

Original Article

Renal mucinous tubular and spindle cell carcinoma: report of four cases and literature review

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Abstract: Mucinous tubular and spindle cell carcinoma of the kidney (MTSCC-K) is an unusual renal tumor. It is important to increase the recognition of the clinicopathological features of MTSCC-K and improve its clinical and differential diagnosis. This report described four cases of MTSCC-K with clinical, imaging, and pathological examination and showed that the tumor boundaries of MTSCC-K were clear, and tumor cells arranged into tubules and cord-like beams, between which was lightly stained myxoid stroma. The tumor cells were smaller and cube- or oval-shaped, with single small eosinophilic nucleoli, low-grade nuclei, and little nuclear fission. The myxoid stroma was scattered around lymphocytes and plasma cells. Immunohistochemical markers including CK7, CD117, EMA (epithelial membrane antigen), vimentin, and CK8/18, showed positive expression in tumor cells, but the tumor cells were negative for CD10 and villin. The proliferation index of Ki-67 was 5-10%. Since MTSCC-K is a rare low-grade malignancy, with unique histological and immunohistochemical characteristics, it is important for clinicians and pathologists to have a defined awareness of this tumor type in order to decrease the rate of misdiagnosis.

Keywords: Mucinous tubular and spindle cell carcinoma, immunohistochemistry, prognosis, differential diagnosis

Introduction

Mucinous tubular and spindle cell carcinoma of the kidney (MTSCC-K), recognized in 2004 by WHO [1], is a rare renal epithelial tumor implicating the urinary and male reproductive systems, which is believed to be a low-grade malignant tumor type [2, 3]. This tumor is often diagnosed as undifferentiated carcinoma or sarcomatoid carcinoma previously. The present report analyzed the clinical results of 4 patients who presented to The Third Affiliated Hospital of Soochow University (Changzhou, China) suffering from MTSCC-K, and performed a review of the relevant literature. Additionally, this study also aimed to raise awareness of this tumor type among clinicians and pathologists in order to decrease the rate of misdiagnosis.

Case report

Case 1

On November 7, 2011, a 77-years old male was admitted in the Third Affiliated Hospital of

Soochow University suffering from lumbodorsalgia on the left side for approximately one month. Imaging examination by abdominal computed tomography (CT) scan revealed a 7 cm solid mass in the inferior pole of the left kidney. A left radical nephrectomy was carried out by laparoscopy. Pathological examination showed that the kidney was 12 × 7 × 5 cm; there was a tumor in the pole of the left kidney measuring 7 × 5.5 × 5 cm. Dissection of the specimen revealed a well-circumscribed, solid, and off-white tumor.

Case 2

On March 31, 2013, a 61-years old male was admitted to our hospital suffering from lumbodorsalgia on the left side for approximately one month. Imaging examination by abdominal CT scan revealed a 10 cm solid mass in the inferior pole of the left kidney. A laparoscopic left radical nephrectomy was carried out. Pathological examination indicated that the kidney was 16 × 13 × 8 cm; the ureter was 12 cm; there was luminal dilatation with a diameter of

