

Partial nephrectomy in horseshoe kidney: Primary carcinoid tumor

Kamil Gokhan Seker¹, Emre Sam¹, Selcuk Sahin¹, Mustafa Gürkan Yenice¹, Ayse Gul Aktas², Abdulmuttalip Simsek¹, Volkan Tugcu¹

¹ Department of Urology, Bakirkoy Dr.Sadi Konuk Training and Research Hospital, Istanbul, Turkey;

² Department of Pathology, Bakirkoy Dr.Sadi Konuk Training and Research Hospital, Istanbul, Turkey.

Summary *Primary neuroendocrine carcinoma of the kidney is a rarely observed clinical condition because neuroendocrine cells are not found in kidney parenchyma. It's not clinically and radiologically possible to distinguish from other kidney tumors. Incidence with horseshoe kidney anomaly, it should be considered as a definitive diagnosis for the patients with this condition. In this case report, we reported about a carcinoid tumor in horseshoe kidney in a 37-year-old woman.*

KEY WORDS: Carcinoid tumor; Neuroendocrine tumor; Horseshoe kidney; Partial nephrectomy.

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INTRODUCTION

Carcinoid tumors are low-grade tumors, exhibiting differentiation from neuroendocrine cells, with malignancy potential. Neuroendocrine cells in the body are also known as enterochromaffin cells or *amine precursor uptake and decarboxylation* cells (APUD) and they present in glandular endocrine organs (like hypophysis, parathyroid, adrenal medulla). *Neuroendocrine tumors* (NETs) are most commonly present at gastrointestinal system by 73.7% and respiratory system by 25.1% where they rarely present in ovary, testis, cervix, breast, biliary tract and gall bladder (1, 2). However, primary renal carcinoid tumor is a rare case and was firstly reported by Resnick in 1966 (1). In this case report, we aimed to focus on a case with well-differentiated primary neuroendocrine carcinoma of the kidney pathologically diagnosed after partial nephrectomy in horseshoe kidney.

CASE REPORT

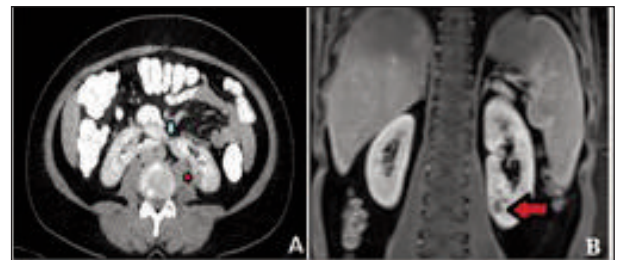
In a 37-year-old woman patient, examined due to incidentally diagnosed splenomegaly, a horseshoe kidney with a hypodense area 18 x 12.5 mm in size at left kidney lower pole was observed by abdominal *computed tomography* (CT) scans. Abdominal *magnetic resonance imaging* (MRI) confirmed a pathological area 18 x 12.5 mm in size at left kidney lower pole, in venous phase monitoring at T2 sequence with mild hypointense signal, with a mild suspicious contrast and exhibiting diffusion limitation in diffusional examination (Figure 1).

The patient was operated by left-open partial nephrecto-

Figure 1.

A. Horseshoe kidney formation and hypodense area 18 x 12.5 mm in size at left kidney lower pole (red, lower arrow). Isthmus in the patient with horseshoe kidney anomaly (blue, upper arrow).

B. Monitoring at T2 sequence with mild hypointense signal of the area exhibiting diffusion limitation.



my zero ischemia with modified Chevron incision (Figure 2). No complication was observed during intra-operative and postoperative periods. At macroscopic pathological examination of the specimen, a 2 cm in diameter and cream color mass was observed, at microscopic examination, negative surgical borders, mitotic index 1/50 *high-power fields* (HPF) and no necrosis were observed. From immunohistochemical tests, CDX2 (-), CD10 (-), vimentin (-), CK7 (-), epithelial membrane antigen (EMA) (-), chromogranin focal (+), synaptophysin: common (+) and 8% Ki-67 were observed. Microscopic and immunohistologic findings were in comply with primary neuroendocrine tumor of the kid-

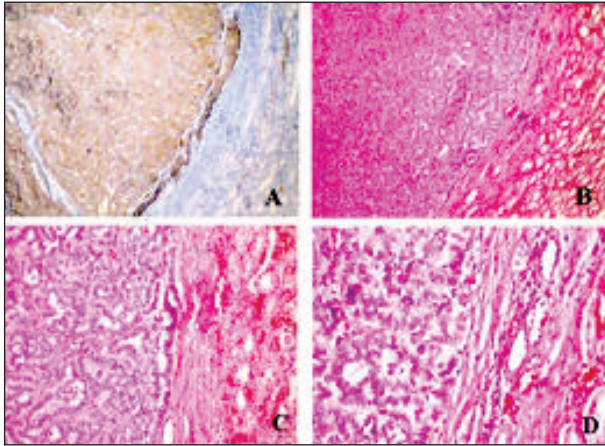
Figure 2.

Exophytic mass lesion. Borders were determined by cautery at left kidney lower pole.



Figure 3.

