



## Primary Renal Carcinoid Tumor: A Case Report

### Primer Renal Karsinoid Tümör: Olgu Sunumu

Renal Karsinoid Tümör / Renal Carcinoid Tumor

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#### Özet

Primer renal karsinoid tümör, nöroendokrin hücrelerden köken alan oldukça nadir görülen bir neoplazmdir. Bu kadar nadir görülmesi nedeniyle immünohistokimyasal karakteristiği net olarak ortaya konulamamıştır. Vakamızda;23 yaşında erkek hastanın rastlantısal olarak saptanan sağ renal kitesi mevcuttu, sağ radikal nefrektomi uygulandı. Postoperatif 22 aylık takiplerinde hastada metastaz veya nüks rastlanmadı.

#### Anahtar Kelimeler

Renal Kanseri; Nöroendokrin Tümör; Nefrektomi

#### Abstract

Primary renal carcinoid tumor is an extremely uncommon neoplasm that arises from neuroendocrine cells. As it is extremely uncommon, its immunohistochemical characteristics have not been clearly determined. In this case, a 23-year-old male patient was incidentally identified with right renal mass. Right radical nephrectomy was performed. Postoperative 22-month follow-up showed no metastasis or recurrence.

#### Keywords

Neuroendocrine Tumors; Renal Cancer; Nephrectomy

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## Introduction

Carcinoids are well differentiated neuroendocrine tumors that are common in the gastrointestinal system (74%), bronchial system and lung (25%). They also rarely (less than 1%) develop in the genitourinary system [1].

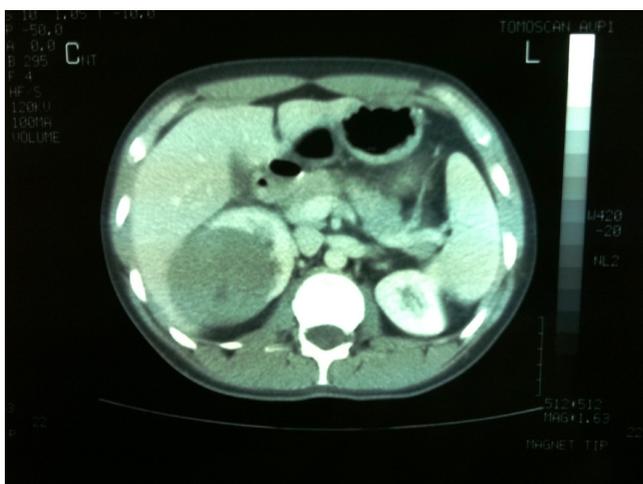
Primary renal carcinoid tumors are extremely uncommon. Less than 65 cases have been reported in the literature [2]. It is relatively more frequent in young adults than renal carcinoid tumors (median age 50). The prevalence in males and females is the same. It was reported that 55.8% of patients are admitted to hospital due to abdominal/flank pain and hematuria; only 13.6% of patients present with carcinoid syndrome [3]. While it is identified incidentally in 30% of patients, palpable mass is detected in 27%.

In some cases, substances such as chromogranin and serotonin can be identified in blood, or serotonin metabolite 5-Hydroxy-indoleacetic acid (5-HIAA) can be identified in 24-hour urine.

## Case Report

A 23-year-old male patient was admitted to general surgery clinic due to constipation. Patient history and physical examination showed no pathologic finding. Biochemical analyses showed urea: 18 mg/dL, creatinine: 0,9 mg/dL, aspartate aminotransferase (AST): 30 U/Lt, alanine aminotransferase (ALT): 35 U/Lt, haemoglobin (Hb): 16 g/ dL, Hematocrit: 47%, Platelet: 250000/dL. Posterior-anterior (PA) lung graphy was normal. Upper abdomen and pelvic spiral computed tomography was taken with and without contrast. Computer tomography (CT) showed a 9x7.5x7.5 centimetre (cm) mass lesion in the right kidney upper median posterolateral, which pushes the kidney towards the median and grew into posterior perirenal fatty tissue and infiltrated renal sinus fatty tissue. The mass contained a necrotic region and showed weaker contrast than the kidney (Images 1).

The patient underwent right radical nephrectomy on 11 May 2011. The patient was diagnosed with carcinoid tumor via pathologic evaluation. Postoperative somatostatin (octreotide) scintigraphy was normal. No tumor was found in esophago-gastroduodenoscopy or colonoscopy. Postoperative 22-month follow-up showed no local recurrence or metastasis.



Images 1. CT of the abdomen revealed a large malignant mass arising from the right kidney.

## Pathology

Grossly, the nephrectomy material was measured 13x11x8 cm. On section, well circumscribed encapsulated solid mass, localized on one pole, beneath the renal capsule and involving both cortex and medulla was detected. The mass measured 7cm in greatest dimension and the cut surface was diffusely haemorrhagic.

Microscopically the encapsulated tumor consisted of uniform cells arranged in trabecules, rows, glandular structures and exhibited ribbon like pattern (Fig 1-A). The tumor cells were of uniform, small to intermediate size with hyperchromatic, round nuclei and moderate amount of cytoplasm. The stroma was diffusely haemorrhagic (Fig 1- B), the cells were necrobiotic and dissociated in most areas and this feature made the evaluation of the mitotic rate difficult. However the Ki-67 index was lower than 1%. Although the tumor cells invaded its own capsule in some foci, they didn't invade the renal parenchyma. Perineural invasion was observed (Fig 1-C). Immunohistochemically the tumor showed diffuse positivity for vimentin, NSE (neuron specific enolaza) (Fig 1-D), synaptophysin ( Fig 1-E ) and CK18. Most of the cells were chromogranin positive (Fig 1-F) whereas a wide range of markers performed (CD10, RCC, EMA, panCK, HMWCK, CK7, CK20, CK8, CEA, inhibin, melanA, S100, TTF-1, WT1, CD99, CD56, SMA) were negative. The tumor was reported as neuroendocrine tumor consistent with carcinoid. It is also noted on the report that after a thorough search of all the organs and the systems and with the exclusion of another tumor, this tumor could be accepted as a primary renal carcinoid.

