A PATHOLOGIST'S EXPERIENCE WITH THE HEAVIEST RENAL TUMOR EXCISED TILL DATE

Pathology

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ABSTRACT

Mixed epithelial and stromal tumor (MEST) was described in 1973 by Block et al. It was introduced in WHO in 2002 and was defined as a complex renal neoplasm composed of a mixture of stromal and epithelial elements. Microscopically it shows abundant stroma that is thrown into broad papillae with microcysts embedded within. This gives it a phyllodes-like appearance. The tumor is benign. There are no reported recurrences of MEST. We here report the histological finding of the heaviest renal mass excised to date and emphasize on the role of careful histological examination to not miss out rare benign conditions like MEST.

KEYWORDS

Kidney, Largest Kidney Mass, Mixed Epithelial Stromal Tumor, Benign Kidney Tumor

INTRODUCTION

Mixed epithelial and stromal tumor (MEST) was introduced in WHO in 2002 [1]. There are little above 100 cases of MEST reported in literature till now [2]. MEST shows a striking female predominance that correlates with the pathogenesis-related to extra ovarian rests of ovarian stroma [1]. Microcyst and tubules lined by Mullerian epithelium, exhibiting apocrine snouts and broad stromal papillae giving it a phyllode like an appearance, expressing ER and PR positivity are typical features of MEST. The tumor is benign with no evidence of recurrence and warrants a surgical excision. We report a case of the heaviest kidney tumor removed which was listed in the Guinness World Records in 2016.

Case Report

A 28-year-old female came with complaints of abdominal pain for 2 years. Computed tomography (CT scan) of the abdomen revealed a huge mass arising from the right kidney (fig 1a). It was encapsulated, multiloculated cystic. The clinical suspicion was multilocular cystic nephroma. The mass was excised by open abdominal surgery. The mass weighed 5500 grams and measured 31 x 19.6 x 19 cm (fig 1b). Externally it appeared solid. Cut surface was predominantly solid with multiple cysts ranging from 5 cm to 0.5 cm (fig 1c). The cysts contained serous fluid; the lining was smooth. Microscopic examination featured a biphasic benign tumor composed of cysts, branching tubules embedded in the stroma. The cysts were lined by cuboidal to columnar cells (fig 1d). The stroma was fibro-collagenous and thrown into broad papillae with microcysts embedded giving it a phyllodes-like appearance. Few cells demonstrated apical snouts, apocrine secretions, giving a hobnail appearance (fig 1e). No mitotic figures, areas of necrosis were noted. The spindle stromal tissue was positive for ER and PR (fig 1f and fig 1g), negative for other markers. This confirmed the diagnosis of MEST. The patient is being followed up. She is clinically asymptomatic to date.

DISCUSSION

MEST was coined by Michal and Syrucek in 1998 [1]. Calio et al studied 53 cases of MEST, out of which 46 were females [3]. Patients present with vague abdominal pain.].

Out of 14 MEST cases studied by Turbner et al, the largest tumor had a diameter of 21 cm [1]. Calio et al stated that 26 % of their cases were exclusively solid [3]. Our case was a solid-cystic renal mass occupying the whole abdominal cavity and was listed in the Guinness World Record as the heaviest kidney tumor removed to date.

The stromal component of the tumor is considered neoplastic while an

epithelial component is Mullerian epithelium. The stromal component resembles ovarian stroma and is theorized been derived from extragonadal rests of ovarian stroma and foetal primitive mesenchymal tissue in the kidney which proliferate during hormonal imbalance [3]. Thus, stromal cells exhibit a widespread expression of ER and PR, as did our case.

Michal and Syrucek in their original report described MEST exhibiting adenomatous and mesenchymal components [4]. The epithelial component is cysts and complex branching tubules. Turbner et al demonstrated hobnail cells in all their cases; seen in our case also [1]. The mesenchyme forms spatulate papillae giving MEST a phyllodes-like appearance.

Cystic Nephroma (CN) is a close differential of MEST. Turbner et al have proposed defining CN and MEST under the title of Renal epithelial and stromal tumor (REST) [1]. However, there are subtle differentiating features. MEST has septae more than 5 mm thick, also the IHC expression of ER, PR is stronger and widespread in MEST. Although benign tumours, malignant transformation in MEST has been recently reported [2]. However, no malignancy was noted in our case.

There are no reported recurrences of MEST. The treatment is surgical excision and closed follow up.

CONCLUSION

We thereby report the histological finding of the heaviest renal mass excised to date and emphasize on the role of careful histological examination to not miss out rare benign conditions like MEST.

Legends:

Fig 1a: CT Scan image showing a large encapsulated multiloculated solid-cystic mass arising from the right kidney and occupying the

entire right abdominal cavity.

Fig 1b: Heaviest renal tumor recorded weighing 5500 grams

Fig 1c: Cut section of tumor reveals predominantly solid areas with multiple cysts ranging from 5 cm to 0.5 cm in diameter. The surface appears pale grey and firm.

Fig 1d: Cysts and tubules are lined by benign epithelial cells. (H and E, 100x)

Fig 1e: Phyllode-like architecture noted. Composed of stromal proliferation with microcystic structures within. The epithelial lining showing apocrine snouts and secretions: hobnail appearance.

Fig 1f-1g: Spindle stromal cells showing immunohistochemistry, Oestrogen receptor (fig 1f) and Progesterone receptor (fig 1g) positivity.

REFERENCES

- 1) Turbinder J, Amin M, Humphrey PA, Srigley JR, Leval AD, Radhakrishnan A, Oliva E. Cystic nephroma and mixed epithelial and stromal tumor of the kidney: A detailed clinicopathological analysis of 34 cases and proposal for renal epithelial and stromal tumor as a unifying term. *Am J Surg Pathol*. 2007; 31: 489-500.
- 2) Jing Ye, Qing Xu, Jing Zheng, Shou-An Wang, Ya-Wei Wu, Jun-Hui Cai, Hu Yuan. Imaging of mixed epithelial and stromal tumor of kidney: a case report and review of the literature. *World J Clin Cases*. 2019; 7(17): 2580-2586.
- 3) Calio A, Eble J, Grignon DJ, Delahunt B. Mixed epithelial and stromal tumor of kidney. *Am J Surg Pathol* 2016; 40: 1538-49
- 4) Michal M, Syrucek M. Benign mixed epithelial and stromal tumor of the kidney. *Pathol Res Pract*. 1998;194:445-448.