SYNCHRONOUS CLEAR CELL RENAL CELL CARCINOMA AND TUBULOCYSTIC RENAL CELL CARCINOMA

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ABSTRACT

Tubulocystic renal cell carcinoma (TCRCC) is a recently described rare subtype of RCC. To best of our knowledge less than 70 cases have been reported till date. The concurrent papillary RCC (PRCC) and TCRCC has been documented in literature, but the co-occurrence of clear cell RCC (CCRCC) and TCRCC is very rare. We are describing a rare case of incidentally detected TCRCC occurring with CCRCC in a 45 years old male who presented with high grade fever with chills and rigors. Grossly, there were two distinct tumors in the total nephrectomy specimen. The larger tumor displayed the histopathological features of CCRCC and the smaller tumor revealed the features of TCRCC.treatment in the present case.

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 was not included in the World Health Organization (WHO) 2004 classification of renal tumors.[1] It consists of a mixture of tubules and micro/macro cysts with low-grade nuclear features and appears to derive from proximal convoluted tubule and distal nephron. The mean age is 54 year and 85% of patients are male[2]. In total, less than 70 cases have been reported until date[3] and the literature describing their clinicopathological information, biologic behavior, immunohistochemical profile, ultrastructural features and the important differential diagnostic considerations is even more limited.[4,5] College of American Pathologists already recognized the new entity in the protocol for invasive carcinoma of renal tubular origin.[6] 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.[3] The concurrent P-RCC and TC-RCC has been documented many times[7] but the co-occurrence of CC-RCC and TC-RCC is very rare. To the best of our knowledge, this is the fourth case[8,9] of TC-RCC occurring with CC-RCC in a 45-year-old male.

CASE REPORT

A 45-year-old male presented with high grade fever with chills and rigors before two months. There was no significant history of weight loss, loin pain, hematuria or lower urinary tract symptoms during this period. On clinical examination, the patient was afebrile. There was mild tenderness in the right flank. There was no palpable superficial lymph node. The results of his routine hematological and biochemical examinations were within normal limits. Ultrasonography(USG-KUB) revealed a solitary exophytic mass measuring 3.8x3.8x2.6cm arising from the lower pole of right kidney. There was no feature of associated hydronephrosis or calculi. There was no hilar lymphadenopathy. He underwent a laparoscopic right radical nephrectomy without any significant post operative complication. There were no adhesions surrounding the kidney. On gross examination, two separate tumors were identified [figure 1]. The larger solid tumor was hemorrhagic and grayish in colour measuring 3x2.5x1.5 cm confined to the cortical region of the lower pole of the kidney. The smaller spongy tumor measuring 1.5x1cm was restricted in the cortical region and corticomedullary junctional region of the upper pole of the kidney. No hemorrhagic or necrotic areas were identified in this part. Both the tumors were confined to the kidney. No renal capsule infiltration or vascular invasion was appreciated macroscopically. On histopathological examinations, the larger nodule was

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well circumscribed composed of multiple cystic spaces lined by tumor cells as well as solid nests of cells [Figure 2A,2B]. These cells were polygonal with abundant clear cytoplasm having well-demarcated distinct cell borders and hyperchromatic nuclei with irregular nuclear membrane and conspicuous nucleoli (Furhman nuclear grade 3) separated by a network of small thin walled capillaries [Figure 2C]. Sarcomatoid features could not be appreciated in the sections. The smaller nodule revealed an unencapsulated well circumscribed tumor composed of small to intermediate sized tubules with areas of cystically dilated larger tubules, dispersed evenly in a bland hypocellular fibrotic stroma [Figure 3A, 3B]. 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 3C]. Intraluminal foam cells were also seen in some of the tubules [Figure 3D]. 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 sinus or pelvicalyceal system involvement. Immunohistochemistry (IHC) showed expression of CD10 and vimentin in both the tumors. CK7and 34betaE12 were expressed only in upper polar nodule confirming the diagnosis of TC-RCC [Figure 4A&4B]. Finally, the case was diagnosed as co-occurrence of TC-RCC and CC-RCC, Fuhrman nuclear grade 3, Stage T1bN0M0. Postoperative was uneventful. He was lost to follow up after one month.

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 1997.[10] In 2004, Amin et al. coined the term "TC carcinoma of the kidney" based on its characteristic morphology.[2]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.[3] 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.[2,4,5,11] 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.[2] The distinctive clinicoradiolgical 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.[4] Further future evaluation of more cases is necessary for better understanding of the biology of this neoplasm and to ascertain its prognosis.

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Figure 1: Right radical nephrectomy showing lower pole an exophytic growth with hemorrhage and necrosis (thick arrow, and upper pole showing a small circumscribed growth (thin arrow).

