



## Metachronous Rapidly-Growing Papillary Type 2 Renal Cell Carcinoma following Contralateral Clear Cell Renal Cell Carcinoma Diagnosis – A Case Report from the Radiologic Point of View

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### Abstract

Papillary Renal Cell Carcinoma Type 2 (PRCC2) is a relatively rare aggressively- behaving tumor associated with poor prognosis. We present the case of a 74 year- old patient who underwent a unilateral radical nephrectomy 9 months prior due to a grade 4 clear cell renal cell carcinoma diagnosis, which had a newly formed 20mm renal mass suspected as malignant incidentally found during a routine MRI which was not evident in a CT scan 4 months before. This report follows the diagnostic process, radiological findings, and management in the unique case of the relatively rare rapidly progressing metachronous malignancy from the radiologic point of view, discussing the challenges of diagnosis and clinical staging affecting the patient's outcome.

**Keywords:** Papillary Renal Cell Carcinoma Type 2 (PRCC2); Metachronous Renal Cell Carcinoma; Clinical Staging; Radiology

### Introduction

Renal Cell Carcinoma arises spontaneously, and account for 2-3% of all malignancies[1]; Papillary tumors are the second-most common renal malignancy, accounting for 10-15% of cases, being either type 1 which is more associated with hereditary syndromes, or type 2 being less common and associated with worse prognosis[2,3]. Both types have a characteristic appearance on CT and are associated with minimal contrast enhancement, resulting in the fact they can be misinterpreted as a hyperdense cyst if an unenhanced examination has not been performed.

Many radiologic modules are used in diagnosis of Renal Cell Carcinoma[4] – US with color Doppler is useful for detecting IVC thrombus and extension of tumor thrombus into the intrahepatic vena cava. Computed Tomography appearance is mostly heterogeneous on unenhanced CT due to their solid consistency with attenuation values of more than 20 HU on unenhanced CT and an increase of attenuation larger than 20 after IV contrast administration indicating malignancy; Tumors occurring in non-functioning kidney may show little enhancement due to papillary subtype or poor renal arterial blood flow. Lastly, MRI can be used to detect and stage renal cell carcinoma, with a sensitivity similar to that of CT, appearing iso- or hypointense compared with the renal cortex on T1 weighted sequence, and slightly hyperintense on T2 weighted sequence; heterogeneous enhancement following IV gadolinium administration occurs immediately, decreasing on delayed images.

Besides the challenges mentioned above in regarding the detection of papillary type 2 renal cell carcinoma, the challenges from the radiologic point of view performing accurate clinical staging are also well known – many

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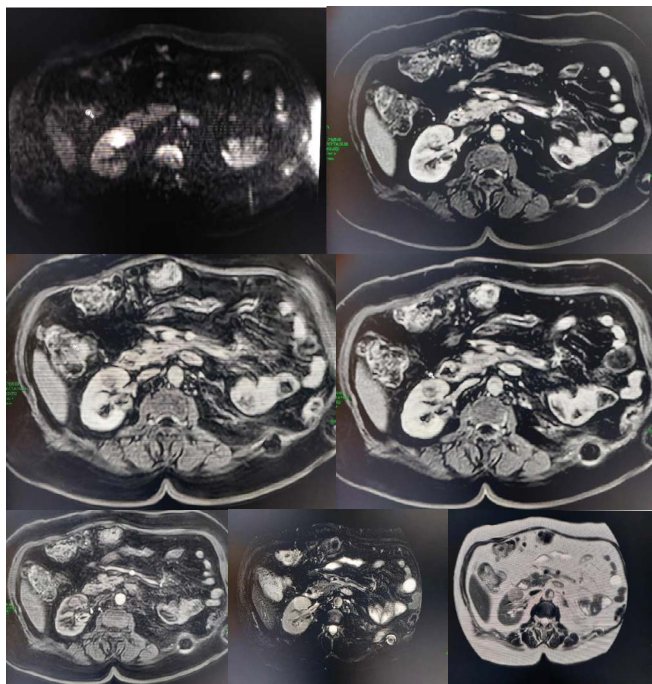
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studies have shown that there is radiologic over-estimation of renal mass sizes in comparison to the pathological staging done after the procedure[5,6,7,8,9,10]; this discrepancy can lead to a more radical approach considering the treatment regimen, resulting in a more extensive surgical procedure performed such as total nephrectomy instead of a nephron-sparing surgery when possible.

