DOI: 10.7860/JCDR/2015/12277.5918

Case Report

Oncology Section

Giant Renal Mixed Epithelial and Stromal Tumour in A Young Female: A Rare Presentation

SIDHARTHA KALRA[1], RAMANITHARAN MANIKANDAN[2], LALGUDI NARAYANAN DORAIRAJAN[3]

ABSTRACT

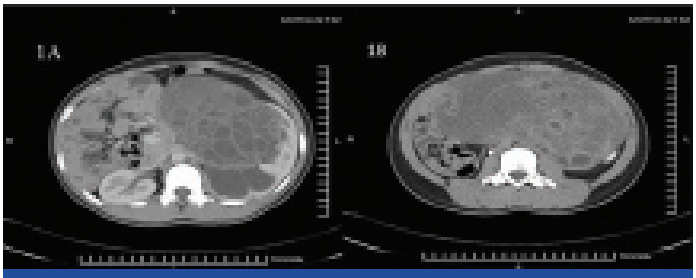
Mixed epithelial and stromal tumour of the kidney (MESTK), is a rare kidney tumour. About 100 cases so far have been reported in the literature. Although MESTK mostly presents in perimenopausal age group some rare cases in young females have been reported. We report a rare presentation of a 25 cm size giant MESTK arising from the left kidney in a young female in the absence of any clinically detectable hormonal abnormalities and successfully managed by nephron-sparing surgery.

Keywords: Cystic tumors, Kidney, Partial nephrectomy

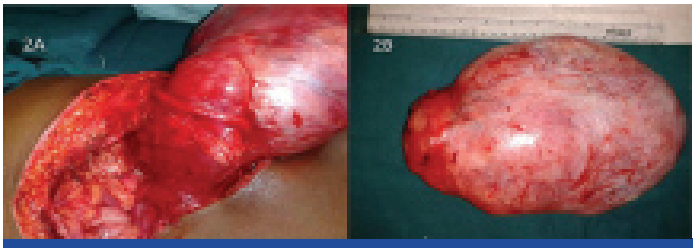
CASE REPORT

A 22-year-old woman who was being evaluated for infertility was referred to our clinic for an incidentally detected left renal mass. She did not report any history of flank pain,hematuria or systemic symptoms. Physical examination revealed a large mass of variable consistency occupying almost the entire left abdomen and crossing the midline. Her urine microscopic analysis,renal and liver function tests were normal. Her infertility evaluation did not reveal any hormonal disturbances or structural abnormality on ultrasonography.

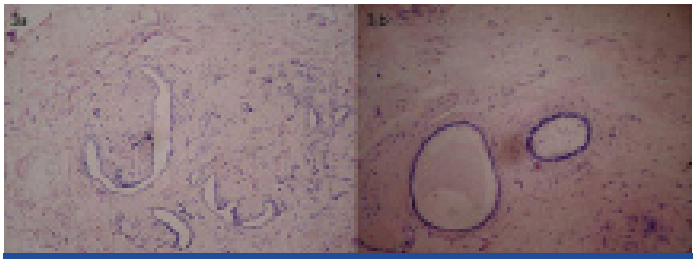
CECT abdomen showed a 24x25 cms heterogeneous, hypodense mass with cystic areas arising from the left kidney with minimal enhancement on contrast phase [Table/Fig-1a&b]. The clinic radiological diagnosis was that of a Bosniak III lesion with suspicion of cystic renal cancer. Intraoperative, the tumour was found to arise from the lower pole with well-demarcated plane with the normal kidney cortex [Table/Fig-2a].This facilitated in performing a left partial nephrectomy in spite of the huge mass. On gross examination, it was a 25 x 24 cm size well-encapsulated lesion [Table/Fig-2b],weighing about 2.7 kg with cut sections showing solid cystic components and thickened septa. Histologic sections showed numerous fatty, fibro vascular and smooth muscle stromal components along with epithelial components [Table/Fig-3a&b]. Stromal component was distinctly higher compared to epithelial component with presence of stromal bundles suggestive of MESTK. The immunohistochemical profile for stromal components was positive for estrogen and progesterone receptors, smooth muscle actin, and desmin while the epithelium showed CD 10 positivity [Table/Fig-4a&b]. Features of malignant transformation such as dysplasia, increased mitotic activity, or tumour necrosis were not found on histopathology. The patient recovered uneventfully and is on regular follow up.