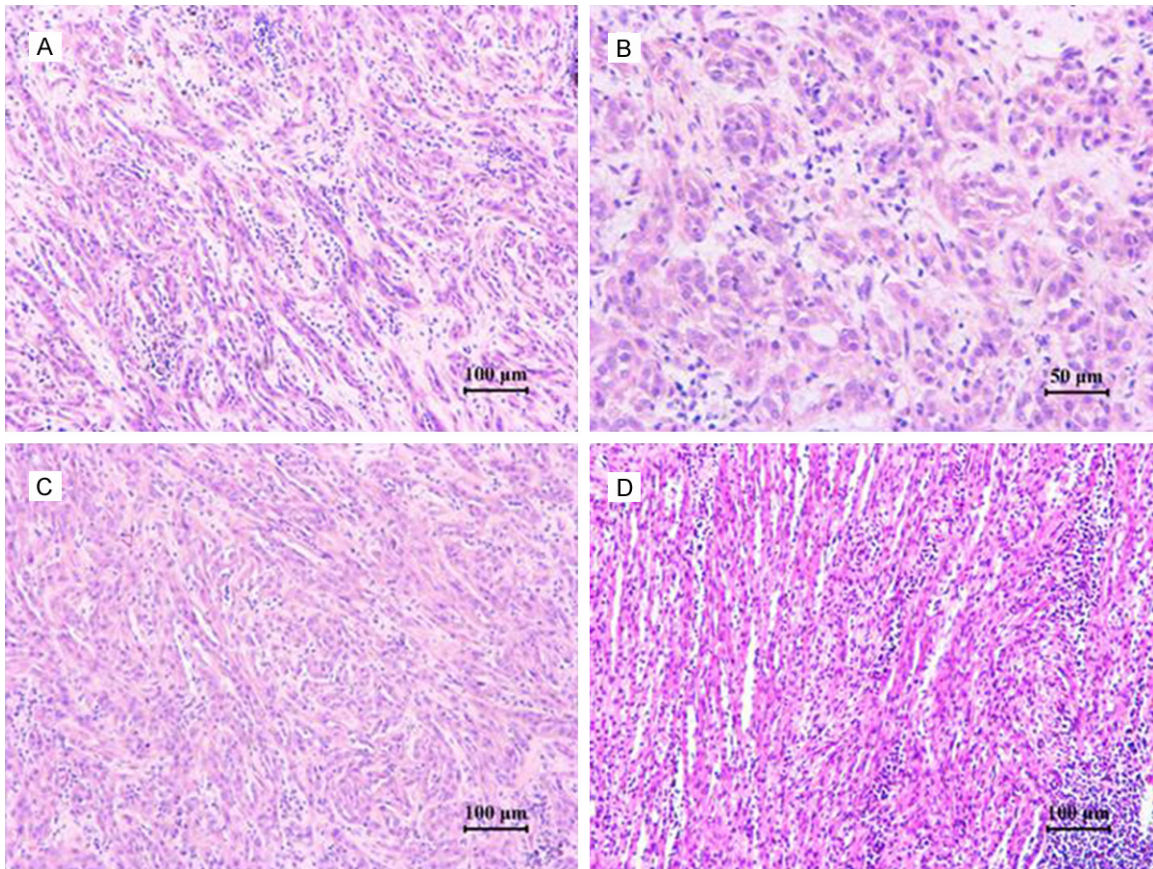


Figure 1. A: Myxoid stroma was interspersed among the tubular cells (magnification, $\times 100$); B: Tumor cells were cube- or oval-shaped, with single small eosinophilic nucleoli, low-grade nuclei, and little nuclear fission (magnification, $\times 200$); C: Tumor cells were relatively smaller (magnification, $\times 100$); D: Myxoid stroma was scattered with lymphocytes and plasma cells (magnification, $\times 100$).

about 1 cm. There was a tumor in the pole of the left kidney measuring $10 \times 8 \times 8$ cm. Dissection of the specimen revealed a well-circumscribed, solid, and off-yellow tumor.

Case 3

On June 7, 2013, a 49-years old female was admitted to our hospital suffering from lumbodorsalgia on the right side for approximately 2 months. Imaging examination by abdominal CT scan revealed a 4 cm solid mass in the inferior pole of the right kidney. A laparoscopic left radical nephrectomy was conducted. Pathological examination showed that the kidney was about $14 \times 9 \times 5$ cm; the ureter was 3 cm. There was luminal dilatation with a diameter of about 0.5 cm. A nodular tumor was observed in the pole of the right kidney near pelvis, measuring $4 \times 3.5 \times 3$ cm. Dissection of the specimen revealed a well-circumscribed, solid, and off-yellow tumor.

Case 4

On August 20, 2013, a 57-years old male presented to our hospital suffering from lumbodorsalgia on the left side for approximately 2 months. Imaging examination by abdominal CT scan revealed an about 4 cm solid mass in the inferior pole of the left kidney. A left radical nephrectomy was carried out by laparoscopy. Pathological examination indicated that the kidney with fat sac was about $12 \times 7 \times 3$ cm. There was a nodular tumor in the pole of the left kidney, measuring $3.7 \times 3.2 \times 3$ cm. Dissection of the specimen revealed a well-circumscribed, off-yellow, and solid tumor.

