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Primary Renal Well-differentiated Neuroendocrine Tumor Extending Into the Inferior Vena Cava

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Abstract

Primary renal neuroendocrine tumors (NETs) are exceedingly rare with roughly 100 reported cases. To the best of our knowledge, this is the first reported case of a primary renal NET with inferior vena caval tumor thrombus. The uniqueness of this case is highlighted by the degree to which vascular invasion occurred.

A 57-year-old male presented with acute right flank pain radiating to the groin and an episode of gross hematuria. Physical examination demonstrated a palpable, nontender right flank mass. CT scan revealed a right renal mass measuring $14.4 \times 13.5 \times 8.2$ cm with tumor thrombus extension into the inferior vena cava (Fig. 1). There were no signs of contralateral or metastatic disease. An uncomplicated right open radical nephrectomy with tumor thrombectomy was performed. Gross examination of the renal mass revealed a tan-brown, hemorrhagic, and nodular mass involving renal sinus and extending into renal vein (Fig. 2). Immunohistochemical analysis revealed tumor cells to be negative for PAX8 and CK7, while positive for synaptophysin and chromogranin (Fig. 3). All 30 resected lymph nodes were benign. The final diagnosis was pT3bN0 well-differentiated NET.

Lamb et al radiographically reviewed 85 samples of primary renal NETs and found no distinctive imaging features to differentiate them from other renal masses.¹ In the absence of a high degree of suspicion for NET based on clinical presentation or radiographic findings, the contemporary preoperative and operative management of primary renal NET remains similar to that of typical renal masses.

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Reference

1. Lamb L, Shaban W. Primary renal carcinoid tumor: a radiologic review. Radiol Case Rep. 2014;9:923. [PubMed: 27186242]

Figure 1.

(A) Axial abdominal CT with contrast. Yellow arrow—Abnormal inferior location, configuration and axis deviation of kidney with overriding IMA consistent with HSK. Red arrow—IVC compression from renal tumor. (B) Axial CT with contrast. Red arrow – Renal tumor thrombus extending into IVC. Yellow arrow—Dilated renal pelvis. (C) Coronal CT. Red arrow—Tumor thrombus invasion of IVC with compression inferiorly. Yellow arrow—Collecting system. Renal malrotation of left kidney from horseshoe kidney configuration.

Figure 2.
Gross examination of renal specimen measuring $14.4 \times 13.5 \times 8.2$ cm.

Figure 3.
Histologic and immunohistochemical features. **(A)** H&E 20×. **(B)** PAX8 immunostain. **(C)** Synaptophysin immunostatin.