- A.** Synaptophysin positive tumor cells.
B. Association of normal kidney tissue with tumor HEx4.
C. Neuroendocrine tumor HEx20.
D. Neuroendocrine tumor HEx40.



ney diagnosis (well-differentiated carcinoid tumor with trabecular and solid pattern) (Figure 3).

No pathology was observed at gastrointestinal system endoscopy, thorax computed tomography and ^{68}Ga -DOTATATE positron emission tomography/computed tomography (PET/CT).

Neuroendocrine tumor markers were not found in urine and blood. The patient was followed by diagnosis of primary neuroendocrine carcinoma of the kidney and during 6 months follow-up period no local relapse or systemic metastasis was observed.

DISCUSSION

Primary neuroendocrine carcinomas of the kidney are rare and their pathogenesis is still unknown as these tumors do not usually present in adult normal renal parenchyma, renal pelvis and ureter (2). Unlike renal cell carcinoma, renal carcinoid tumor is observed at an early age and in most cases the age at diagnosis is about 49 years and no gender dominance is present. Symptoms (e.g. carcinoid syndrome) are rare and it is incidentally diagnosed in 25-30% of cases (2-4). They are clinically misdiagnosed with type 1 papillary renal cell carcinoma, mesonephric tumors, urothelial tumors, Wilms tumor and undifferentiated carcinoma (3).

Primary neuroendocrine carcinoma of the kidney do not have specific CT or MRI imaging. In general, primary renal neuroendocrine tumors show a mild contrast uptake as it's observed in radiological examinations of our case. Apart from this, it's characterized by cystic mass and sometimes by calcification at CT (5, 6).

The most dominant histological types of carcinoid tumors are trabecular or ribbon-like types. As a result of immunohistochemical studies, synaptophysin, chromogranin and CD56 showed positive and thyroid transcription factor (TTF-1), Wilms tumor protein (WT-1) and CDX2 showed negative (2, 7). Chromogranin, synaptophysin and CD56 positivities and CDX2 negativities of our case supported our neuroendocrine tumor diagnosis,

and thus, we discarded renal cell carcinoma diagnosis. It's known that primary neuroendocrine carcinomas of the kidney are frequently presented in kidneys with anomaly. So far in studies in the literature, 100 patients have been diagnosed by similar cases and it has been observed that tumor development mostly occurs in horseshoe kidney. The incidence of neuroendocrine carcinoma in horseshoe kidney is 62 times higher than in normal kidneys (8). In their report examining clinical characteristics of 21 renal carcinoid tumors, *Hansel et al.* reported horseshoe kidney formation in 19% of the patients (7).

Other kidney anomalies apart from horseshoe kidney are found as renal teratoma or teratoid malformation and polycystic kidney anomalies (1).

Our case of primary neuroendocrine tumor 2 cm in size, diagnosed in horseshoe kidney supports the literature. Primary neuroendocrine carcinomas of the kidney are known to stem from neuroendocrine cells and be low-grade malignancy potential tumors. To determine prognostic state of these tumors, some indicators and clinical findings are proposed. Aung et al. reported about morphologic and molecular structures of 11 well-differentiated renal carcinoid tumors showing that distant metastasis were not present in tumors 5 cm in size or ≤ 2 mitotic 10 HPF (3). In an other review, *Romero et al.* reported that 50% of cases was metastatic and of which 45.6% was at the time first diagnosis.

They also stated that the risk of metastasis increases especially in patients over 40 years of age and with solid characterized and high mitotic activity tumors (2). Complete surgical resection is the primary treatment against primary neuroendocrine tumors of the kidney. Partial nephrectomy might be applied depending on the size of the tumor (9).

Octreotide or ^{68}Ga -DOTATATE PET/CT is offered during follow-up as useful diagnostic tool in order to diagnose residual or hidden metastatic carcinoid tissue after surgical resection (10). Patients may develop local lymph node or distant organ metastasis after a long period of clinical course (7).

In our study, a local tumor 2 cm in size with low mitotic index was diagnosed and no distant metastasis was present. During follow-up after partial nephrectomy, no local or systemic relapse was observed.

CONCLUSION

Primary neuroendocrine carcinoma of the kidney are rarely observed and treated by curative treatment after complete surgical resection and show low-grade malignancy potential. Its presence in kidneys with anomalies should be kept in mind and, if there is not a clear definitive diagnosis of other kidney masses, it should be taken into consideration.

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Correspondence

Kamil Gokhan Seker, MD

Emre Sam, MD

Selcuk Sahin, MD

Mustafa Gürkan Yenice, MD

Volkan Tugcu, MD

Abdulluttalip Simsek, MD (Corresponding Author)

simsek76@yahoo.com

Bakirkoy Dr.Sadi Konuk Education and Research Hospital

Department of Urology

Tevfik Saglam Caddesi No:11 Zuhuratbaba/Bakirkoy

Istanbul 34147, Turkey

Ayşe Gul Aktas, MD

Bakirkoy Dr.Sadi Konuk Education and Research Hospital

Department of Pathology

Tevfik Saglam Caddesi No:11 Zuhuratbaba/Bakirkoy

Istanbul 34147, Turkey