

Papillary Renal Cell Carcinoma Type-II: A Distinct clinicopathological Subtype of Renal Epithelial Neoplasm

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ABSTRACT

Papillary Renal Cell Carcinoma (PRCC) has been recognized as a distinct clinicopathologic subtype of Renal Cell Carcinoma (RCC). Herewith we present a case of 45 year old male patient having complaints of left lower flank pain and hematuria of 1 month duration. Ultrasonography showed large homogenous mass measuring 9.1x9x6.1cm at upper pole of left kidney. Doppler showed mild to moderate vascularity with low resistance to flow. Small cystic areas with calcifications were noted. On radiological finding, diagnosis suggestive of renal cell carcinoma was given. Left radical nephrectomy was done. On histopathology evaluation; diagnosed as PRCC Type-II. As the prognosis of PRCC Type-II is poorer than Type-I, these tumors should be properly evaluated.

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Introduction

PRCC comprises about 10-20% of RCC and it is distinct morphological entity.^[1,2] The term chromophil RCC has been used to describe these tumour.^[3] PRCC having distinct histologic criteria for its diagnosis and grading which is helpful for clinical outcome of these malignancies when treating the patient.

Case Report

A 45 year old male patient came with history of pain in abdomen and left abdominal palpable mass since 1 month which gradually increased in size. He is tobacco chewer since 15 years. On routine hematological examination Hb-10.5gm%, polymorphs-78%, lymphocytes 20%, eosinophils 2%, platelets-3lacs/cu³. Routine urine examination showed mild hematuria and pyuria. Renal function tests were normal. X-ray chest showed no evidence of metastases. Ultrasonography pelvi-abdomen showed large homogenous mass measuring 9.1x9x6.1cm at upper pole of left kidney. Doppler showed mild to moderate vascularity with low resistance to flow. Small cystic areas with calcifications noted. Radiological diagnosis -suggestive of renal cell carcinoma was given. Other kidney was normal and rest of pelvi-abdominal organs showed no significant abnormality. No any other significant contributory personal, past or family history was there. Left radical nephrectomy was done and specimen was sent for histopathological examination. The postoperative period was uneventful.

Gross: We received left nephrectomy specimen with perinephric fat, totally measuring 15x11x10 cm and weighing

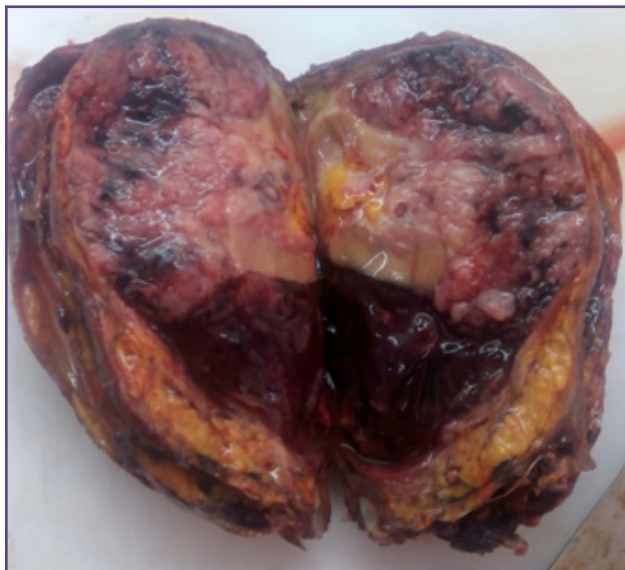


Fig. 1: Left nephrectomy specimen, cut section showing large yellow to brown tumor mass with areas of hemorrhage and necrosis.

700g. Renal surface was irregular, thickened and showed two nodules. Also perinephric fatty tissue at hilum showed three nodular structures measuring approximately 1cm in diameter. Cut section of kidney showed large, circumscribed grey brown to dark tan colored mass with variegated appearance measuring 9x6x5 cm. Areas of hemorrhage, necrosis were prominent (Figure-1). Tumor was replacing almost all renal tissue and extending over renal capsule, pelvis and perinephric fat. Renal vessels were not involved.

Microscopy: Multiple sections showed renal tissue with a tumor composed of predominantly papillary pattern (90%) with solid areas (Figure-2). The papillae were large, discrete forming fronds and lined by one-two layered large round to elongated cells having moderately pleomorphic nuclei and 1-2 prominent nucleoli and abundant eosinophilic cytoplasm (Figure-3). In areas pseudostratification was noted. Papillary core was infiltrated by inflammatory cells. Large areas of hemorrhage, necrosis was noted. Few psammoma bodies were seen. Tumor was infiltrating renal capsule, pelvi-ureter and nodules in the perinephric fat. Lymph nodes were negative for tumor infiltration. The histopathological diagnosis was given as papillary renal cell carcinoma type-II grade-III of left renal mass.

Discussion

The incidence of RCC is rising worldwide. RCCs on histopathology has been classified into approximately clear cell (70%), papillary (15%), granular cell (8%), chromophobe (5%), sarcomatoid (1-2%) and collecting duct type (0.5%).^[3]

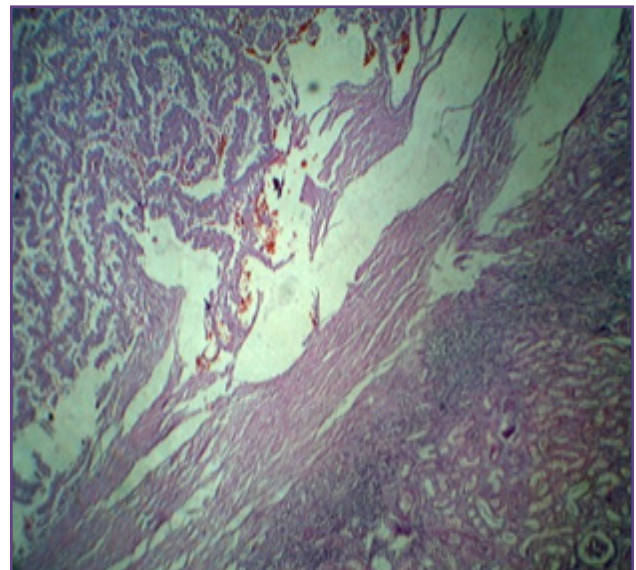


Fig. 2: photomicrograph showing renal tissue with a tumor composed of predominantly papillary pattern. (H&E stain, x100).

