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Case Report

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A Case Report of Partial Nephrectomy of Mucinous Cystadenocarcinoma in Kidney and Its Literature Review

Sung Han Kim, MD, PhD[1]
Heong Dong Yuk, MD[2]
Weon Seo Park, MD, PhD[3]
Sun Ho Kim, MD, PhD[4]
Jae Young Joung, MD, PhD[1]
Ho Kyung Seo, MD, PhD[1]
Kang Hyun Lee, MD, PhD[1]
Jinsoo Chung, MD, PhD[1]

[1]*Department of Urology, Center for Prostate Cancer, National Cancer Center, Goyang,*
[2]*Department of Urology, Seoul National University Hospital, Seoul, Departments of*
[3]*Pathology and* [4]*Radiology, Center for Prostate Cancer, National Cancer Center, Goyang, Korea*

Correspondence: Jinsoo Chung, MD, PhD
Department of Urology, Center for Prostate Cancer, National Cancer Center, 323 Ilsan-ro, Ilsandong-gu, Goyang 10408, Korea
Tel: 82-31-920-1676
Fax: 82-31-920-1790
E-mail: cjs5225@ncc.re.kr

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Mucinous cystadenocarcinoma (MC) of the kidney is a rare epithelial tumor originating from the renal pelvic urothelium and few study cases have been reported. Because of the rarity of these tumors and their unknown histogenesis, its diagnosis is difficult until surgical exploration. We report here on a 55-year-old man referred to the urology department from the hepatology department because of a cystic renal mass measuring approximately 5 cm in size, which was detected incidentally under ultrasonography during the routine examination of liver. The renal mass was finally diagnosed as MC originating from kidney after partial nephrectomy and the patient still showed no evidence of recurrence until 12 months postoperatively. This is the first report on a case of renal MC in a patient who underwent partial nephrectomy. The aim of this report is to present our unusual case of MC and also review the previous literature on the pathological and radiological aspects of MC of kidney.

Key words
Mucinous cystadenocarcinoma, Kidney, Nephrectomy

Introduction

Mucinous cystadenocarcinoma (MC) of kidney is a rare tumor comprising less than 1% of kidney malignancies of the renal pelvis [1,2]. Because of the rarity of these tumors and their unknown histogenesis, it is difficult to determine their exact origin and proper treatment regimen. Diagnosis can often be confusing preoperatively because they can be mistaken for renal cystic disease. Surgical exploration is needed for the diagnosis and treatment, and many authors recommend complete excision of retroperitoneal tumor [3,4]. We report here on a 55-year-old male patient with MC of the kidney who underwent partial nephrectomy (PN) of tumor excision without evidence of recurrence 12 months after surgery. To the best of our knowledge, this is the first PN case ever reported of this type of tumor from kidney origin. We also review the previous literature on MC and share our experience of renal MC with other clinicians in order to be aware of the characteristics of the tumor in their clinical settings.

