

A rare co-occurrence of tubulocystic and clear cell type renal cell carcinoma

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ABSTRACT

Tubulocystic renal cell carcinoma (TC-RCC) is a recently described rare subtype of RCC. Less than 70 cases have been reported until date. The concurrent papillary RCC (P-RCC) and TC-RCC has been documented many times in the literature, but the co-occurrence of clear cell RCC (CC-RCC) and TC-RCC is very rare. We are probably describing the third case of TC-RCC occurring with CC-RCC in a 62-year-old male who presented with heaviness of left upper abdomen and dull intermittent pain in the left flank. Grossly, there was two distinct parts in the total nephrectomy specimen. The larger solid part displayed the histopathological features of CC-RCC and the smaller spongy part revealed the features of TC-RCC.

Key words: Low-grade collecting duct carcinoma, renal cell carcinoma, synchronous, tubulocystic carcinoma

INTRODUCTION

Tubulocystic renal cell carcinoma (TC-RCC) is a recently described rare subtype of RCC, which has not yet been included in the World Health Organization (WHO) 2004 classification of renal tumors.^[1] In total, less than 70 cases have been reported until date^[2] and the literature describing their clinicopathological information, biologic behavior, immunohistochemical profile, ultrastructural features and the important differential diagnostic considerations is even more limited.^[3,4] College of American Pathologists already recognized the new entity in the protocol for invasive carcinoma of renal tubular origin.^[5] The most recent International Society of Urological Pathology (ISUP) Vancouver Modification of WHO 2004 classification popularly known as the ISUP Vancouver Classification of Renal Neoplasia recommends the inclusion of TC-RCC along with four other new distinct epithelial tumors in the classification system.^[2] The concurrent P-RCC and TC-RCC has been documented many times^[6] but the co-occurrence

of CC-RCC and TC-RCC is very rare. To the best of our knowledge, this is the third case^[7,8] of TC-RCC occurring with CC-RCC in a 62-year-old male.

CASE REPORT

A 62-year-old male presented with heaviness of left upper abdomen for last 9 months and dull intermittent pain in the left flank, off and on for last 5 months. There was no significant history of weight loss and hematuria during this period. History of fever with associated urinary complaints was also conspicuously absent. On clinical examination, there was mild pallor and mild tenderness in the left flank. There was no palpable superficial lymph node. The results of his routine hematological and biochemical examinations were within normal limits. Contrast-enhanced computed tomography (CECT) scan revealed a solitary heterogeneously enhancing well-delineated mass with a small area of cystic change in the upper pole of the left kidney without any noticeable infiltration of adjacent organ. Retroperitoneal lymph nodes did not appear to be enlarged on CECT. There was no feature of associated hydronephrosis or calculi. Further, no distant metastases were appreciated on CECT chest or bone scan. He underwent a left total nephrectomy without any significant post-operative complication.

On gross examination, the mass was well-circumscribed having two distinct parts separated by intact fibrous

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capsule. The larger solid part of the mass was yellow to reddish measuring 4.5 cm × 3.8 cm × 3.5 cm confined to the cortical region of the upper pole of the kidney. Small areas of hemorrhage were noted in this area. The relatively smaller spongy multicystic part consisting of thin-walled translucent small to intermediate sized cystic structures containing clear serous fluid measuring 2.4 cm × 2.2 cm × 1.8 cm was restricted in the cortical region and corticomedullary junctional region. No solid components or hemorrhagic or necrotic areas were identified in this part. The whole mass was confined to the kidney. No renal capsule penetration or vascular invasion was appreciated macroscopically.

On histopathological examinations, the larger mass displayed solid nests of cells with abundant clear cytoplasm having well-demarcated distinct cell borders and hyperchromatic nuclei with inconspicuous nucleoli (Furhman nuclear grade 2) separated by a network of small thin walled capillaries. Nuclear pleomorphism was mild and noticed at places. Sarcomatoid features could not be appreciated in the sections. This part of the mass was clearly separated from smaller spongy part and from kidney tissue by distinct boundary [Figure 1a and b]. The smaller spongy lesion revealed an unencapsulated circumscribed mass composed of small to intermediate sized tubules with areas of cystic dilatation, dispersed evenly in a bland hypocellular fibrotic stroma [Figure 2a]. The tubules and cysts were lined by a single layer of flat, cuboidal to columnar epithelial cells with abundant eosinophilic or amphophilic cytoplasm, slightly irregular nuclear membranes and inconspicuous nucleoli (Fuhrman nuclear grade was 2). Cells also displayed focal hobnail configuration [Figure 2b]. No necrosis was seen and mitotic figures were extremely rare. There was no stratification or papillary configuration in the lining epithelial cells of the tubules and cysts. No areas displayed solid growth. Desmoplastic reaction or cellular ovarian like stroma was not appreciated in any part of the slides. The lesion also lacked the lymphovascular involvement, tumor necrosis, renal vein or adrenal involvement. Finally, the case was diagnosed as co-occurrence of TC-RCC and CC-RCC, Fuhrman nuclear grade 2, Stage T1bN0M0. The patient did not receive any adjuvant therapy and was alive and doing well after 3, 6 and 12 months of surgical resection with no evidence of recurrence or metastasis.

