Clear cell tubulopapillary renal cell carcinoma: A case report

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INTRODUCTION

Clear cell (tubulo) papillary renal cell carcinoma (CCPRCC) was recently established as one of the five new distinct renal tumor entities by The Classification Working Group of the International Society of Urological Pathology in a consensus conference held at Vancouver, Canada.[1] The majority of cases have been reported as sporadic, although cases associated with end-stage renal disease are also well documented.[2] Microscopically, CCPRCC is composed of an admixture of cysts, tubules, acinar glands, papillae, and even solid sheets. Although the characteristic linear arrangement of the low-grade nuclei away from the basal aspect (the so-called inverted polarity) is fairly unique to CCPRCC, the classification committee strictly requires the immunostaining pattern should be satisfied simultaneously.[1] Immunohistochemically, the tumor cells should have a profile of CK7+, CA-IX+, high-molecular weight cytokeratin (HMWCK)+, CD10−, and Alpha Methyl Acyl Coenzyme-A Racemase (AMACR). No cases with metastasis have been reported.[3] If further studies confirm their apparent indolent course, it is possible that neoplasms will subsequently be reclassified as being of “low malign potential” rather than as carcinoma.[1]

CASE REPORT

The patient was a 73-year-old man. Computed tomography revealed a mass lesion measuring 25 mm in the middle portion of right kidney [Figure 1]. Partial nephrectomy was performed. Macroscopic material was measuring $3.7 \times 3.5 \times 2$ cm. There was a well demarcated mass in diameter 2.4 cm and renal parenchyma was tumor-free macroscopically. Cut surface of tumor was solid and focally cystic. Microscopically, the tumor is composed of clear cell of low nuclear grade, variable papillary–tubular–acinar architecture [Figure 2]. The characteristic linear arrangement of nuclei deviating from basal aspect of lumen was prominent [Figure 3]. In immunohistochemical analyses following antibodies CK7, AMACR, CD10, TFE-3, RCC, Vimentin, HMW-CK, WT-1 were used. Tumor cells were positive for CK7, Vimentin, HMW-CK and negative for CD10, TFE-3, AMACR, WT-1, RCC [Figure 4].

DISCUSSION

CCPRCC is composed of cells with clear cytoplasm lining cystic, tubular, and papillary structures. CCPRCC is one of these entities. Williamson et al. reviewed their resection specimen diagnosed clear cell renal cell carcinoma/papillary renal cell carcinoma and found 14 CCPRCC (14/469; 3%).[4] The majority of cases have been reported as sporadic, although cases associated with end-stage renal disease are also well documented.[3] CCPRCC is generally well defined and well encapsulated. In our case the tumor was 2.4 cm diameter and well demarcated. Microscopically, the presence of branching tubules/acini and/or complex clear cell ribbons in the cysts with fibrotic stroma is the key to the identification of these tumors. Nuclei of most CCPRCC have horizontally linear arrangement apart from the basement membrane.[5] The tumor has a distinct immunophenotype. All tumors showed strong diffuse staining for CK7 but negative for CD10.[1] Our case was positive for CK7, Vimentin and negative for AMACR, CD10, TFE-3, WT-1. Differential
diagnosis between CCPRCC and multilocular cystic renal cell carcinoma is possible with the presence of macroscopically and microscopically solid areas in CCPRCC and negativity CD10, RCC.[6] Partial nephrectomy or total nephrectomy is generally the treatment provided for a solitary tumor when surgical resection is feasible. As clear cell papillary renal cell carcinoma has been only recently described, they generally present at low stage and have a low nuclear grade.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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