OPEN ACCESS – CASE REPORT

Mucinous Tubular and Spindle Cell Carcinoma of the Kidney: A Case Report and Concise Review of Literature

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ABSTRACT

Mucinous tubular and spindle cell carcinoma (MTSCC) is a rare neoplasm of the kidney. Recognition of this rare entity is important with regards to a patient's prognosis and therapeutic management.

Key words: mucinous tubular and spindle cell carcinoma, kidney neoplasms, immunohistochemistry, surgical pathology

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INTRODUCTION

Mucinous tubular and spindle cell carcinoma (MTSCC) is a rare neoplasm of the kidney, accounting for less than 1% of renal cell carcinomas.[1] Females are more frequently affected than males, with a majority of cases diagnosed in adults. Most tumors are indolent and discovered incidentally.[1,2]

First described in 1996, MTSCCs were previously described as "low-grade collecting duct carcinoma," and formally introduced in the 2004 edition of the WHO Classification of Tumors.[1] Tumors are predominantly located in the renal cortex. Histologic features of MTSCCs include elongated tubules and spindle cells within a mucinous to myxoid stroma, with components occurring in varying proportions.[1,2] Tumors are generally low-grade, but high-grade features such as pleomorphism, nuclear atypia and extensive necrosis have been recognized.[3] Metastases are remarkably rare, especially in tumors with low-grade morphology. Despite its nonspecific clinical features, recognizing MTSCC's distinct histomorphology is important due to different therapeutic and prognostic implications.[1]

We present a case of a 50s female with an incidental finding of a renal mass on imaging, and describe its gross pathology, microscopic features and a concise review of literature.

CASE REPORT

A female in her 50s with chronic kidney disease was found to have an incidental ultrasound finding of a left kidney mass. Further investigation with triple-phase computed tomography of the abdomen showed a predominantly endophytic mass with high complexity in the superior to middle pole of the left kidney (2+3+3+x+3 = 11+x by R.E.N.A.L. Nephrometry scoring). The patient was then admitted for surgery. A radical nephrectomy was performed, and the specimen received consisted of a left kidney with no ipsilateral adrenal gland attached. Serial sections of the kidney showed a well-circumscribed, cream-tan mass with a central hemorrhagic area, located in the superior to middle pole of the kidney and measuring 6.8

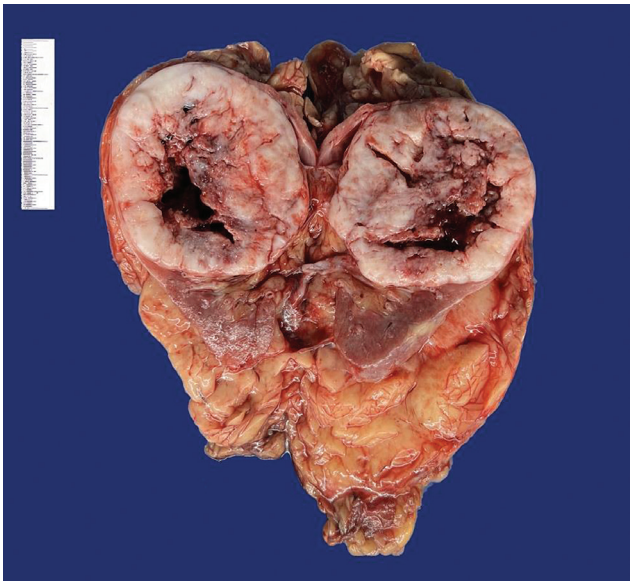

Figure 1. Gross appearance of the left kidney with a superior to mid-pole mass (bivalved).

x 5.2 x 5 cm (Figure 1). The renal pelvis, renal sinus soft tissues, perinephric fat, ureter and hilar blood vessels were grossly uninvolved by the mass.

Microscopic sections showed an unencapsulated, fairly defined cellular tumor (Figure 2) composed of elongated and anastomosing tubules seen within a basophilic myxoid to mucinous stroma (Figure 3). These structures were composed of cuboidal cells with low-grade nuclei, merging with bland spindle-shaped cells (Figure 4). No evidence of sarcomatoid or rhabdoid differentiation was seen.