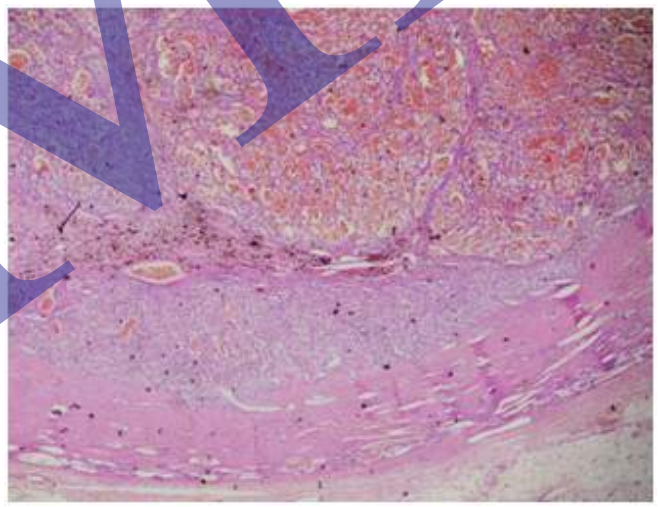

Figure 2A (2X): Well circumscribed hemorrhagic tumor composed of cystic and solid areas

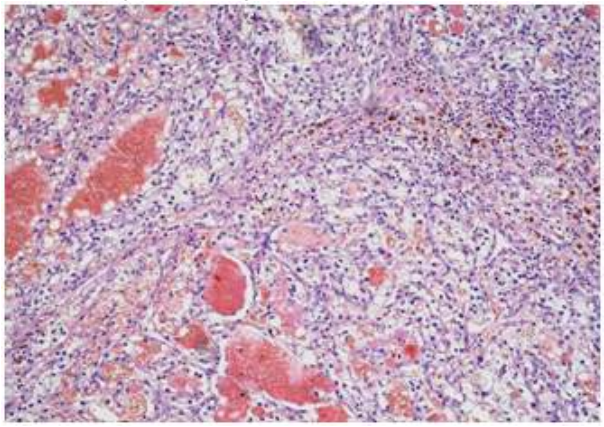

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Figure 2B(10X): multiple cystic spaces lined by tumor cells as well as solid nests of cells.

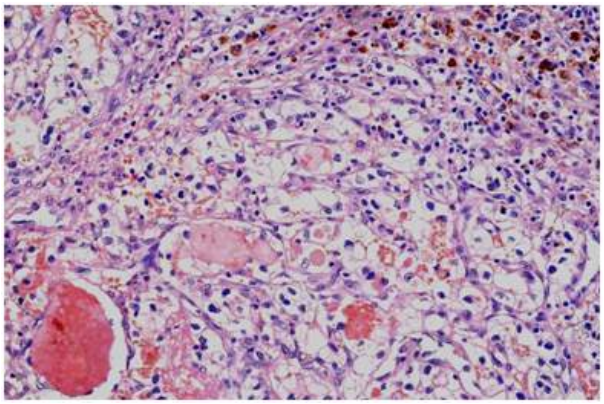

Figure 2C(40X): polygonal cells with abundant clear cytoplasm, irregular nuclear borders and conspicuous nucleoli

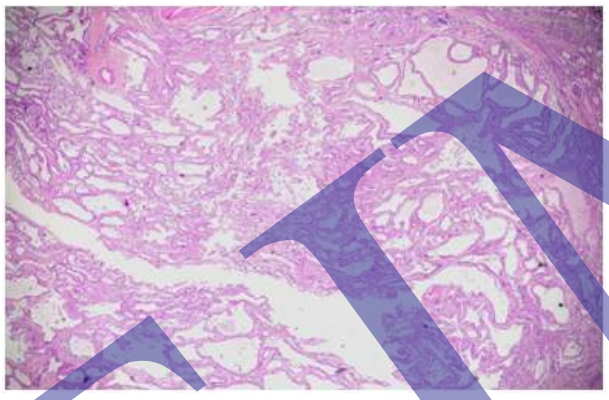

Figure 3A: 2X- well circumscribed tumor composed of small to intermediate sized tubules with areas of cystically dilated larger tubules