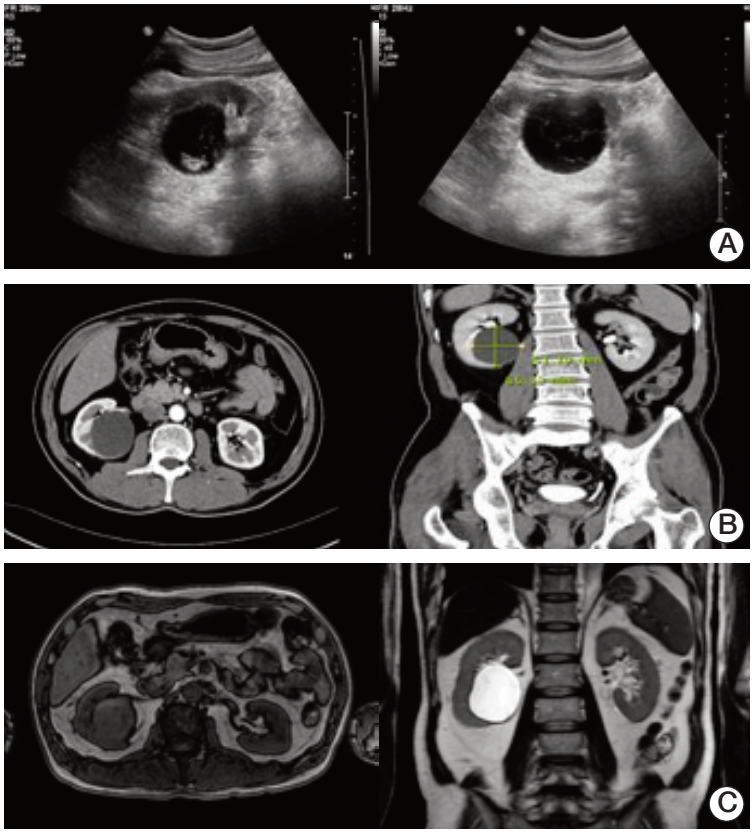

Fig. 1. Computed tomography (A), magnetic resonance (B), and ultrasonographic (C) images showing a large unilocular renal cyst of the right kidney with multiple septation, a single mural nodule, and fluid materials filled in the cysts, but separate from other adjacent organs.

Case Report

A 55-year-old man was referred to the urology department from the hepatology department because of a cystic renal mass measuring approximately 5 cm in size, which was detected incidentally under ultrasonography during the routine examination of liver. Ultrasonography showed a 5.5-cm-sized complex cystic mass with irregular septa and an echogenic portion in the right kidney classified as Bosniak III or higher cyst (Fig. 1A). According to records from an outside facility, the patient had a history of liver cirrhosis and hypertension, and a benign renal cyst diagnosed 10 years ago without further follow-up evaluation. He was originally placed on observation because he was asymptomatic. He had no clinical symptoms including palpable mass, weight reduction, anorexia, hematuria, and flank pain. Physical examination, including palpation of the renal mass over the abdomen, did not show any abnormal findings. Laboratory tests, including complete blood count, chemistry profile, urinalysis, and chest X-ray, were all within normal limits, except mildly elevated hepatic enzyme (glutamate oxaloacetate transaminase/glutamate pyruvate transaminase 101/35 IU/L), total bilirubin 1.3 mg/dL, alkaline phosphatase 128 IU/L, and γ glutathione transaminase 133 IU/L. Abdominopelvic computed tomography (CT) and magnetic resonance imaging scanning showed a 5.5-cm-sized Bosniak III hemorrhagic cyst with a small amount of irregular enhancing septa in the posteroinferior part in the right kidney (Fig. 1B and C). There was no evidence of extracystic extension or distant metastasis in the abdominopelvic cavity.

Open retroperitoneal PN was performed successfully without complication such as ruptured cyst. On surgical exploration, an endophytic 5.8×3.5×2.5-cm-sized cystic tumor revealed originating from the middle area of the right kidney with a free resection margin of 3 mm from the normal renal parenchyme resulting in a total resected size of 6.9×5.0×1.4 cm including cystic tumor and renal tissue weighed approximately 27 g (Fig. 2). The cystic tumor was constituted with unilocular cystic and solid structure filled with dark brown gelatinous mucus contained materials adjacent to the renal capsule. The inner surface of the cyst

