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Primary renal carcinoid tumor: A radiologic review

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Carcinoid tumor is the classic famous anonym of neuroendocrine neoplasms. Primary renal carcinoid tumors are extremely rare, first described by Resnick and colleagues in 1966, with fewer than a total of 100 cases reported in the literature. Thus, given the paucity of cases, the clinical and histological behavior is not well understood, impairing the ability to predict prognosis. Computed tomography and (occasionally) octreotide studies are used in the diagnosis and followup of these rare entites. A review of 85 cases in the literature shows that no distinctive imaging features differentiate them from other primary renal masses. The lesions tend to demonstrate a hypodense appearance and do not usually enhance in the arterial phases, but can occasionally calcify. Octreotide scans do not seem to help in the diagnosis; however, they are more commonly used in the postoperative followup. In addition, we report a new case of primary renal carcinoid in a horseshoe kidney.

Case report

40-year-old male initially presented to a community hospital with a 20-lb weight loss over a few months. In retrospect, the patient recalled mild left-flank discomfort and fatigue, but denied any hematuria. Blood work revealed an elevated serum glucose, and he was diagnosed with type 2 diabetes. Further workup included ultrasound, which revealed a tumor in his retroperitoneum abutting a left moiety of a horseshoe kidney.

The patient's past medical history was significant for recently diagnosed type 2 diabetes and a knee ligamentous injury at the age of 14. Medications included metformin, diamicron, ventolin, and symbicort. His family history consisted of a maternal grandfather requiring a nephrectomy; the patient was unsure of the cause. There was no known family history of renal-cell carcinoma.

Physical examination was unremarkable, with no flank or abdominal pain. All biochemical and hematological workup was normal, including CBC, LFTs, creatinine, and calcium.

Imaging findings

CT of the abdomen and pelvis, done in the portal venous phase, demonstrated a solid, hypodense, 4.5-cm renal mass containing calcifications, located in the posterior aspect of the medial portion of the left renal moiety of the horseshoe kidney (Fig. 1). The mass did not enhance strongly in the venous phase, and there was no apparent metastatic disease. Subsequent MRI attempted to further characterize the renal mass; it revealed an enhancing left renal upper pole mass measuring approximately 4.1 x 3.8 cm, which demon-

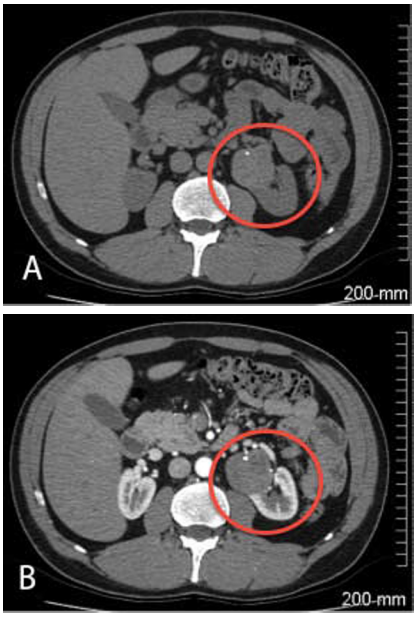

Figure 1. Computed tomography imaging of axial slices, unenhanced (A) and enhanced (B), showing left renal carcinoid (circled in red) in horseshoe kidney.

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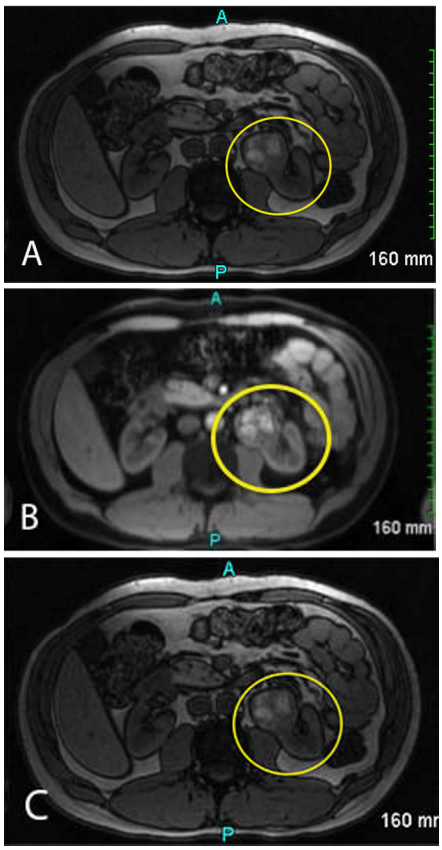