Immunohistochemical studies using CK7, alpha-methylacyl-CoA racemase (AMACR), PAX8, CD10, and renal cell carcinoma antigen (RCCA) (Dako Agilent) were also performed. PAX8 showed moderate to strong, diffuse nuclear expression, while CK7, AMACR, and RCCA showed strong, diffuse, cytoplasmic expression in the tumor cells (Figure 5 A-D). No expression of CD10 was observed in the tumor (Figure 5 E).

DISCUSSION

The case described highlights the classic clinical and histologic features of mucinous tubular and spindle cell carcinoma. This tumor is mostly seen in adult patients, and predominantly affects females, with a reported female-to-male ratio of up to 4:1.[2] MTSCCs usually present asymptomatically as an incidental finding, which was the case for our patient. However, MTSCCs may also present as an abdominal mass, or with flank pain and hematuria.[1,2] It has also been proposed that there may be some association with nephrolithiasis and chronic kidney disease.[4]

Grossly, MTSCCs are well-circumscribed, sometimes partially encapsulated tumors located within the renal cortex.[2] Cut surfaces are solid, and homogenous tan to grey or pale yellow. Hemorrhage and necrosis are uncommonly present.[1] While MTSCC may be easy to recognize when it presents with classic histomorphology, a mucin-poor

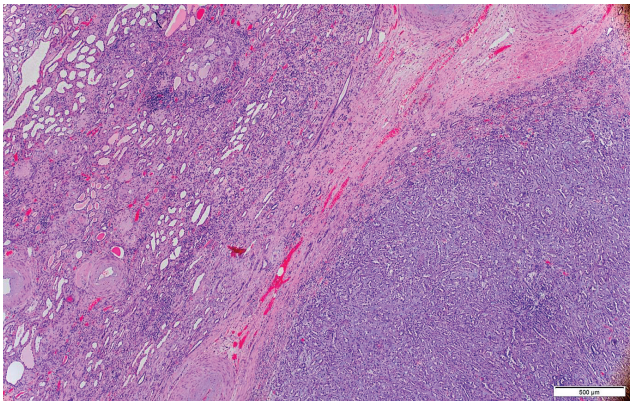

Figure 2. Well-delineated and unencapsulated cellular tumor with uninvolved renal parenchyma (H&E, 40x).

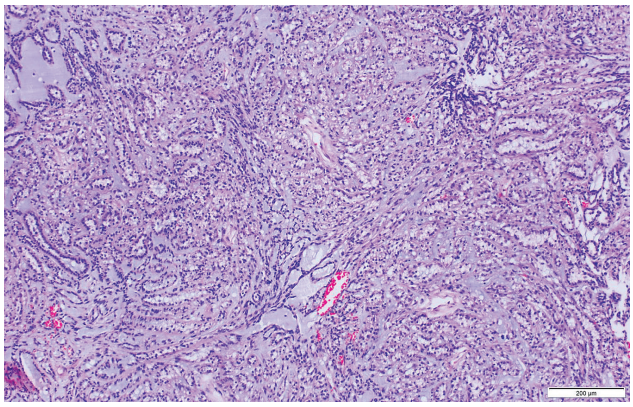

Figure 3. The tumor consists of tubules seen within a basophilic myxoid to mucinous stroma (H&E, 100x).