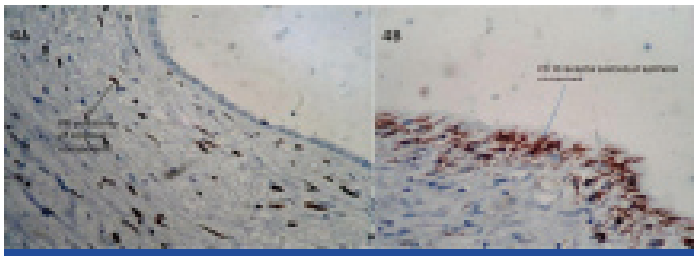
[Table/Fig-1a&b]: CECT abdomen showing a heterogenous mass with solid and cystic components arising from the lower pole of the left kidney and crossing the midline.



[Table/Fig-2a,b]: Intra-operative picture showing a clear demarcation between the tumour and normal renal parenchyma (b) tumour specimen measuring 25 cm



[Table/Fig-3a,b]: Histological features of the tumour. (a) Epithelial components comprising of tubules and cysts scattered among stromal fascicles with stromal bundles (b) Some tubules were lined by epithelial cells with a hobnail appearance, similar to renal collecting ducts. Mitotic figures and atypical cells are not observed in epithelial components and stroma



[Table/Fig-4a,b]: Immunohistochemical features of the tumour. (a) Spindle-Shaped stromal cells expressing progesterone receptors with a nuclear pattern(400x,PR+ve) (b) Epithelial cells in some neoplastic tubules were immunoreactive for CD10 (400x,CD 10 +ve)

DISCUSSION

Mixed epithelial-stromal tumour of the kidney (MESTK) was first described by Michal M and Syrucek M in 1998 [1]. Subsequently, Michal et al., described 22 cases of MESTK [2]. MESTK, previously described as congenital mesoblastic nephroma, leiomyomatosis renal hamartoma, solid and cystic biphasic tumour, cystic hamartoma, solitary multilocular cyst, and adult metanephric stromal tumour are now considered as a separate entity by the 2004 World Health Organization classification of renal neoplasms. About 100 cases so far have been reported in the literature [3]. Although MESTK mostly presents in perimenopausal age group some rare cases in young females have been reported [4]. The typical presentation is that

of perimenopausal women on hormonal therapy presenting with non-specific symptoms of flank pain, hematuria or genitourinary infection. The tumour size reported in literature ranges 2 to 24cm (mean 6cm) [3,5]. Considering the size, this is probably the largest MESTK reported so far.

Various theories have been postulated in its etiology but the one most favored is the altered hormonal milieu, typically observed in perimenopausal women or may be induced by therapeutic use of hormones. This hormonal imbalance might induce pluripotent differentiation of fetal mesenchyme into epithelial and mesenchymal components. This theory has been corroborated by the presence of estrogen and progesterone receptor expression in the spindle cell [3,6].

Grossly MESTK is mostly an encapsulated mixture of solid and cystic areas.Involvement of renal hilum and compression of the pelvicalyceal system is common, but gross infiltration of adjacent renal parenchyma is not seen. Microscopic features include epithelial and stromal cells with stromal bundles while the immunohistochemistry is positive for smooth muscle actin and desmin and negative for vimentin [3,5,6].Most of the MESTK are benign with very few malignant cases reported so far. It is the stromal component which has been found to give rise to the malignant expression and not the epithelial counterpart. Furthermore, malignant MESTK solely demonstrates progesterone receptors positivity while negative for estrogen receptors [7]. The differential diagnoses of MESTK include renal tumours with cystic components such cystic nephroma and cystic renal cell carcinoma which are radiologically classified as Bosniak class III to IV lesions [5].

