

Invasive neuroendocrine tumor of the kidney: a case report

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Abstract

Neuroendocrine tumors (NETs) involve the genitourinary system in less than 1% of cases, with primary renal carcinoids comprising only 19% of reported genitourinary NETs (56 cases worldwide). We report a case of a renal NET presenting as a large renal mass with extensive local invasion, requiring definitive radical *en bloc* resection via a thoracoabdominal approach.

Introduction

The neuroendocrine cell system is divided into gland-forming cells, such as the pituitary, parathyroids, adrenal medulla etc., and disseminated neuroendocrine cells, such as those found in the lung, gastrointestinal tract and genitourinary tract among other organs. Tumors arising from the disseminated neuroendocrine system within the genitourinary tract are referred to as carcinoids by the WHO classification.¹ In a recent comprehensive review of the world's literature, Romero *et al.* reported on 56 cases of primary renal carcinoids, extirpated by partial or radical nephrectomy.² To our knowledge, we report the first case of a renal NET exhibiting wide local invasion (diaphragm, pancreas, spleen and left colon mesentery), requiring *en bloc* radical resection through a thoracoabdominal approach.

Case Report

A 61-year old woman, an immigrant from Panama, presented in her home country with vague abdominal pain. She was diagnosed with diverticulitis *in lieu* of imaging, and was treated conservatively. Six months subsequently, she had progressively worsened abdominal pain as well as a 20lb weight loss. An abdominal ultrasound and follow-up CT scan revealed a large left renal mass which was deemed unresectable. She traveled to the

United States for a second opinion.

On presentation, she was an otherwise healthy thin woman with a palpable minimally tender mass in her left flank. A CT scan of the abdomen and pelvis (Figure 1) showed a 16 cm necrotic renal mass invading into the tail of the pancreas, spleen, and omentum, with marked displacement of intraabdominal organs. No metastatic lesions were identified. Clinical staging by chest CT and bone scan was negative for metastases. A magnetic resonance imaging (MRI) of the abdomen showed no evidence of renal vein or IVC thrombus.

She underwent uncomplicated *en bloc* resection, through a thoracoabdominal approach, of the left kidney, left adrenal, cuff of diaphragm, spleen, tail of pancreas and part of the left colon, with a primary colocolostomy (Figure 2). The diaphragm was closed primarily and a tube thoracostomy placed. Her post-operative recovery was unremarkable, with discontinuation of the chest tube and advancement to a regular diet by post-operative days 3 and 6, respectively. Pathological diagnosis was a low-grade neuroendocrine tumor of unknown primary, but with total replacement of the left kidney and involvement of all resected organs except for the adrenal gland and the wall of the left colon (but the colon mesentery was involved). Resection margins were negative. There was histological variation, with a dominant multinodular pattern composed of nests and trabeculae (Figure 3A and B) with focal spindled morphology (Figure 3C). Tumor cells were small, with small nuclei and moderate cytoplasm. Immunohistochemistry was positive for keratin, chromogranin (Figure 3D and E), neuron-specific enolase, and synaptophysin (consistent with neuroendocrine carcinoma), and negative for markers of fibroblasts and smooth muscle. An octreotide scan one month post-operatively was limited, but was negative for evidence of residual or recurrent disease.

Discussion

Disseminated neuroendocrine cells are also known as enterochromaffin cells or amine precursor uptake and decarboxylation (APUD) cells. They are rare in the genitourinary tract, but have been identified in the bladder trigone, prostate, and the renal collecting system. They are absent in the renal parenchyma.^{3,4} Tumors arising from these cells are extraordinarily rare, particularly within the kidney. Indeed, to date, only 56 reported cases of primary renal carcinoid tumors have been described. Pathogenesis is controversial,⁵ and there is a common association between the presence of these tumors and renal anomalies, particularly horseshoe kidneys.⁶

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Previous case reports have described resection of renal carcinoids up to 30 cm in size by partial or radical nephrectomy.⁷⁻¹⁰ However,

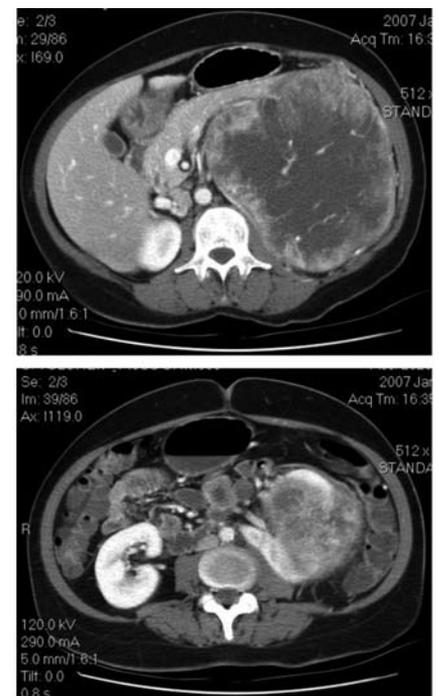


Figure 1. CT scan of the abdomen and pelvis.