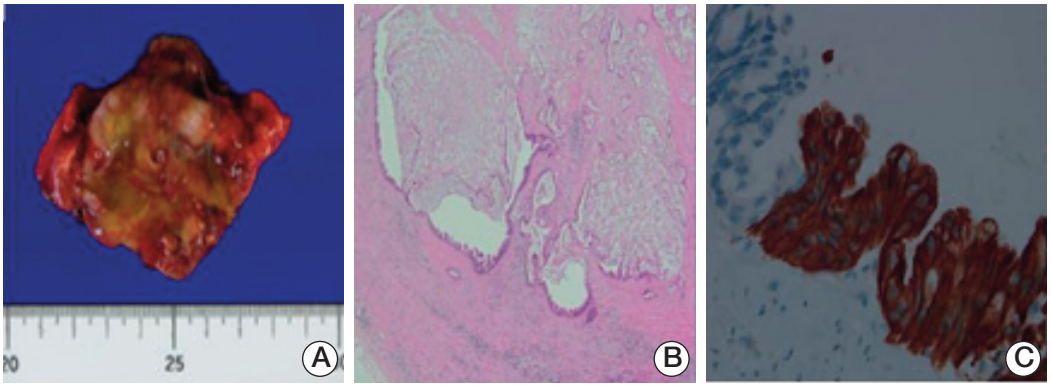

Fig. 2. (A) Fresh and formalin fixed specimen of renal cystic mass after partial nephrectomy. (B) Areas of the cyst wall lined by atypical mucinous epithelium showing stratification and stroma-free papillae (H&E staining, ×40). The tumor cells invade into the capsular stroma. There is no endometrial stroma-like structure under the tumor epithelial lining. (C) Atypical mucinous epithelium with patchy cytokeratin 7 staining (×400). Atypical mucinous epithelium with diffuse strong staining for the *MUC2* and *MUC5AC* gene products (×400).

was smooth and the cystic wall measured 0.2 cm at the maximum thickness. There was no evidence of capsular rupture. Microscopically, there was no necrosis, no vascular invasion, and no capsular, perirenal, or peritumoral fat invasion was found, and there were no tumor cells in the mucus component (Fig. 2). However, the tumor extended to the tumor stroma. When viewed microscopically, the cyst wall was mostly lined by cuboidal to columnar mucinous cells of the intestinal type. In most areas, the cells were single-layered, with mild atypia. Broad projections with fibrous cores covered by similar mucinous epithelium were occasionally seen. In foci, the cells were stratified, with mild to moderate atypia and mitotic activity. The fibrous cyst wall showed the presence of spaces containing mucin and was lined by atypical cells. The tumor cells invaded into the capsule stroma (Fig. 2). Immunochemical staining showed that pan-cytokeratin, cytokeratin-7, and cytokeratin-20, epithelial membrane antigen, and MUC-2, MUC-5AC were positive, and α-methylacyl-CoA racemase and vimentin negative in carcinoma cells. Focal positive membranous staining for carcinoembryonic antigen (CEA) was found in the epithelial cells of the benign and atypical components. Thus final diagnosis with pathologic staging was T1bNxM0 with retroperitoneal MC of the kidney.

On postoperative seventh day, the patient was discharged without complication and on the 10th day, the serum concentration of CEA was decreased from preoperative 7.3 to within normal limits postoperatively (3.2 ng/mL). The patient was followed up until 12-months without any signs of recurrence on the follow-up abdominopelvic CT after the operation.

Discussion

Since the first report of MC of retroperitoneum (RMC) in 1977 by Roth and Ehrlich [5], approximately 50 cases of RMC have been reported in the English medical literature [3,4-7]. MC of kidney is a much rarer epithelial tumor, comprising less than 1% of malignancies, from epithelium of the renal pelvis [1,2]. In RMC there is a distinctive gender difference in mean presenting age, with 55 years old in men and 42.2 years old in women, and the size of MC in kidney ranged from 5 to 24 cm in diameter. Several theories of histogenesis of renal MC have been suggested to explain the metaplastic formation of glands in the pleuripotent uroepithelium of the collecting system. Three histogenetic suggestions are theories of chronic irritation, differentiation of coelomic epithelium, and maldevelopment of kidney [8]. To date, the increasing supported hypothesis is the theory of chronic irritation which causes high pressure resulting in metaplastic formation of transitional uroepithelium on the renal pelvis. The next supported theory was that of the differentiated coelomic epithelium [1,9,10].

