Primary Renal Carcinoid Tumor

CHI-LUN WENG1  CHI-JEN CHEN2  SEN-PING LIN1  SHIH-HUNG HUANG3  YUNG-CHEN WANG1

Department of Radiology1, Department of Pathology3, Cathy General Hospital, Taipei, Taiwan
Department of Radiology2, Taipei Medical University-Shuang Ho Hospital, Taipei, Taiwan

ABSTRACT

Primary renal carcinoid tumor (PRCT) is extremely rare since normally no enterochromaffin cells will be found in renal parenchyma. The clinical manifestations of PRCT are often non-specific, sometimes even without symptoms. There were about 90 cases reported in the literatures to our knowledge. We reported a PRCT in a 67-year-old woman, which was incidentally found in the health examination. The literature was reviewed. We suggest that PRCT should be considered when a renal tumor coexists with horseshoe kidney or a renal mass shows hypovascularity on the angiography.

CASE REPORT

The 67-year-old woman received a health examination on May 20, 2009. Abdominal sonography showed some hepatic cysts and a tumor in the left kidney. She had a history of hypertension under regular medication and denied recent body weight loss. During admissional work-up, only mild microcytic anemia (Hb=11.6 g/dl, MCV=75 fL) was noted. An abdominal CT examination revealed hepatic cysts and a left renal tumor (Fig. 1). There was a well-circumscribed mass lesion of soft tissue density with punctate calcification in the left kidney on the axial view of non-contrast CT. It shows mild homogenous enhancement after injection of contrast medium and the left pelvicalyceal system was displaced by the tumor mass without hydronephrosis. She received left radical nephrectomy 2 days later. The operative findings revealed a tumor, measured 5.9 × 5.7 × 3.8 cm in size, confined to renal parenchyma without blood vessel and hilar lymph node extension. The renal pelvis and calyces were free of tumor except focally compressed. The histopathological study of the specimen showed uniformly polyhedral cells arranged in characteristic cord-like or thin trabecular patterns. The nuclei of the tumor cells were round to oval shape with neuroendocrine-like chromatin features and scanty to moderate amounts of eosinophilic cytoplasm. The neuroendocrine cell markers, including chromogranin A, synaptophysin and CD56, were all positive in these tumor cells (Fig. 2). All of above features were compatible with the diagnosis of a PRCT. The post-operative course was smooth. No tumor recurrence was observed in her follow-up CT one year later.
Carcinoid tumors are neuroendocrine origin and often contain neurosecretory granules with a variety of biogenic amines and/or hormones [2], which are arising from enterochromaffin cells or cells with amine precursor uptake and decarboxylation. The most common locations are the primitive gut derivatives, such as gastrointestinal tract (74%) and respiratory tract (25%) [3], whereas otolaryngeal, breast and genitourinary carcinoids make up the remaining 1%-2% [4].

Primary carcinoid tumor in the kidney is extremely rare because enterochromaffin cells are absent in normal renal parenchyma. Several pathogenetic theories of PRCT have been proposed, including intestinal metaplasia of the pyelocalyceal urothelium due to a sequela of chronic inflammation [5, 6], metastases from undiscovered primary origins, or entrapped neural crest, pancreatic cells and primitive stem cell differentiation [7]. PRCT are often found in horseshoe kidneys, possibly because of the existence of aberrant epithelium or teratomatous elements in the horseshoe kidney [5, 8-9].

The initial symptoms of the majority of patients were abdominal or flank pain. Other symptoms included palpable abdominal mass, hematuria, and certain constitutional symptoms such as body weight loss, fatigue, malaise, fever and carcinoid syndrome. The tumor could also be incidentally found in minority of patient without any symptoms, like our case [10].

The radiological features of PRCT are not well characterized. McKeown et al. reported 10 patients of PRCT in the literature. Five of them were predominantly solid and one was a complex cystic tumor with soft polypoid projection.
from the cystic wall. The remaining four cases, three arose from the teratoid malformation of kidney and one without radiologic pictures. Either central or peripheral calcification in the PRCT was noted in these cases [11]. Shurtleff et al., after analyzing 43 cases, described that 74% of PRCT were solid tumor with occasionally cystic component (49%). 26% of PRCT had obvious necrosis [10]. Kurl et al. reviewed 16 patients, including 8 patients with renal angiography or aortography. One out of 8 was an avascular mass. Three revealed poor vascularity and four showed hypovascularity [12]. The CT findings of PRCT are variable and non-specific in comparison with those of renal cell carcinoma. However, they always showed avascular or hypovascular feature in the angiography, which is contrary to the common hypervascular nature of renal cell carcinomas [13, 14]. PRCT rarely involved the vascular structure such as inferior vena cava or renal vein except two case reports of the inferior vena cava and right renal vein invasion [15, 16]. Our PRCT case showed some punctate calcification, mild enhancement and no adjacent structural invasion. Urothelial carcinoma is less likely but can not be excluded completely because the renal mass only showed mild enhancement. These radiologic findings were too non-specific to differentiate carcinoid tumor from other renal mass. The final diagnosis was confirmed histologically by positive reactions to neuron-specific enolase, synaptophysin, and chromogranin. Somatostatin receptor scintigraphy (OctreoScan) is a good diagnostic and staging tool. Sensitivity of this method in detecting carcinoid tumors had been reported to be greater than 85% [17, 18].

All patients of PRCT reported in English literatures underwent partial or radical nephrectomy. The prognosis of PRCT is relatively good. In a review literature of 43 cases, 10 patients (23%) developed metastasis, and the most common sites of metastases were the lymph nodes and liver. Overall, only 4 deaths were reported with a mean time follow-up of 27.6 months (range 3-48 months) [10]. The response rate of PRCT to chemotherapy is poor and toxic. The best treatment strategy for PRCT is still undetermined [19].

In summary, we concluded the PRCT is a rare renal tumor and cannot be differentiated from the renal cell carcinoma exclusively by CT. However, they generally demonstrate hypovascularity on the angiography. We suggest PRCT should be considered when a renal tumor coexists with horseshoe kidney or a renal mass shows hypovascularity on the angiography.
REFERENCE