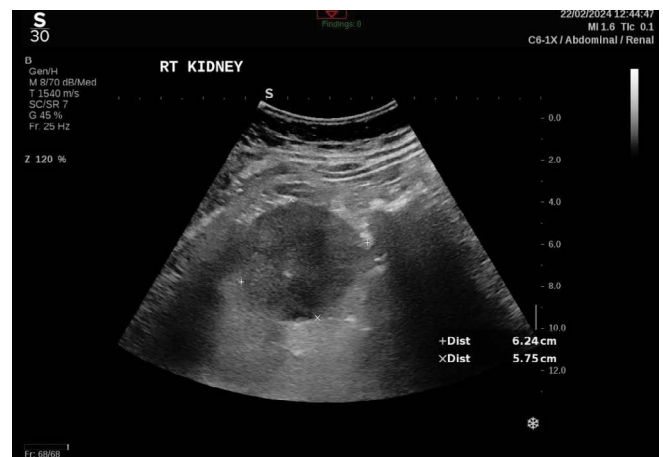
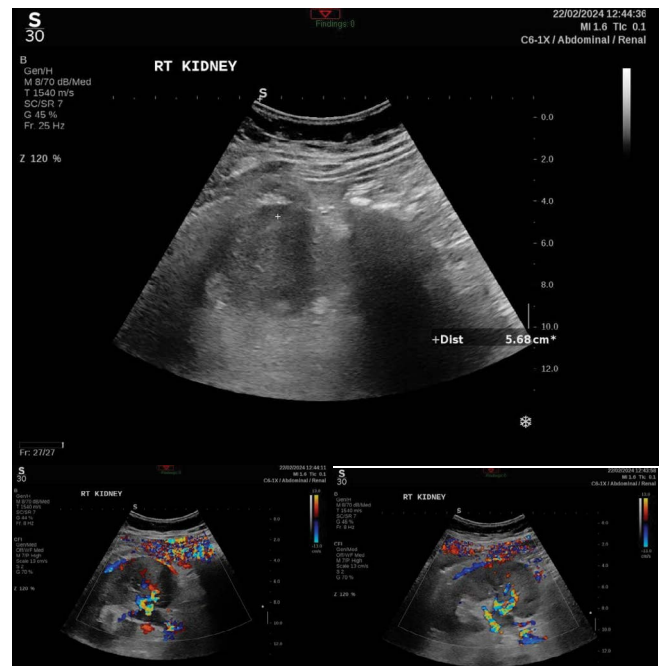
## Case Presentation

A 74 year-old patient with a history of unilateral radical nephrectomy following clear cell RCC (grade 4) had a routine Abdominal MRI+MRCP scan 9 months after surgery due to a hypodense pancreatic lesion suspected to be IPMN incidentally discovered in abdominal CTA prior to surgery, presented with a hyperintense T<sub>2</sub> weighted 20mm mass DWI-restricted gaining slight heterogeneous enhancement following IV gadolinium administration, suspected as malignancy in the remaining kidney which appeared healthy in a CT scan with and without IV contrast administration 4 months prior to the MRI.



Further investigation of CT Urography three months after initial MRI detection demonstrated a new 58mm exophytic process undergoing heterogeneous enhancement after IV contrast administration accompanied by surrounding perirenal fat infiltration.

Following the CTU findings, one month later the patient was referred to our institute and scheduled for an US-guided biopsy which demonstrated the mass measuring at 50\*60\*70mm and the presence of a tumor thrombus occupying the renal vein.



Due to the US-guided biopsy results returning negative, after a multidisciplinary discussion, the patient had a CT Renal Mass Protocol performed three days later demonstrating the irregularly-shaped bordered mass measuring 60\*60\*80mm having no distinct separation from the renal vein, undergoing heterogeneous enhancement after IV contrast administration, intruding the renal pelvis, causing the involvement of the collecting system with lining thickening and enhancement of the renal pelvis, proximal, and middle ureter accompanied by perirenal and periureteral fat infiltration. Furthermore, a filling defect of the renal vein by the tumor thrombus had been present.







