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The Mixed Epethelial Stromal Tumor in Medullary Sponge Kidney - A Rare Case Report

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Abstract

Mixed epithelial stromal tumor is rare distinctive kidney tumor which has been described over past few years. It is new entity included in WHO 2004 renal tumor classification. Mixed epithelial stromal tumor occurring in medullary sponge kidney is very rare and to best of our knowledge, it is first case in Indian literature. We report a case of 43 year old female who presented with left renal mass and loin pain.

Keywords -- *Mixed epithelial stromal tumor, medullary sponge kidney.*

INTRODUCTION

Mixed epithelial stromal tumor (MEST) is a distinctive benign tumor of kidney that should be distinguished from other renal tumors. It is predominantly seen in females mostly in perimenopausal period. Although these tumors are known to arise from renal pelvis, our case is distinct that it had intrapelvic component and growing in exophytic fashion in medullary sponge kidney. MEST of kidney is rarely mentioned in literature. Very few cases have been reported worldwide. Only recently it was included in renal tumor classification [1].

CASE REPORT

A 43 year old female presented to us with left abdominal pain on & off for last six months. She had no associated LUTS/ hematuria. Physical

examination was unremarkable. Her blood and urine examinations were Normal. Ultrasound (USG) showed left renal mass. Contrast CT showed exophytic solid cystic lesion involving upper pole of left kidney with solid component extending along upper pole of calyx into infundibulum and renal pelvis [7 x 6 cm size] along with medullary sponge kidney(Figure 1,2 – CT scan images). It was difficult to determine whether mass was arising from renal parenchyma or pelvis.

On abdominal exploration, there was well defined exophytic mass arising from parenchyma of upper pole of left kidney extending into renal pelvis. Hence upper pole partial nephrectomy was done with negative frozen section margins.

On Histopathological gross examination tumor was well-defined and capsulated. On cut section it showed white solid area of necrosis and hemorrhage. Microscopic examination showed tumor composed of large collagenised area containing bodies of spindle cells. They were interspersed with thick walled blood vessels, fatty tissue and several microcyst lined by cubical epithelium. Immunohistochemistry showed strongly positive for desmin in spindle cell area.

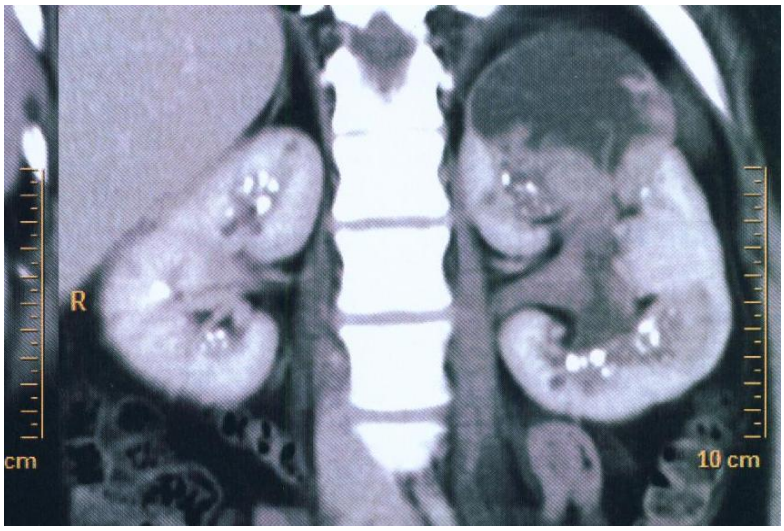

Figure 1 – Ct Scan Image

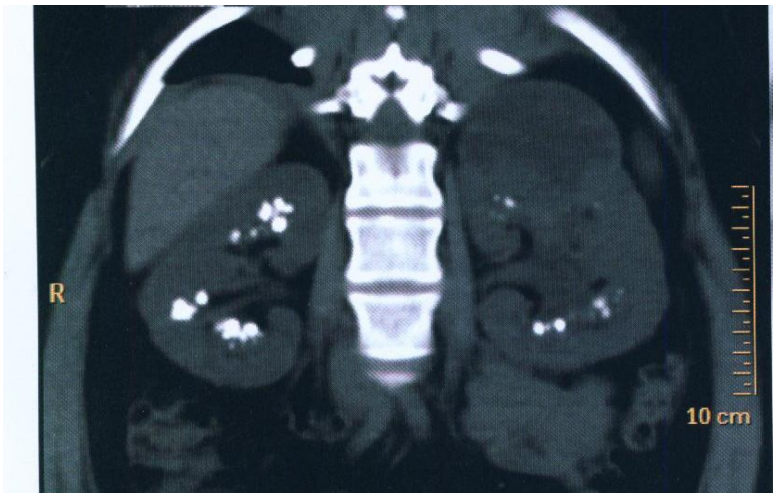

Figure 2 – Ct Scan Image

DISCUSSION

MEST is rare tumor involving kidney. The term MEST was first introduced by Michael and Syrucck in 1998. MEST is characterized by mixture of solid and cystic areas and is microscopically composed of proliferation of epithelial and stromal cells. [2,3] It is previously reported under different names like Adult Mesoblastic Nephroma, Cystic hamartoma of pelvis, Cystic Nephroma.[2,3]

Most patients presents with flank pain or hematuria or related urinary complaints. It predominantly occurs in women of perimenopausal age group and shows frequent estrogen and progesterone receptors. [2] Although benign, cases of malignant MEST have been reported.[4,5] Commonly it arises from renal parenchyma and pelvis and nephrectomy is advocated to manage this tumors.

However, in our case it was restricted to only upper pole of kidney and hence we could do partial nephrectomy preserving rest of the kidney.

CONCLUSION

In summary, mixed epithelial stromal tumor (MEST) is a rare kidney tumor. MEST arising from pelvis and growing exophytically in medullary sponge kidney is rarely reported. Intraoperative frozen section is necessary to exclude malignancy and attempt for conservative surgery is worthwhile. The overall prognosis is favorable. Such unusual case reports would greatly contribute towards understanding the disease as a whole.

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ABBREVIATIONS

Mixed epithelial stromal tumor - MEST, Lower urinary tract symptoms – LUTS

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