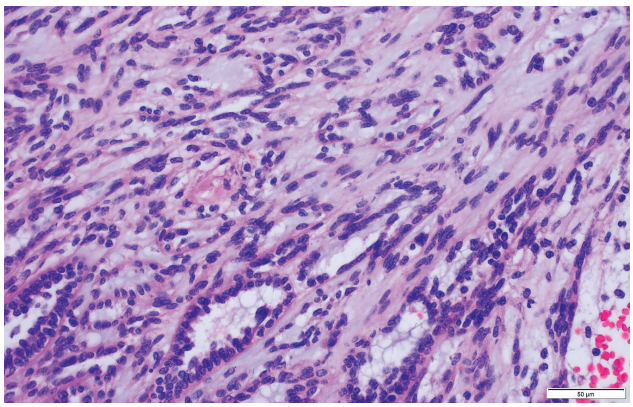

Figure 4. Tubules are composed of cuboidal cells with low-grade nuclei, merging with spindle-shaped cells (H&E, 400x).

variant has been described, which may be confused with a papillary renal cell carcinoma.[2,4] High-grade features such as sarcomatoid changes and necrosis have also been reported to occur in MTSCCs.[3,5] Other variations in the morphology of MTSCC may prompt consideration of other renal tumors. MTSCCs with predominantly spindle cells may be confused with sarcomatoid transformation of other renal cell carcinomas or mesenchymal neoplasms.[2] Clear cell changes have also been reported and may be confused with the more common clear cell renal cell