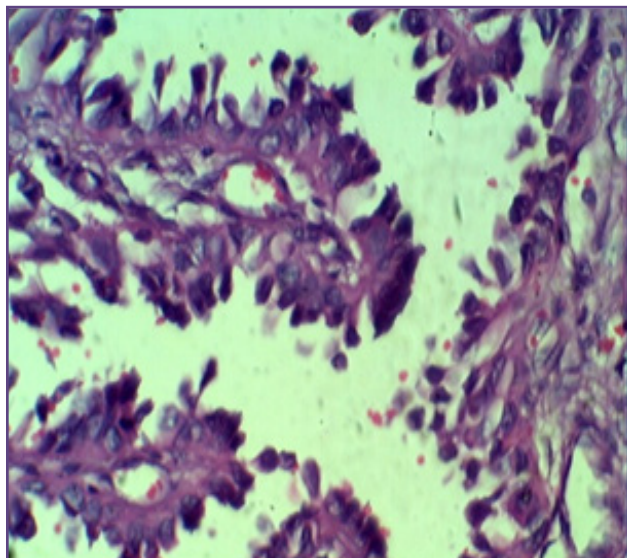


Fig. 3: photomicrograph showing large papillae forming fronds and lined by one-two layered large round to elongated cells having moderately pleomorphic nuclei and 1-2 prominent nucleoli and abundant eosinophilic cytoplasm (H&E stain, x400)

PRCC represent about 10-20% of all RCCs, with male predominance (male: female ratio 2 to 5:1).^[4] The mean age of patient presentation is 61.8 years (range 22-83).^[5] Various etiological factors includes hereditary, autosomal dominant transmission, heavy metal exposure, drugs, tobacco etc. have been described. Development of multiple tumors and bilateral renal masses are noted in PRCC.^[6] In our case the other kidney was normal. Clinically mostly they present with abdominal mass, pain and hematuria. On gross examination PRCC are usually well circumscribed and may show a fibrous pseudocapsule. On cut section tumor appears yellow to brown with large areas of hemorrhage and necrosis, occasional cases shows cystic change.

PRCC has been recognized as a distinct clinic-pathologic subtype of RCC. Diagnostic histological pattern given for PRCC is carcinoma of kidney with a predominant papillary pattern. The two types have been identified on the types of cells lining papillae.^[7] In type-I papillae are lined by small cells with clear to basophilic cytoplasm. The other associate frequent findings are foamy macrophages, edema in papillary cores, few psammoma bodies and glomeruloid papillae and these are usually called low grade PRCC. While in type-II papillae are lined by large cells with abundant eosinophilic cytoplasm. The nuclei show frequently pseudo stratification and prominent nucleoli. These are considered high grade PRCC. Our case was PRCC

Type-II with nuclear grade III according to Fuhrman's grading.^[8] PRCC predominantly shows papillary pattern on histomorphology, however areas tubular, solid and spindle cell (sarcomatoid) component were also reported. Tumors with cytoplasmic features of eosinophilia (42%), basophilia (34%) and mixed (24%) were distinguished.^[5] Eosinophilic tumors were predominantly of high grade. As necrosis and phagocytic activity within a tumor become quiet extensive, cytoplasmic clearing in some tumors may cause morphologic confusion with clear cell RCC.

The differential diagnosis for PRCC is renal adenoma, clear cell RCC with foci of papillary areas, clear cell tubo-papillary carcinoma, papillary oncocyoma etc. In papillary adenoma it shows similar morphology and tumor with or without fibrous capsule but size of tumor is 5mm or less by WHO criteria. In case of Clear cell carcinoma with focal papillary architecture the neoplasm have dyshesive tumor areas may have pseudopapillary appearance. Papillary renal cell carcinoma with clear cells is a novel entity with a unique clinical and histopathological features.. The presence of clear cells is associated with aggressive pathological characteristics and poorer prognosis.^[9] In case papillary Oncocyoma on microscopy no classic areas of chromophobe carcinoma, no prominent cell membranes, no crinkle or raisinoid nuclei, no binucleation, no abundant microvesicles. These lesions should be kept in mind and carefully evaluated on routine H&E histopathology for proper diagnosis and typing of the tumor. In most of these cases histopathological features are sufficient to make correct diagnosis.

The cases of PRCC usually present at an early stage and has better 5 year survival rates (82-90%) than does RCC of the same stage (65-70%).^[5] The comparison of the type 1 PRCCs with type 2 PRCCs revealed that type 2 tumors were associated with a greater stage and grade and microvascular invasion significantly.^[10] The overall and disease-free survival rate was 89% and 92% in type 1 tumors and 55% and 44% in type 2 tumors, respectively.^[10] Our patient received surgical resection and in on regular follow-up and doing well.

Conclusion

The diagnosis of RCC with both papillary architecture and cell with clear cytoplasm can be challenging for pathologist. Histologic subtype of renal carcinoma as PRCC Type-II appears to have poor prognostic implications and therefore it is important to diagnose and grade this tumor for better management of the patients.

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Competing Interests

None Declared

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