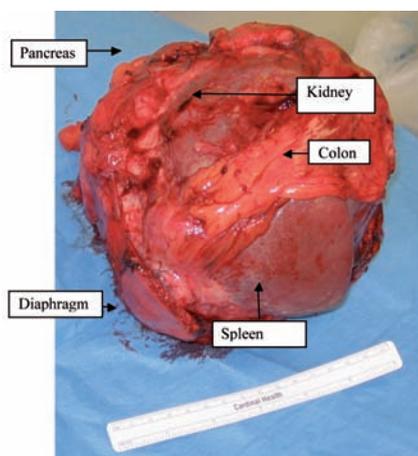


Figure 2. Surgical specimen.

although liver and lymph node metastases were commonly observed with larger tumors, extensive local invasion in the absence of metastatic disease, as described in the above case, has not been previously reported. Within the gastrointestinal tract, invasive behavior of carcinoid tumors is rare, and is associated with poorly differentiated or sporadic tumors, with angioinvasion and metastatic spread at time of diagnosis commonly identified.¹¹ As demonstrated in the case reported, extensive local invasion in the absence of metastatic spread does not preclude complete surgical resection. Evidence of complete extirpation was negative by octreotide scintigraphy one month post-operatively. Octreotide scintigraphy has been described as a useful adjunctive diagnostic tool in detecting residual or occult metastatic carcinoid tissue following surgical resection.⁹ However, long-term follow-up is required as metastatic disease up to seven years following treatment has been reported.

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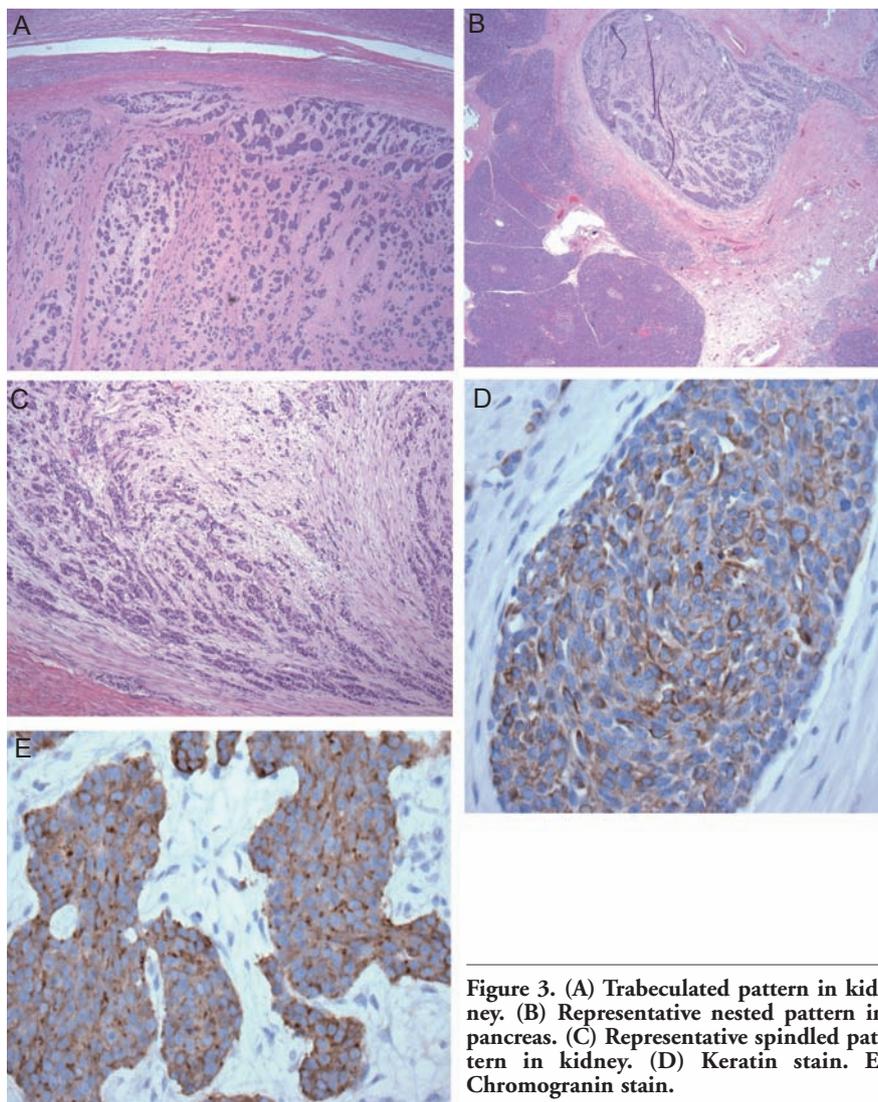


Figure 3. (A) Trabeculated pattern in kidney. (B) Representative nested pattern in pancreas. (C) Representative spindled pattern in kidney. (D) Keratin stain. E. Chromogranin stain.

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