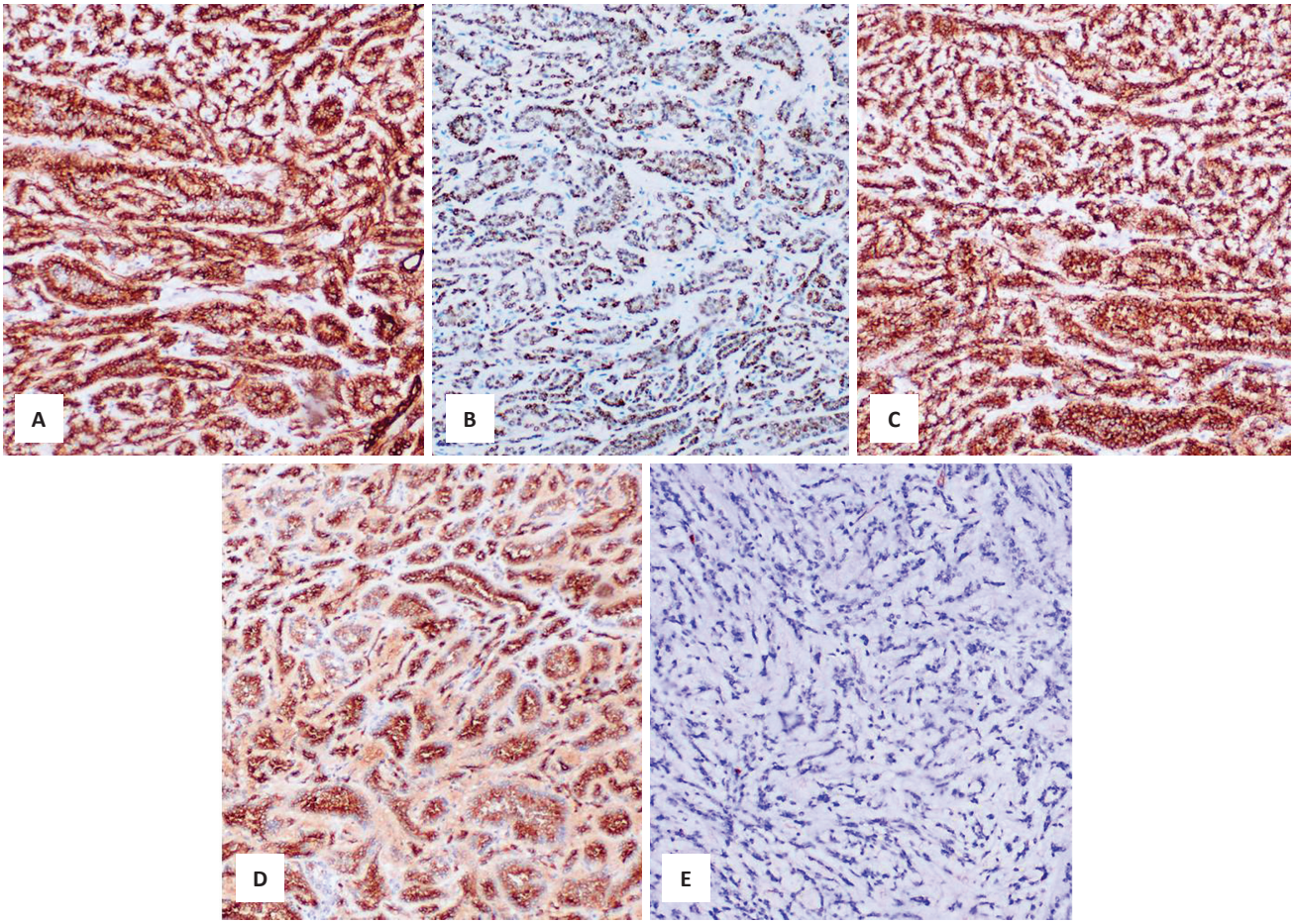

Figure 5. Immunohistochemistry expression of the renal tumor (400x). The tumor stained positively for CK7 **(A)**, PAX8 **(B)**, AMACR **(C)**, and RCCA **(D)**. No staining was seen using CD10 **(E)**.

carcinoma.[6] Psamomma bodies and aggregates of foamy macrophages may also be seen in MTSCCs, papillary renal cell carcinoma, and Xp11 translocation renal cell carcinoma.[7]

Immunohistochemistry studies may further complicate the distinction of MTSCCs from its closest differential. MTSCCs, like papillary renal cell carcinomas, typically demonstrate expression of CK7, AMACR, PAX8.[2,4,5,8] This immunohistochemical profile has led some authors to speculate the origin of MTSCCs from the proximal renal tubules,[1,2] but at present, the exact histogenesis of MTSCCs has remained unclear.[5,9] However, MTSCCs have also been reported to have lower expression of CD10 as compared to papillary renal cell carcinoma.[3,10]

When distinction cannot be made by examination of the histomorphology and immunohistochemical expression, MTSCCs may be distinguished from other renal neoplasms using molecular studies. Cytogenetic studies such as karyotyping, fluorescence in situ hybridization (FISH) and comparative genomic hybridization (CGH) studies frequently demonstrate chromosomal losses involving chromosomes 1, 4, 6, 8, 9, 13, 14, 15, and 22.[7,8,10-12] This is in contrast to papillary renal cell carcinomas, which are associated with trisomy 7, 17, or loss of Y chromosome.[7,8,10-12] Loss of 3p or mutations in the *VHL* gene, seen in clear cell renal cell carcinomas, are also not observed in MTSCCs.[7,11] Other investigations involving the use of novel markers, such as VSTM2A by RNA in situ hybridization, have also shown its possible use as a sensitive and specific marker for the diagnosis of MTSCC.[13]

The management of MTSCCs is surgery, with successful treatment via complete surgical excision or radical nephrectomy.[2,3,5,14] However, tumor recurrence, lymph node and/or distant metastasis have been occasionally reported, and these findings may be correlated with high-grade features.[1,2] Tumor progression, associated with loss of CDKN2A/B and additional complex genomic abnormalities, is seen in these more aggressive MTSCCs.[5,11] Nevertheless, the majority of MTSCCs are still considered to have favorable prognosis.[1-3]

CONCLUSION

Mucinous tubular and spindle cell carcinoma of the kidney is a rare, frequently indolent neoplasm of the kidney with recognizable histomorphologic features, as seen in the presented case. Immunohistochemical examinations done for this case also parallel previous studies. Molecular studies, such as karyotyping, FISH, or CGH analysis may be performed when distinction from more aggressive entities is difficult. Prompt recognition of this tumor is important, as it influences treatment and prognosis.

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ETHICAL CONSIDERATIONS

Patient consent was obtained prior to submission of this manuscript.

STATEMENT OF AUTHORSHIP

The authors certified fulfillment of ICMJE authorship criteria.

AUTHOR DISCLOSURE

The authors declared no conflict of interest.

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