Int J Clin Exp Med 2015;8(8):14180-14183
www.ijcem.com /ISSN:1940-5901/IJCEM0010585

Case Report

Mixed epithelial and stromal tumor of the kidney: a rare case report and review of the literatures

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Received May 23, 2014; Accepted July 11, 2015; Epub August 15, 2015; Published August 30, 2015

Abstract: Mixed epithelial and stromal tumor of the kidney (MESTK) is a rare complex renal neoplasm composed of a mixture of cystic and solid components. Until date only few cases of MESTK have been reported. We present here a rare case of MESTK that was diagnosed in a 56-year-old female. The patients were referred to our hospital due to a mass on the right kidney identified incidentally in a routine physical examination. A pre-operative diagnosis of cystic renal cell carcinoma was made and a right radical nephrectomy was carried out. Macroscopically, a cystic tumor was noticed in the upper portion of the right kidney. Various-sized cysts accompanied by multiple cysts and few solid areas were observed. Immunohistochemically, various epithelial markers as well as stromal markers were identified. Taken together with all the immunohistochemical results and morphological pattern of the tumor, a diagnosis of MESTK was made. MESTK is relatively rare and generally benign. However, it is difficult to distinguish between benign or malignant tumors according to the current radiological method. Therefore a complete resection of the tumor by partial or radical nephrectomy is suggested.

Keywords: Mixed epithelial and stromal tumor of the kidney, kidney neoplasm

Introduction

Mixed epithelial and stromal tumor of the kidney (MESTK) is a rare complex renal neoplasm composed of a mixture of cystic and solid components [1]. It was first reported by Block et al and originally as a congenital mesoblastic nephroma in 1973 [2]. In 1998 Michal M et al first described the term MESTK. Other terms such as cystic biphasic tumor, adult mesoblastic nephroma (MN), leiomyomatous renal hamartoma, multilocular renal cyst with müllerian-like stroma, adult mesoblastic nephroma, solid and cystic biphasic tumor and cystic hamartoma of the renal pelvis have also been described. During the past decades, emerging features of MESTK have been explored, leading to an increased awareness of this type of renal tumor. Here we present a rare case of MESTK that was diagnosed in a 56-year-old female and a review of the literature.

Case presentation

A 56-year-old female patient were referred to our hospital due to a mass on the right kidney identified incidentally in a routine physical examination. Apart from two thyroidectomies due to thyroid carcinoma that were performed 1 and 2 years ago respectively, no accompanying disease was present. She reached menopause at the age of 42 and didn't complained a history of irregular menses. A physical examination revealed pain on percussion on right lumbar region. All of her laboratory data were within normal limits except for a slightly increased percentage of reticulocyte (RET%, 1.07%). On ultrasonographic examination a large cystic mass in the right kidney area was noticed. A computed tomography (CT) scan revealed a 6.0 × 6.8 cm well-defined, unevenly mass lesion with cystic components and patchy high density shadows and the tumor was not enhanced uniformly

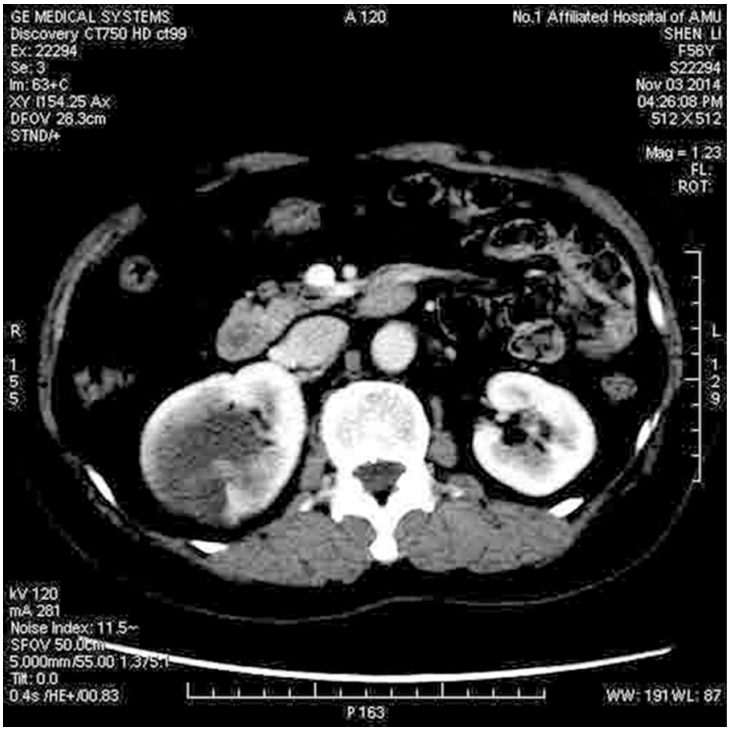

Figure 1. CT scan revealed a 6.0* 6.8 cm well-defined, unevenly mass lesion with cystic components.

