Case Report
A Case of Primary Renal Carcinoid Tumor

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Primary renal carcinoid tumors are extremely rare kidney lesions, with fewer than 100 reported cases previously. We describe a 75-year-old man with an incidentally detected cystic renal mass. Computed tomography showed a 3 cm tumor with a cystic component enhanced with contrast. No evidence of metastasis was detected. We treated the patient with radical nephrectomy. Pathological examinations revealed a cellular arrangement specific to carcinoid tumor and positive for chromogranin A, neural cell adhesion molecule, and somatostatin receptor type 2. The tumor cells had a mitotic count of 4 mitoses/10 high-power fields, and the level of the proliferation marker Ki-67 was 5%. The pathological diagnosis was renal neuroendocrine tumor grade 2. No local recurrence and no systemic metastasis were detected during the 18-month follow-up period. To our knowledge, this is the 6th case of renal neuroendocrine grade 2 tumor reported thus far.

1. Introduction
Carcinoid tumors can arise in almost any organ, but they occur most commonly in the gastrointestinal and bronchopulmonary systems and less frequently in the hepatobiliary system [1, 2]. A primary renal carcinoid tumor is extremely rare, with only 97 cases reported in the medical literature [2–9]. We describe a 75-year-old man with a primary renal carcinoid tumor without any evidence of metastasis or carcinoid syndrome.

2. Case Presentation
A 75-year-old man was referred to our institution after being diagnosed incidentally with a 3.0 cm, solid, right renal mass on computed tomography (CT) performed to assess thoracic trauma. He had no specific symptoms. The results of laboratory investigations were within normal limits. On abdominal CT, a renal mass was identified in the upper portion of the right kidney (Figure 1). The mass measuring approximately 3.0 cm × 2.0 cm was well defined, and it consisted of a partial cystic mass within a mainly solid renal mass. The cystic component contained calcifications. No definite hydronephrosis or calicetasis was detected. The patient had no other abnormal findings, such as suspected distant metastasis or lymph node metastasis. At that time, we considered cystic renal cell carcinoma (RCC), oncocytoma, and adult-type cystic nephroma as the differential diagnosis of this mixed cystic and solid renal tumor. In May 2012, we planned an open partial right nephrectomy; however, during the operation, we noticed that the tumor had invaded the renal capsule. Therefore, we performed open right nephrectomy. After histopathological evaluation, the lesion was diagnosed as a primary renal carcinoid tumor (neuroendocrine tumor). The tumor met both the histological and immunohistochemical criteria for designation as a carcinoid tumor. Grossly, the resected tumor included a white-yellow soft mass attached to a cystic lesion (Figure 2). On microscopic examination, the tumor consisted of a ribbon-like, trabecular, or rosette-like pattern of cells with a high nucleus:cytoplasm ratio (Figure 3). Immunohistochemically, the tumor cells were positive for antibodies for the neuroendocrine markers, chromogranin A (Figure 4), neural cell adhesion molecule (NCAM) (Figure 5), and somatostatin receptor type 2 (SSTR2) (Figure 6). The tumor cells had a mitotic count of 4 mitoses/10 high-power fields and the level of proliferation marker Ki-67 was 5%.
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(a) Abdominal plain CT shows calcified mass in the right kidney

(b) Abdominal plain CT shows a heterogeneous mass (size: 3 × 2 cm)

(c) Abdominal contrast enhanced CT shows a solid lesion within a slightly enhanced mass in the right kidney

Figure 1: Abdominal computed tomography (CT).

The microscopic and immunohistological findings were compatible with the diagnosis of a primary renal neuroendocrine tumor (grade 2). No local recurrence or systemic metastasis was detected during the 18-month follow-up period.

3. Discussion

Carcinoid tumors are low-grade malignant neoplasms with neuroendocrine differentiation. They are located mainly in the gastrointestinal (74%) and respiratory (25%) tracts [1]. In <1% of all cases, carcinoid tumors are reported in the testes, ovaries, kidneys, and the prostate [2]. A primary renal carcinoid tumor is extremely rare, with only 97 cases reported in the medical literature. Primary renal carcinoid tumors are equally prevalent in both genders [3]. Such tumors usually

Figure 2: Macroscopic findings. The gross features of the resected tumor included a white-yellow soft mass attached to a cystic lesion.

Figure 3: Microscopic findings. Tumor cells are arranged in cords and with a ribbon-like appearance (hematoxylin and eosin, magnification: ×400).

Figure 4: Chromogranin A-stained section. Tumor cells are positive for chromogranin A immunostain (magnification: ×200).

Figure 5: Neural cell adhesion molecule-stained section. Tumor cells are positive for neural cell adhesion molecule immunostain (magnification: ×200).
The World Health Organization (WHO) has proposed a classification system of carcinoid tumors located elsewhere. The World Health affinity receptors for somatostatin in 87% of cases [7].

Recent several treatments for carcinoid tumors have been reported, such as somatostatin analogues, everolimus, and radionuclide therapy [20–22]. Adjuvant therapy is not currently indicated in patients with completely resected NETs [23]. But the results of a placebo-controlled randomized double blind prospective phase IIIB study on the effect of octreotide on tumour growth in patients with metastatic midgut NETS (PROMID Study) showed that octreotide significantly lengthens time to tumour progression (14.3 months in octreotide group versus 6 months in the placebo group) in patients with functionally active and inactive NETs [20]. Conventional methods of imaging are inadequate for detecting smaller carcinoids, so octreotide scintigraphy or 68Ga-labeled PET-CT should complement computed tomography and magnetic resonance imaging when searching for occult or metastatic disease postoperatively. And we should consider whether to adjuvant therapy based on the nuclear examination.

The prognosis of renal carcinoid tumors is not predictable because of their rarity. Omiyale and Venyo found metastasis in 55% of the 29 patients under study and reported that 73.1% of the 29 patients did not present evidence of disease after treatment [3]. Although prognosis of renal carcinoid tumors is better than that for other renal neoplasms, renal NETs grade 2 are associated with poor prognosis. All of the five renal NET grade 2 cases previously reported had metastasis and 2 of 5 (40%) patients died because of the disease [8, 16–18]. In our case, no local recurrence and no systemic metastasis were detected in the 18-month follow-up period. However, we did not perform any nuclear medicine images for searching occult or metastatic disease. A nuclear medicine image and strict follow-up after surgery were needed because of the poor prognosis associated with renal NET grade 2 tumors.
Consent

Written informed consent was obtained from the patient for the publication of this case report and the use of the accompanying images.

Conflict of Interests

The authors declare no conflict of interests regarding the publication of this paper.

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