Cystic nephroma has a bimodal age distribution with diagnostic peaks in the first 2 to 3 years of life, predominantly in males, and again in the fourth and fifth decades, especially in females. MESTK have a higher stromal-to-epithelial ratio, with smaller cysts as compared to cystic nephroma were there is lower stromal to epithelial ratio, and the cysts are larger with only thin septa. Some authors consider MESTK and cystic nephroma as different morphological variants of the same tumour entity due to similar gross and microscopic features [5-7].However, due to lack of specific molecular markers for differentiating these two tumour entities a definitive classification remains outstanding [6]. Although most of the MESTK are reported in perimenopausal age group, some rare cases in young females have been reported. Similar to our patient, these patients also had normal hormonal profile although one had a history of recurrent masturbation during childhood [4]. Probably the sex surge during puberty could have promoted differentiation of primitive periductal mesenchyme into neoplastic cells.

Nephron sparing surgery (NSS) is the current standard of care whenever feasible. Wang et al., [3], reported eight cases of MESTK two of which with a mean size of 3.5 cm were managed by NSS. A preoperative diagnosis in the form of percutaneous biopsy maybe considered whenever there is a clinic-radiological suspicion.

CONCLUSION

This case report demonstrates an atypical presentation of a giant MESTK in a young female with infertility. This probably is the largest MESTK reported so far.Considering the benign nature of the tumour, nephron sparing surgery must be considered as a therapeutic option even in large tumours.

REFERENCES

[1] Michal M, Syrucek M. Benign mixed epithelial and stromal tumour of the kidney. *Pathol Res Pract.* 1998;194:445–48.
[2] Michal M, Hes O, Bisceglia M, Simpson RH, Spagnolo DV, Parma A, et al. Mixed epithelial and stromal tumours of the kidney. A report of 22 cases. *Virchows Arch*. 2004;445:359–67.
[3] Wang C, Lin Y, Xiang H, Fang D, Jiang P, Shen B. Mixed epithelial and stromal tumour of the kidney: report of eight cases and literature review. *World J Surg Oncol*. 2013;11(1):207.
[4] Vergine G, Drudi F, Spreafico F, Barbisan F, Brachi S, Collini P, et al. Mixed epithelial and stromal tumour of kidney: an exceptional renal neoplasm in an 8-year-old prepubertal girl with isolated clitoral hypertrophy. *Pediatr Hematol Oncol*. 2012;29(1):89-91.
[5] Margulis V, Matin SF, Wood CG. Benign Renal tumours. In: Alan J. Wein Editor-in-chief. Campbell-Walsh Urology.Philadelphia: Saunders; 2010. pp. 1501-03.
[6] Turbiner J, Amin MB, Humphrey PA, Srigley JR, De Leval L, Radhakrishnan A, et al. Cystic nephroma and mixed epithelial and stromal tumour of kidney: a detailed clinicopathologic analysis of 34 cases and proposal for renal epithelial and stromal tumour (REST) as a unifying term. *Am J Surg Pathol*. 2007;31(4):489-500.
[7] Nakagawa T, Kanai Y, Fujimoto H, Kitamura H, Furukawa H, Maeda S, et al. Malignant mixed epithelial and stromal tumours of the kidney: a report of the first two cases with a fatal clinical outcome.*Histopathology*. 2004;44(3):302-04.

PARTICULARS OF CONTRIBUTORS:
1. Senior Resident, Department of Urology, JIPMER, Puducherry, India.
2. Associate Professor, Department of Urology, JIPMER, Puducherry, India.
3. Professor and Head of Department, Department of Urology, JIPMER, Puducherry, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Sidhartha Kalra,
Senior Resident, Department of Urology, JIPMER, Puducherry-605006, India.
Email: sid6121984@yahoo.co.in

FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Nov 30, 2014**
Date of Peer Review: **Mar 18, 2015**
Date of Acceptance: **Mar 27, 2015**
Date of Publishing: **May 01, 2015**