DISCUSSION

The histological features of the high-grade collecting duct carcinoma (CDC) of the kidney with hobnail cells occurring in the central region of the kidney were described for the 1st time in 1956 by Pierre Masson as “Bellinian epithelioma” or “carcinoma of Bellini (collecting) duct.”^[1] The neoplasm which is distinctly different from classic CDC in many ways, was described as “low-grade CDC” by MacLennan *et al.* in

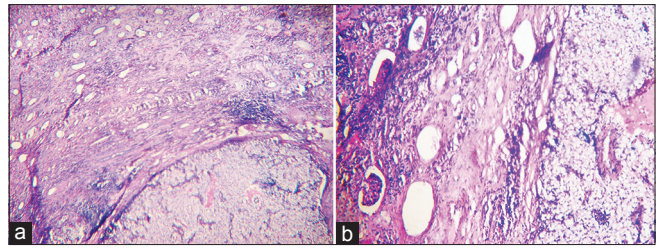


Figure 1: (a) Photomicrograph showing the interface of clear cell type of renal cell carcinoma (CC-RCC) and tubulocystic RCC (H and E, ×40) (b) Photomicrograph of CC-RCC with adjacent kidney tissue (H and E, ×100)

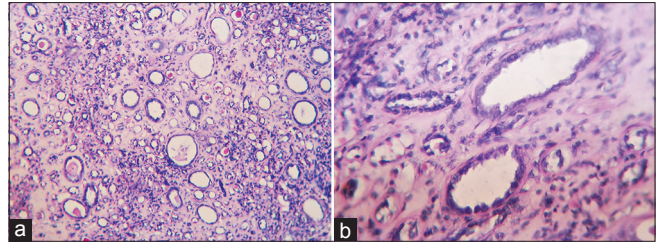


Figure 2: Histologic features of tubulocystic carcinoma. Lower magnification (a) Variably sized uniformly distributed tubular and cystic structures lined by a single layer of epithelium in a fibrotic to hyalinized stroma (H and E, ×100). Higher magnification (b) Displaying flat to cuboidal lining eosinophilic epithelial cells with focal hobnail configuration having mild nuclear membrane irregularities and inconspicuous nucleoli (H and E, ×400)

1997.^[9] In 2004, Amin *et al.* coined the term “TC carcinoma of the kidney” based on its characteristic morphology.^[1] The term should not be used in situations in which there is a TC pattern admixed with the usual elements of P-RCC or CDC.^[2]

The major differential diagnosis of TC-RCC includes cystic nephroma, multilocular cystic RCC, oncocytoma with prominent tubules and cysts and mixed epithelial and stromal tumor of the kidney and lastly the CDC which is characterized by aggressive behavior and highly infiltrative growth pattern.^[1,3,4,10] Interestingly, there were neither discriminating immunohistochemical markers nor conclusive evidence to identify a specific lineage of histogenesis for TC-RCC. The distinctive morphologic features of these tumors are enough for the diagnostic purposes.^[1] The distinctive clinico-radiological features, typical gross and microscopic features supported the diagnosis in the present case.

TC-RCC appears to have a favorable prognosis and presents at the lower stage with very few reports of local recurrence or metastatic disease.^[3] Further future evaluation of more cases is necessary for better understanding of the biology of this neoplasm and to ascertain its prognosis.

REFERENCES

1. Amin MB, MacLennan GT, Gupta R, Grignon D, Paraf F, Vieillefond A, *et al.* Tubulocystic carcinoma of the kidney:

- Clinicopathologic analysis of 31 cases of a distinctive rare subtype of renal cell carcinoma. *Am J Surg Pathol* 2009;33:384-92.
2. Srigley JR, Delahunt B, Eble JN, Egevad L, Epstein JI, Grignon D, *et al.* The International Society of Urological Pathology (ISUP) Vancouver Classification of Renal Neoplasia. *Am J Surg Pathol* 2013;37:1469-89.
 3. Alexiev BA, Drachenberg CB. Tubulocystic carcinoma of the kidney: A histologic, immunohistochemical, and ultrastructural study. *Virchows Arch* 2013;462:575-81.
 4. Yang XJ, Zhou M, Hes O, Shen S, Li R, Lopez J, *et al.* Tubulocystic carcinoma of the kidney: Clinicopathologic and molecular characterization. *Am J Surg Pathol* 2008;32:177-87.
 5. Srigley JR, Amin MB, Delahunt B, Campbell SC, Chang A, Grignon DJ, *et al.* Protocol for the examination of specimens from patients with invasive carcinoma of renal tubular origin. *Arch Pathol Lab Med* 2010;134:e25-30.
 6. Zhou M, Yang XJ, Lopez JL, Shah RB, Hes O, Shen SS, *et al.* Renal tubulocystic carcinoma is closely related to papillary renal cell carcinoma: Implications for pathologic classification. *Am J Surg Pathol* 2009;33:1840-9.
 7. Quiroga-Garza G, Piña-Oviedo S, Cuevas-Ocampo K, Goldfarb R, Schwartz MR, Ayala AG, *et al.* Synchronous clear cell renal cell carcinoma and tubulocystic carcinoma: Genetic evidence of independent ontogenesis and implications of chromosomal imbalances in tumor progression. *Diagn Pathol* 2012;7:21.
 8. Gönül II, Cakr A, Sözen S, Ataoglu O, Alkibay T. A case of tubulocystic carcinoma simultaneously occurring with clear cell type renal cell carcinoma and micropapillary urothelial carcinoma of bladder. *South Med J* 2009;102:754-7.
 9. MacLennan GT, Bostwick DG. Tubulocystic carcinoma, mucinous tubular and spindle cell carcinoma, and other recently described rare renal tumors. *Clin Lab Med* 2005;25:393-416.
 10. Azoulay S, Vieillefond A, Paraf F, Pasquier D, Cussenot O, Callard P, *et al.* Tubulocystic carcinoma of the kidney: A new entity among renal tumors. *Virchows Arch* 2007;451:905-9.

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