (Figure -3 d). The specimen was sent for histopathology. The patient was kept on Albendazole to decrease the risk of recurrence. The post-operative recovery was unremarkable.

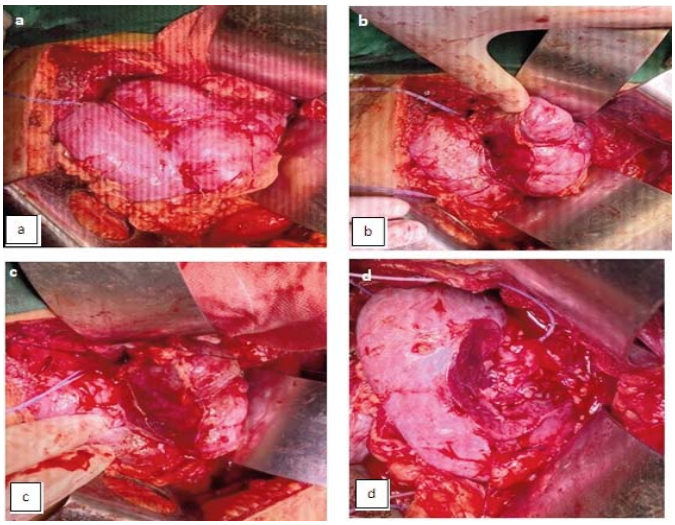

Figure-3: Intraoperative Pictures of Mass. (a): Right kidney with exophytic mass over anterior surface of mid and lower pole. (b): Plane created between normal renal parenchyma and mass. (c): Mass going into the renal hilum. (d): Mass excised with clear margins.

The gross histopathology findings were a single encapsulated, multicystic specimen with an attached kidney parenchyma measuring 13x10x4 cm. Multiple cysts were seen on the cut surface. No hard areas were identified and the surgical resection margins were unremarkable.

The microscopic examination in the low-power view showed normal kidney parenchyma on right side and a well-circumscribed biphasic lesion composed of cystic dilated

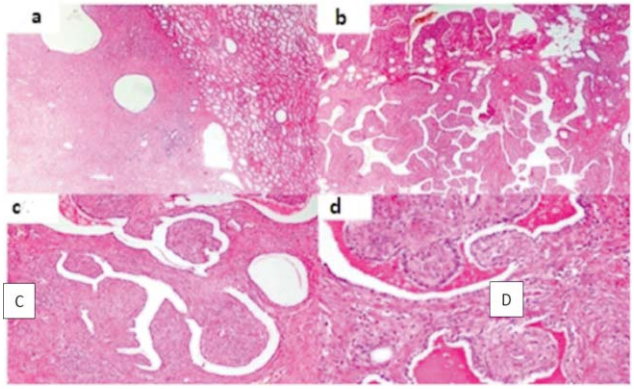

Figure-4: Histopathology. (A): Low-power view showing normal kidney parenchyma on right and a well-circumscribed biphasic lesion composed of cystically dilated tubules embedded in spindle cell stroma, on the left side of the field. (HE, ×50). (B): Low-power view showing areas of broad papillary fronds covering the stromal cores with foci of lipometaplasia in the stromal component. (HE, ×100). (C): Medium-power view showing papillary fronds lined by monolayered cuboidal epithelium and comprising of spindly stroma. (HE, ×200). (D): High-power view showing papillary fronds with focal nuclear hobnailing. No cytologic atypia or necrosis is seen. (HE, ×400).

tubules embedded in spindle cell stroma, on the left side of the field. (HE, ×50). (b): The low-power view projected areas of broad papillary fronds covering the stromal cores with foci of lipometaplasia in the stromal component. (HE, ×100). (c): Medium-power view showed papillary fronds lined by monolayered cuboidal epithelium and comprising of spindly stroma. (HE, ×200). (d): High-power view revealed papillary fronds with focal nuclear hobnailing. No cytologic atypia or necrosis was seen. (HE, ×400).Figure 4-a

At 6 months follow up the patient remained asymptomatic with no evidence of recurrence on imaging and laboratory parameters.

Discussion

Mixed epithelial and stromal tumor is a rare cystic come solid renal mass. Pawade et al[4] described it as a cystic hamartoma of the renal pelvis. Michal and Struck nominated it "mixed epithelial and stromal tumour" of the kidney[5], generally considered as a benign condition with strong predilection for perimenopausal women receiving estrogen replacement therapy. However some cases have been reported in, male and paediatric patients[6]. Microscopically, the tumour is biphasic, with both epithelial and stromal cell elements with expression of estrogen and progesterone receptors[7]. Turbiner et al. confirmed 62% and 85% of estrogen and progesterone receptor expression respectively[8]. Wang et al[9] denied association of hormone replacement therapy and MEST in a series of 8 cases. There are no specific diagnostic clinical or radiological features attributed to MEST. On a CT scan, it mimic multilocular cystic renal cell carcinoma due to manifestation of mixed density tumor with contrast enhancement[10]. Immunohistochemically, the epithelial components show positivity for epithelial membrane antigen and cytokeratin. Spindle cells are usually diffusely strong positive for desmin, actin and vimentin[10]. Our patient had no history of estrogen therapy. There is reported evidence of malignant transformation in few cases of MEST[3]. Malignant features can occur in either component of the tumor[10]. In our patient, we did not see features of malignant transformation. The tumour should be distinguished from other cystic neoplasms. Radical nephrectomy or nephron-sparing surgery should be performed where indicated. Routine surveillance and follow-up is advised owing to the risk of malignant transformation and recurrence. There are some limitations to accurate diagnosis as we lack single diagnostic investigation or imaging.

Conclusion

Mixed Epithelial and Stromal Tumour is a rare renal neoplasm that can mimic infective process, it warrants strong susceptibility index and vigilance for correct diagnosis and prompt adequate management.

Patients Consent: Patient's written consent was taken to publish this case report

Disclaimer: None.

Conflict of Interest: The person who signed ethical review statement is also a co-author of the same manuscript.

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