The clinical course appeared to be to be mostly indolent, but asymptomatic hematuria, mucusuria, flank pain, and palpable mass might also be presented. The overall prognosis and tumor biology of these tumors were uncertain because of their rarity and the short-term follow-up reports; however, renal MC was presumed to become aggressive so that radical or simple nephrectomy has been suggested as a definitive treatment in previous studies [1]. Some suggested an additional ureterectomy in case of presenting mucusuria to prevent implantation of tumor cells by mucusuria, because

renal MCs were able to develop in any location, such as bladder, ureter, and renal calyces where urothelium was present. Reported prognosis appeared to be slightly better for male gender than female gender and the longest reported follow-up period with good prognosis was 79 months [6]. Our case was a 55-year-old male with a localized renal MC without evidence of metastasis or invasion of adjacent organ or any evidence of mucusuria. After careful history taking and review of the radiologic evaluation, the slowly growing characteristics of the cystic tumor for more than 5 years and early detection of a renal mass found localized enough for PN enabled surgeons to perform PN successfully for preservation of renal function without tumor violation including cystic rupture. Despite a postoperative short follow-up period of 12 months, the patient was still in a healthy state without evidence of recurrence or elevation of tumor markers. However, further follow-up would be required due to its unknown clinicopathological property.

Radiologic tests including ultrasonography, CT, or magnetic resonance imaging are used to localize the tumor and to evaluate its nature. A mural nodule demonstrated within a septated cyst by ultrasonography or CT has been reported to suggest Bosniak classification III as a malignancy and the presence of a mural nodule within the cyst wall portends worse prognosis, given that 75% of patients who died had a mural nodule [4]. However, it is often difficult to differentiate a benign from a malignant neoplasm, or even to determine the origin site of the tumor using preoperative radiologic images. Those previous reports did not provide information regarding correlation of the mural nodules with a frankly malignant histology. Our patient also had a mural like nodule composed of pleomorphic sarcomatoid cells with mitotic figures in both the imaging studies and in the final pathology. Cases with a mural nodule may have an aggressive prognosis like ovarian cancers; therefore, careful histological assessment and careful postoperative follow-up were recommended for all mural nodules [4]. Other reports suggested that serum tumor markers rarely assist in the diagnosis or follow-up and the presence of glandular epithelial cells and high levels of CEA in the cystic fluid was very useful for making the diagnosis [11]. However, despite the presence of a mural nodule and a preoperative high CEA level of 7.3 ng/dL in this case, the tumor was successfully treated surgically and no tumor recurrence on CT follow-ups was detected until postoperative 1 year.

Pathologically, the benign and borderline mucinous epithelium in their cases showed apical membranous staining for CEA, whereas the areas of MC showed more extensive, cytoplasmic staining with this marker [11]. The staining pattern for both cytokeratin 7 and cytokeratin 20 in the current case is also similar to the pattern seen in ovarian mucinous tumors of the intestinal type [12]. The pattern of expression of the mucin peptide core antigens in this case is interesting, with strong diffuse expression of both gastric-type (MUC5AC) and intestinal type (MUC2) antigens [12,13]. *MUC5AC* gene expression has been found in 98%-100% of MC by immunohistoc-hemistry [13], whereas *MUC2* gene expression has been noted in 70%-100% of ovarian mucinous carcinomas [13,14]. Lower levels of MUC2 expression were observed in borderline and benign ovarian mucinous tumors [13]. These immunostaining results, similar to those of our current case, might support another differentiation of coelomic epithelium theory of MC histogenesis originating from the peritoneum (mesothelium) undergoing mucinous metaplasia and mucinous cystadenoma [15]. These may then progress on to borderline and malignant mucinous tumors.

Conflicts of Interest

Conflict of interest relevant to this article was not reported.

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