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Oncology

Case report: A case of primary renal carcinoid tumor

Chung-Tso Chen^{a,1}, Shu-Wei Hsieh^b, Teh-Sheng Hsieh^{a,*}

- ^a Division of Urology, Department of Surgery, Cathay General Hospital, Taipei, Taiwan ROC
- ^b Department of Anatomic Pathology, Cathay General Hospital, Taipei, Taiwan ROC



Introduction

Carcinoid tumors are low-grade malignant tumors that arise from neuroendocrine cells. There are mostly found in the intestinal tract. Primary renal carcinoid tumors are extremely rare, with fewer than 100 reported cases since the first description in 1966. We describe a 37-year-old female with a primary renal carcinoid tumor without any evidence of metastasis.

Case presentation

This 37-years-old female patient had past history of hypertension and obesity (103 kgs decreased to 73 kgs in 2 years) but denied any other systemic diseases. According to the patient, she has had hypertension for 5-6 years. Although with regular medical control, her blood pressure was still around 130-140 mmHg. Under the suspicion of secondary hypertension, kidney echo was arranged on 2015/05/26 and showed a 2.5cm right renal tumor (Fig. 1a). She had no fever, hematuria, flank pain nor loss of body weight. Urine analysis revealed no positive finding. Abdominal CT showed a nearly 3 cm mild contrast enhancing mass lesion in right renal sinus (Fig. 1). CT guided biopsy reported an epithelial neoplasm with neuroendocrine features. After discussion with the patient, right radical nephrectomy was performed on 2015/8/13. Pathology reported a primary carcinoid tumor about $2.4 \times 2.3 \times 2.1$ cm in size (Fig. 1b) and consisted of trabecular or ribbon-like arrangement of monotonous tumor cells with oval nuclear and salt-and-pepper chromatin (Fig. 2). Immunohistologically, the tumor cells were positive for Synaptophysin and CD56, the level of proliferation marker Ki-67 was < 2% (Fig. 3). She recovered smoothly and was discharged on 8th post-operation day. There was no evidence of metastasis and the hypertension was under better control during the 10 months follow-up period.

Discussion

Carcinoid tumors are low-grade malignant neoplasms with neuroendocrine differentiation. They are located mainly in the gastrointestinal (74%) and respiratory (25%) tracts. The primary carcinoid tumor originated from kidney is very rare. There are less than 100 cases after the first case was reported by Resnick et al., at 1966. The mean age of previous report of primary renal carcinoid tumor was 48 years, ranged 29–75 years old. The peak ages of incidence are between 50 and 60 years old. The occurrence rate of primary renal carcinoid tumor was no difference in both genders. According to previous study, renal carcinoids were associated with other renal abnormalities, like horseshoe kidneys (17.8%), teratomas (14.3%), and polycystic kidney disease (1.8%).

There was about 25–30% of primary carcinoid tumor of the kidney was incidentally found with no clinical symptoms.² Presenting symptoms in the remainder include abdominal or flank pain, hematuria, constipation, urinary frequency, fever, weight loss and testicular pain. The carcinoid syndrome occurs in approximately 12.7% of patients with renal carcinoids in previous study reported. The clinical manifestations of carcinoid syndrome such as flushing or diarrhea was caused by vasoactive substances (serotonin) produced from the tumors which enter the systemic circulation.² Serotonin is the principal endocrine product of carcinoid tumors, but simultaneously increased production of catecholamines from these tumors have been reported in some cases. Therefore, poor blood pressure control in this case, maybe caused by the carcinoid tumor.

Conventional methods of imaging such as computed tomography or magnetic resonance imaging are inadequate for detecting smaller carcinoids, because of occasionally lack of sensitivity in detecting these tumors. In recent case report, somatostatin receptor scintigraphy can be helpful in preoperative and postoperative follow-up assessment of disease status and is helpful to early detect metastasis. Carcinoid tumors abundantly express somatostatin receptor (SR), and radiolabeled octreotide, a somatostatin analogue that has a high affinity for SR, is useful in detecting even the smallest carcinoid tumors. SR scintigraphy has a reported sensitivity of 85% in recent studies.