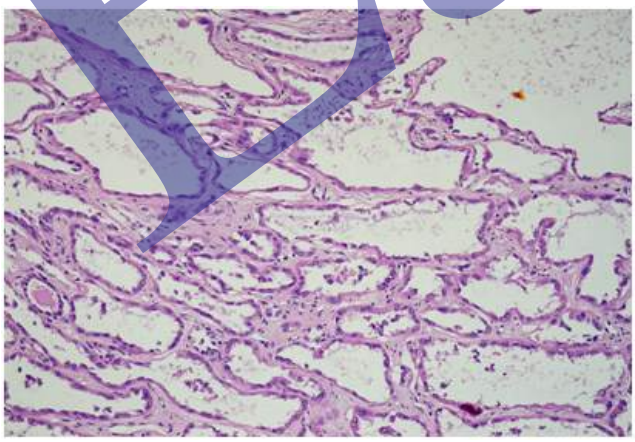

Figure 3B: 10X- Tubules dispersed evenly in a bland hypocellular fibrotic stroma

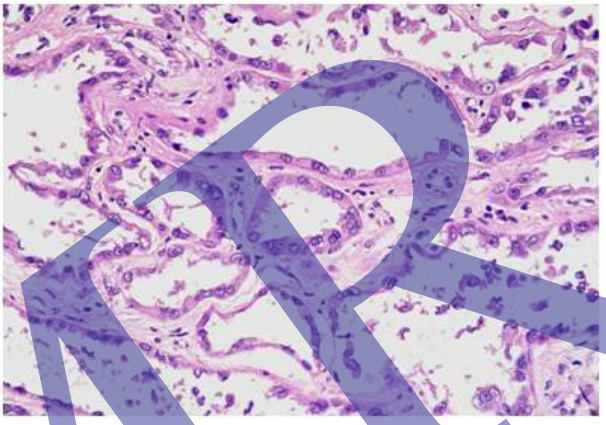

Figure 3C: 40X- The tubules and cysts lined by a single layer of flat, cuboidal to columnar epithelial cells with abundant eosinophilic or amphophilic cytoplasm, slightly irregular nuclear membranes and inconspicuous nucleoli with focal hobnail configuration

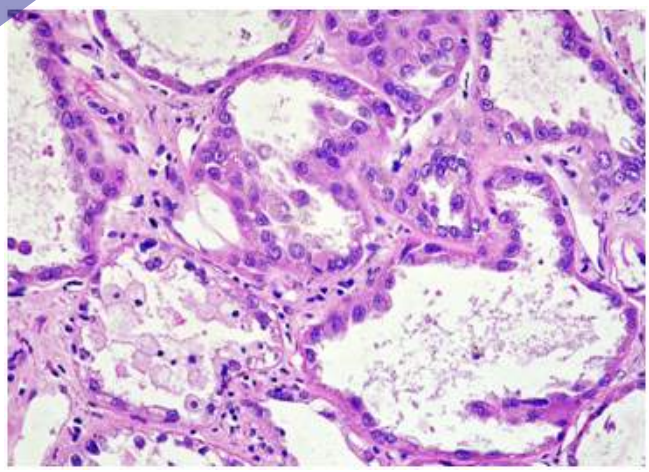

Figure 3D: 40X- Intraluminal foam cells in some of the tubules

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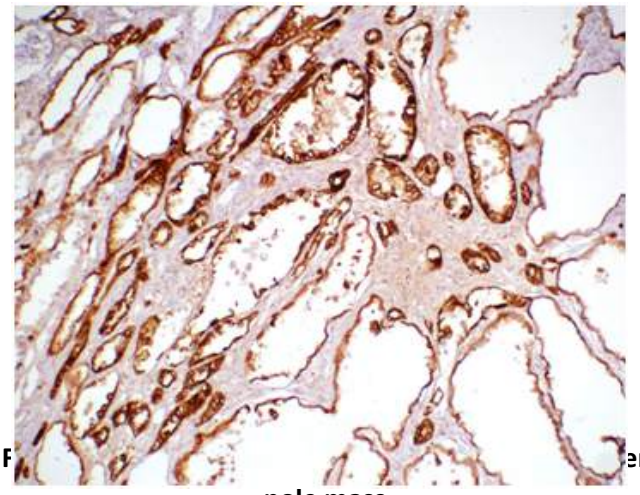

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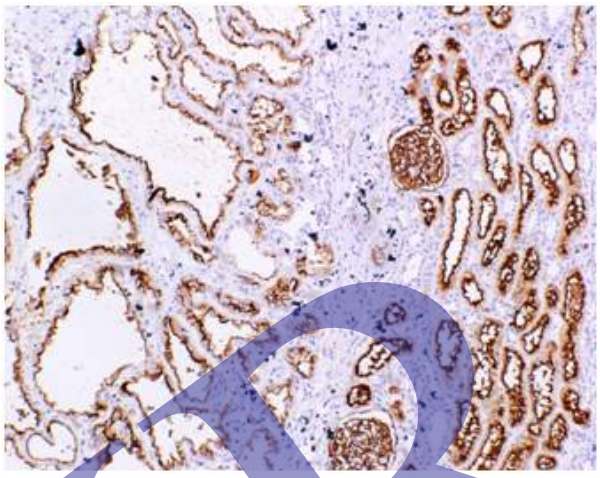
pole mass



upper pole mass

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