Figure 2. MRI axial T1 Dixon VIBE out of phase (A), T1 VIBE gadolinium-enhanced fat-saturated (B), and T1 FLASH gadolinium-enhanced fat-saturated sequences (C) showing left renal mass (circled in yellow) in a horsehoe kidney.

strated a heterogeneous signal intensity on both T1 and T2-weighted images with areas of bright signal on T1-weighted images, likely due to intralesional hemorrhage (Fig. 2). Although the tumor abutted the psoas muscle, it did not appear to invade it.

Management

A pre-operative chest radiograph was negative for metastatic disease. The patient was taken to the operating room and underwent an uncomplicated partial left nephrectomy. Intra-operatively, it was obvious that the mass did not invade the psoas muscle. Pathology revealed a well-differentiated neuroendocrine tumor, its histology compatible with carcinoid. Numerous immunostains were performed. The neoplastic cells were negative for inhibin, TTF-1, keratin 903, carbonic anhydrase IX, PAX-2, RCC antigen, S-100, CD10, CK7, and CK20. The tumor cells were strongly and diffusely positive for neuroendocrine markers, including synaptophysin, chromogranin A, and CD56. The cells were also positive for vimentin and weakly for racemase (AMACR). The proliferation index, as demonstrated by Ki-67 staining, was low, at 1-2%.

Followup

Postoperative CT thorax, octreotide study, endoscopy, and colonoscopy were all negative, with no source of gastrointestinal tumor identified. The patient is now approximately two years post surgery, with no evidence of local recurrence or metastatic disease.

Discussion

Etiology and demographics

Carcinoid tumor is the classic famous anonym of neuroendocrine neoplasms, first described in 1888 by Lubarsch (1). It gained its name from the earlier description of benign behavior of the lesion despite its malignant appearance under microscopy (2). It arises from a wide variety of tissues and organs, most commonly from specialized endocrine cells in the gastrointestinal and respiratory tracts, with prevalence values of 66.9% and 24.5%, respectively (3). The tumors produce hormones and protein products associated with specific clinical symptoms, and their malignant potential varies by location and cell type. Primary renal carcinoid tumors are extremely rare; they were first described by Resnick and colleagues in 1966 (4), with fewer than a total of 100 cases reported in the literature (5, 6). Thus, given the paucity of cases, the clinical and histological behavior is not well understood, impairing the ability to predict prognosis.

Carcinoid tumors arise from neuroendocrine cells and are believed to originate from enterochromafin or amine precursors and decarboxylation cells with malignant potential; however, neuroendocrine cells are not identified in the kidney or renal pelvis (7-10). Although renal carcinoid tumors exhibit morphologic and immunohistochemical features consistent with a hindgut neuroendocrine phenotype, the precise pathogenesis is controversial (11). Several hypotheses have been proposed, on the basis that these tumors arise from interspersed neuroendocrine cells associated with acquired and/or congenital abnormalities. The first hypothesis suggests that chronic inflammation induces metaplasia of the pyelocalciceal urothelium (12-14). The second suggests that they are metastases from an unknown primary (15). The third is that the neural crest or pancreatic cells have been misplaced or abnormally migrated during embryogenesis (14, 16). The fourth suggests concurrent congenital renal abnormalities (17, 18). And the last hypothesis suggests activation of gene sequences in multipotent primitive stem cells (5, 19, 20-28).

Methods

We performed an extensive literature search for all reported renal primary carcinoid tumors. In total, we reviewed 85 cases, with one unpublished case from our institution. We evaluated demographical, clinical, histopathological, and prognostic data, with a focus on radiologic findings.

Demographics

Of the 85 cases, 50 were males (59%) and 35 females (41%), with ages ranging from 12 to 75. The average age of diagnosis was 47.7 years, and the median age was 49 years. Forty-eight tumors were right-sided, and 37 were left-sided. Of the 85 cases, 50 were between 0-4 cm, 34 cases were more than 4 cm, and one size was not specified.

Several cases in the literature have reported coexisting renal anomalies such as horseshoe kidney and teratomas, suggesting that this disease may perhaps be more common