Microscopy

The tumor was invasive with an expansive growth. The tumor cells arranged in shapes of tubules and cord-like beams, between which was lightly-stained myxoid stroma. The tubular

Renal mucinous tubular and spindle cell carcinoma

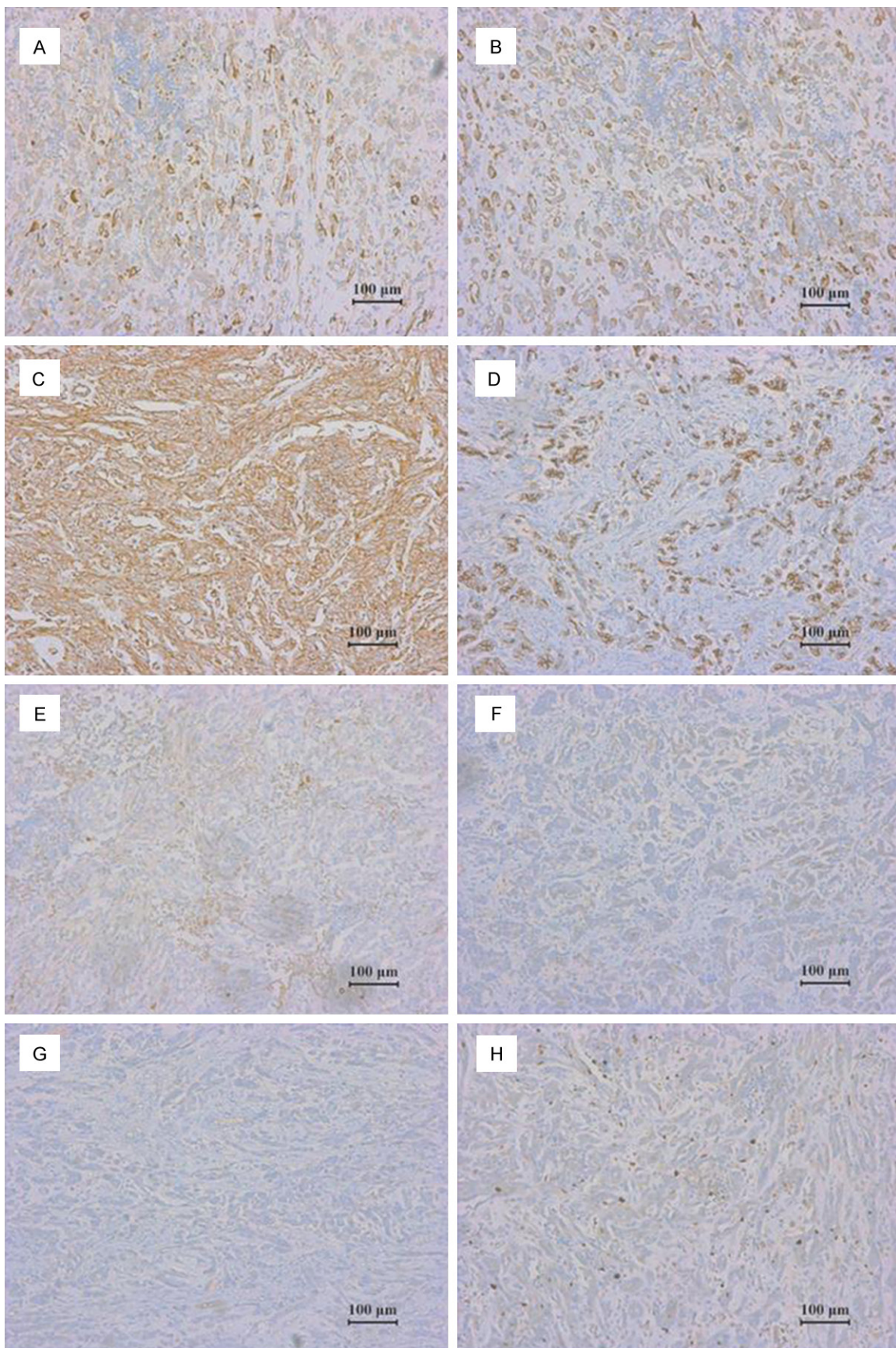


Figure 2. A-E: Tumor cells showed positive staining for CK7, CK8/18, vimentin, EMA, and CD117, respectively; F and G: Tumor cells displayed negative staining for CD10 and villin respectively; H: The proliferation index of Ki-67 was 5-10% (magnification, $\times 100$).

cells arranged in parallel and gradually appeared to exhibit slender tubular spindle cell-like structures. The tumor cells were smaller and cubic- or oval-shaped, with single small eosinophilic nucleoli and low-grade nuclei. Occasionally, necrosis and foam cell infiltration were identified. The myxoid stroma was stained by acidic mucus.