The most common histological pattern is a trabecular arrangement of tumor cells. Tumor cells may also form solid nests with peripheral palisading as well as gland-like or ribbon-like structures. In immunohistochemical staining, particularly strong and diffuse staining

^{**} Corresponding author. Division of Urology, Department of Surgery, Cathay General Hospital, 280 Renai Rd. Sec.4, Taipei, Taiwan ROC. *E-mail addresses*: gn01118415@hotmail.com (C.-T. Chen), tshuro.tw@yahoo.com.tw (T.-S. Hsieh).

¹ First author: Division of Urology, Department of Surgery, Cathay General Hospital, 280 Renai Rd. Sec.4, Taipei, Taiwan.

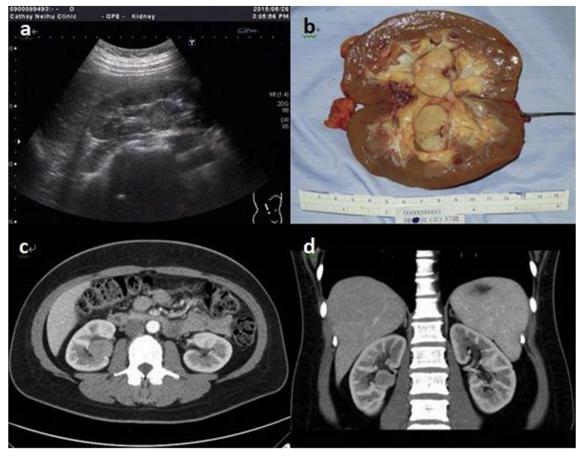


Fig. 1. (A) a heterogeneous mass (2.5cm) was noted in the right kidney. (b) Fig. 3: One brown to yellow, solid and well-defined tumor ($2.4 \times 2.3 \times 2.1$ cm) over middle portion, of right renal sinus. (c) (d)3 cm mild contrast enhancing mass lesion in right renal sinus. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

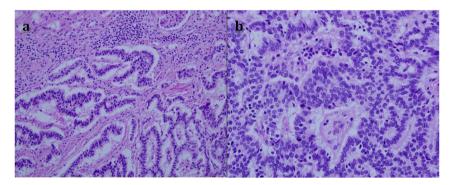


Fig. 2. Microscopic findings. (a) ribbon-like arrangement of tumor cells. Upper portion of this picture show normal renal parenchyma and glomerulus. (b) Oval nuclear and salt-and-pepper chromatin in HPF.

with synaptophysin and chromogranin could supports the diagnosis.⁵ In our case, the tumor consisted of trabecular or ribbon-like arrangement of monotonous tumor cells with the positive Immunohistologically staining of Synaptophysin and CD56. The pathologic finding is compatible with the previous study.

Surgical treatment which may be radical or partial nephrectomy with lymph node dissection has been reported as the first choice of treatment in the management of localized primary renal carcinoid. In previous review studies, 73.1% of patients without evidence of disease

after surgical treatment in 20 months average follow-up time had been reported. $^{\!1}$

Conclusion

Primary renal carcinoid tumors are rare tumors with an indolent course. SR scintigraphy can be helpful in assessment of renal carcinoid tumor. Surgical treatment was the curative treatment that provide good prognosis.

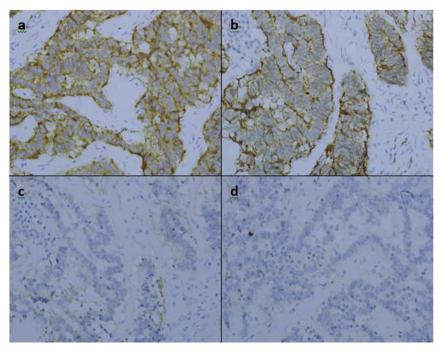


Fig. 3. (A) Synaptophysin: diffusely positive. (b) CD56: diffusely positive. (c) Chromogranin A: negative. (d) the level of proliferation marker Ki-67 was < 2%.

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