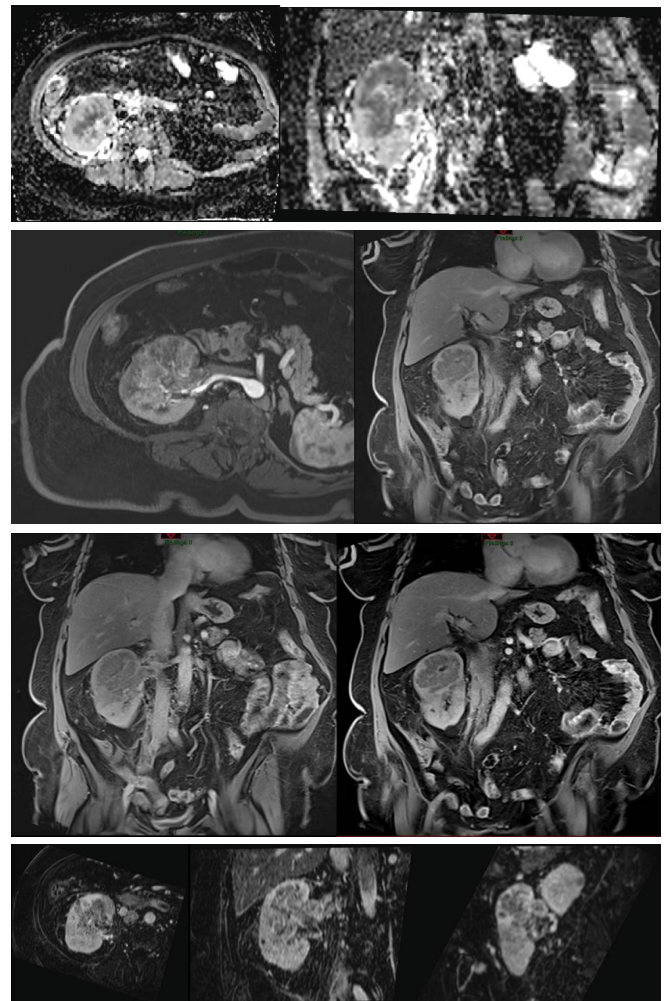
Several hours following the CT Renal Mass examination, a CT-guided biopsy was performed, with results confirming the diagnosis of papillary type 2 (grade 2) renal cell carcinoma.

#### RIGHT KIDNEY. TC BIOPSY:

Core of renal parenchyma infiltrated by RENAL CELL CARCINOMA, morphologically suggestive for papillary type 2 (grade2).

**Comment:** Immunohistochemical stains will follow to confirm the type of this carcinoma.

In order to try and perform a nephron-sparing nephrectomy of the only remaining kidney, another abdominal MRI+MRA were performed which illustrated the solid heterogenic space-occupying lesion measuring 76\*61\*70mm undergoing a clear DWI-restriction and also under immediate enhancement following contrast IV administration, fully involving the anterior-inferior and anterior-superior segments of the kidney and partially-involving the posterior segment, accompanied by collective system lining thickening with distinct nodular enhancement, and by the 4cm long tumor thrombus occupying the renal vein toward the entrance of the IVC.



Due to the recent MRI examination showing the involvement of collective system, the course of action decided by the multidisciplinary team was performing curative surgery in the form of a radical nephrectomy.

#### Discussion

The radiologist's point of view directly affects the patient's outcome at different checkpoints: whether it concerns the initial suspicion of a mass requiring further investigation, taking an active part as a member of the multidisciplinary team to assess the progression of the disease, and determining the proper clinical staging in order to administer the most suitable treatment option.

The probability of metachronous contralateral renal cancer is at overall risk of about 1.5% at 10 years[11,12]. This rare occasion of a patient having not just an aggressive metachronous renal malignancy less than a year after undergoing total nephrectomy, but also a relatively rare which is known to be more elusive in its radiologic detection, was detected in a radiologic examination performed due to a different etiology as an incidental finding.

This aggressively behaving tumor has been under close radiologic monitoring since the moment of its initial detection, demonstrating the rapid growth and local invasion to neighboring tissues, with 7 different examinations using all 3 main radiologic modalities to properly assess it, in hopes to be as accurate as possible at determining the clinical staging, in order to avoid unnecessary radical surgery resulting in the patient requiring renal replacement therapy for the rest of their life.

This case has also shown the challenges faced by the radiologist – the probability of the metachronous contralateral renal malignancy is relatively low, and the fact this relatively rare tumor has behaved so aggressively could have led to an under- estimation at determining the clinical staging that could have resulted in an uncurative surgery being performed and allowing the malignancy to spread.

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