Diagnosis

All the 4 cases were diagnosed with MTSCC-K (Figure 1).

Immunohistochemistry

The tumor cells were stained positive for CK7 (Figure 2A), CK8/18 (Figure 2B), vimentin (Figure 2C), EMA (Figure 2D), and CD117 (Figure 2E), but were negative for CD10 (Figure 2F) and villin (Figure 2G). The proliferation index of Ki-67 was 5-10% (Figure 2H).

Follow-up

Although the 4 cases had no recurrence and metastasis so far, a long-term observation and follow-up are still needed for further evaluation.

Discussion

MTSCC-K is a newly identified rare low-grade malignant renal epithelial tumor in the past 20 years. To date, about 100 cases have been reported world-wide [1]. MTSCC-K is more common in adults, and female patients are approximately 2- or 3-fold more than male patients [4]. The disease has a wide age range (20-81 years; mean, 53 years) [5]. Most patients have no obvious symptoms; while some patients may have hematuria, back pain, repeated urinary tract infections, and kidney stones by physical examination [6].

MTSCC-K usually locates in the renal parenchyma with clear boundary and no membrane [2]. Microscopic examination showed some invasive region with relatively clear boundaries. The tumors are ranked with 3-phase morphology, tubular, myxoid, and cord-like. The tubular area

shows lumen in various sizes, and in some areas the lumen is not obvious [7]. The tumor cells in myxoid areas arrange into small tube, chordoid and/or beam cords, floating in the myxoid region. Most of the MTSCC-K is smaller and cube- or oval-shaped, with single small eosinophilic nucleoli, low-grade nuclei, and little nuclear fission [8]. The myxoid stroma is often scattered with lymphocytes and plasma cells. It has been reported that MTSCC-K is positive for CD10 (10/74), CD15 (12/40), P504S (48/51), CK7 (57/66), vimentin (16/34), EMA (46/54), AE1/AE3 (23/28), and CK19 (21/23) [9].

Differential diagnosis of this tumor included: 1. Angiomyolipoma. MTSCC-K is lack of adipose tissue and characteristic thick wall vessel, and the tumor cells arrange into differential tubular and mucinous structures. In addition, angiomyolipoma is HMB45⁺. 2. Oncocytic adenoma. Macro examination indicates that oncocytic adenoma of the cross section is red-brown, with the radial scar in the center of tumor; the tumor cells are round-shaped or polygonal, and the oncocytic granular cytoplasm is with arranged nested acinar; MTSCC-K has no central radial scar, and the tumor cells are smaller and cube- or oval-shaped, with spindle cell areas. 3. Metanephric adenoma. Metanephric adenoma is composed of uniform small cells, arranged with closed and circular, tubular, and round cell nests of solid areas. It usually has no spindle cells and myxoid areas. 4. Renal juxtaglomerular cell tumor. The boundary of tumor cells is clear and composed of sheet-arranged spindle and polygonal cells. The tumor diameter is usually less than 2 cm. Renal juxtaglomerular cell tumor usually expresses renin, actin, CD34, etc. 5. Sarcomatoid carcinoma. It cannot be differentiated from MTSCC-K in gross examination. Sarcomatoid carcinoma is highly malignant under microscope, with cell atypia, caryocinesia, and visible coagulation necrosis. Compared with MTSCC-K, the prognosis of sarcomatoid carcinoma was poor. 6. Papillary renal cell carcinoma. The tumor cells arrange into closely papillary, entity shape, and it is difficult to identify the papillary axis without foam cell-like macrophages. However, MTSCC-K usually appears as areas with spindle cells.

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Disclosure of conflict of interest

None.

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