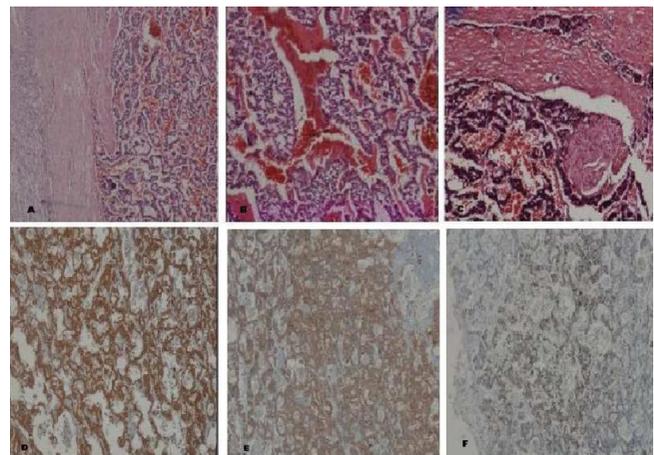


Figure 1. Microscopically tumor consisted of uniform cells arranged in trabecules, rows, glandular structures(A). The stroma was diffusely haemorrhagic (B), Perineural invasion was observed (C). Immunohistochemically the tumor showed diffuse positivity for vimentin, NSE (D), synaptophysin ( E ) and chromogranin positive (F).

## Discussion

Primary renal carcinoid tumor was first reported in 1966, by Resnick. Since then, due to the rarity of this disease, prognosis, clinical behavior and immunohistochemical characteristics have not been clearly determined [4].

Renal carcinoid tumor arises from neuroendocrine cells, as with other localizations of carcinoid tumor. Normal kidney tissue does not contain neuroendocrine cells.

Several hypotheses have been suggested in an attempt to explain how neuroendocrine cells, which play a role in pathogenesis of carcinoid tumor, reach the kidney. These hypotheses include congenital and acquired renal anomalies. Metaplasia in

pyelocaliceal urothelium due to chronic inflammation; metastasis from non-established primary tumor; trapped or wrong localization of pancreatic tissue or neural crest cells in the kidney during embryogenesis; activation of common gene sequences in multipotent primitive root cells with neuroendocrine programmed cells; and simultaneous congenital renal anomalies [5,6].

A review by Romero et al. found other accompanying renal pathologies in 26.8% of patients. Horseshoe kidney was identified in 17.8%, renal teratoma was identified in 14.3% and polycystic renal disease was identified in 1.8% of patients [7]. Relative risk for renal carcinoid in patients with horseshoe kidney was reported to be 62–82% [5].

These tumors are often identified during the fourth decade (range 13–68, mean 47). The prevalence in males and females is the same. Common symptoms are abdominal/flank pain, abdominal mass, loss of weight and hematuria.

Laboratory diagnosis involves identification of neuroendocrine substances in blood and urine. These include serum chromogranin, serotonin, glucagon, gastrin, somatostatin, calcitonin, pancreatic peptide, vasoactive intestinal peptide, adrenocorticotrophic hormone (ACTH) and 5HIAA in urine. However, there is currently no chemical test to endocrinologically identify whether silent renal masses are renal carcinoid.

Radiological findings observed in carcinoid tumors are common with renal cell carcinoma (RCC). Radiological imaging shows calcification in approximately 26.5% of renal carcinoid tumors. Lesion is monitored heterogeneously. CT shows minimal contrast involvement, while angiography shows hypo-vascular or avascular lesion. Recently, octreotide scintigraphy began to be used as sensitive imaging modality in diagnosis and staging of the tumor. Primary carcinoid tumor and metastases show high affinity to more than 85% of somatostatin receptors [8].

Metastasis rate of carcinoid tumors is directly related to the dimensions of the primary tumor. Primary renal carcinoid tumors show less aggressive biological character than RCC [6]. In general, metastases are present in 45.6% of cases at the time of diagnosis and metastasis develops in 59% of patients. The tumors that develop metastasis are larger than 4 cm. Generally, metastases occur in regional lymph nodes, liver and bones.

Primary treatment in primary renal carcinoid tumors involves surgical resection and regional lymph node dissection. Lymph node metastasis is present during surgery in 47% of patients. In 43-month postoperative follow-ups, although lymph node metastasis was present in pathology specimens, the absence of evidence for the disease was taken to indicate the possibility of surgical and curative treatment [7].

When renal carcinoid is detected, potential primary lesions should be analyzed via another focus [8]. Octreotide scintigraphy is the first and most important diagnostic test. Serum chromogranin and urine 5-HIAA identification is used in the absence of neuroendocrine symptoms.

It was reported that 50% metastasis can develop in postoperative 7th year. Therefore, long-term follow-up is necessary. Biomarkers should be examined at 3–6 month intervals; imaging should be conducted at 6–12 month intervals.

### Competing interests

The authors declare that they have no competing interests.

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