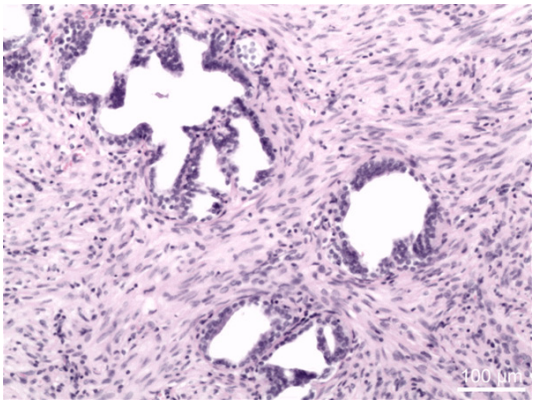

Figure 2. Few amount normal appearing tubules with round lumina scattered throughout the spindle cell component (H&E × 100).

(**Figure 1**). No positive findings were revealed from chest X-ray.

A pre-operative diagnosis of cystic renal cell carcinoma was made and a right radical nephrectomy was carried out. Macroscopically, a cystic tumor measuring 7 cm in maximum diameter was noticed in the upper portion of the right kidney. After sucking out the fluid, various-sized cysts accompanied by multiple cysts and few solid areas were observed. No obvious necrosis and hemorrhage was identified. No renal parenchymal invasion was observed.

Microscopically, the lining of the cystic wall revealed a spindle cell proliferation. The spindle cells were arranged in braided and bundles and no obvious cytologic atypia was noticed. A small amount of normal appearing tubules with round lumina that were distributed in the spindle cell component were also identified (**Figure 2**).

Immunohistochemically, the epithelial cells revealed positive expression of CK7 and PCK. The stromal components showed strong expression of SMA, Vimentin and Desmin. None expressed HMB-45, CD34 or PR. A minor proportion of the cells displayed expression of Ki-67. Taken together with all the results and morphological pattern of the tumor, a diagnosis of MESTK was made.

Discussion

MESTK is a rare genitourinary tract tumor composed of both epithelial and mesenchymal components. It was first defined by Michal and Syrucek in 1998 [3]. MESTK mostly occurs in perimenopausal women with average age of 46 year-old [4]. Until date, only two cases of MESTK were diagnosed in male [5].

The most typically presenting symptoms of MESTK include flank pain, hematuria and symptoms related to genitourinary infections. However, approximately 25% of MESTKs were incidentally diagnosed [6]. The mostly described imaging features of MESTK were circumscribed mass that located in cortex or protruding from cortex, with multiple septa with solid and cystic proportions of tumor. A delayed enhancement of the solid component during the nephrographic phase in the contrast-enhanced CT was also frequently observed [7]. Therefore MESTKs may be diagnosed as renal angiomyolipomas, adult cystic nephroma, cystic renal cell carcinoma and complex cyst preoperatively [8].

Microscopically, MESTK is composed of both mesenchymal components that are characterized by fascicles of spindle cells and epithelial components that generally consist of glands scattered throughout the stroma [9]. Mesenchymal components frequently express ER, PR, SMA, Vimentin, Desmin, and inhibin, while usually negative for HMB-45, S-100, CD34 and CD99. Epithelial components were shown to express cytokeratins, especially CK7, Ulex europaeus and caretinin [1, 4, 10].

The pathogenesis of MESTK is largely unexplored. One hypothesis revealed that the presence of fetal primitive mesenchyme may proliferate under the conditions of imbalanced hormonal level. Another focused on the abnormal migrated ovarian stromal cells that incorporated in the ureteric bud and metanephric mesoderm, which may become activated, secrete paracrine factors and induce proliferation of adjacent epithelium [11]. Regarding to the medical history, a great number of the female patients revealed a history of prolonged estrogen exposure and the male patients reported a sex-steroid administration. In combination with the frequent expression of ER and PR, hormonal factors may play a vital role in tumorigensis of MESTK [12, 13].

Due to the difficulty in preoperative diagnosis of MESTK, preoperative biopsy and intraoperative frozen sample diagnosis may be an optional method. Complete resection of the tumor by partial or complete nephrectomy according to the patient's condition is the suggested treatment of MESTK. In general, the great majority of MESTKs behaved in a benign manner, although few malignant cases and malignant transformation were reported [1, 14-16].

Conclusion

MESTK is relatively rare and generally benign. However, it is difficult to distinguish between benign or malignant tumors according to the current radiological method. Therefore a complete resection of the tumor by partial or complete nephrectomy is suggested. Comprehensive studies aiming at the pathogenesis and pre-operative diagnosis will improve the current understanding of the disease.

Disclosure of conflict of interest

None.

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