Horseshoe kidney and primary renal carcinoid tumour: a case report of a rare entity

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Abstract

Carcinoid tumours are unique neuroendocrine neoplasms, displaying low-grade malignant potential and arising frequently within the gastrointestinal tract and respiratory tree. Primary carcinoid arising within the renal system is uncommon, whilst carcinoid tumour arising within a horseshoe kidney is rare. We describe a case of primary renal carcinoid occurring within a horseshoe kidney.

Keywords

Carcinoid; renal; kidney; neoplasm.

Case report

A 51-year-old man was found to have a mass in a previously unknown horseshoe kidney whilst undergoing computed tomography (CT) lumbar spine investigation for lumbar back pain and sciatica. History and examination were unremarkable for carcinoid syndrome or urinary tract symptoms. Full blood analysis, biochemistry, and urinalysis were within normal parameters. Staging CT chest, abdomen and pelvis revealed a horseshoe kidney containing a solid, heterogeneous, enhancing, $60 \times 42 \times 35$ mm mass involving the right lower pole and extending up to the site of lower pole fusion, with areas of focal calcification. There was no evidence of metastatic spread (Fig. 1). An uncomplicated tumour resection occurred through a midline laparotomy with partial nephrectomy and resection of the horseshoe isthmus. A $40 \times 50 \times 35$ mm specimen was resected, containing a well-circumscribed pale tumour, measuring $30 \times 25 \times 20$ mm (Fig. 2). The patient recovered well and 1 year postoperatively an octreotide study with single photon emission-computed tomography/CT images demonstrated no evidence of metastatic tumour.

Diagnosis

Histopathology demonstrated a solid mass composed of tightly packed trabeculae, in a background of minimal sclerotic stroma. The tumour cells showed uniform round to ovoid...
nuclei with characteristic fine stippled chromatin. These cells appeared in parallel to each other forming trabecula and ribbons. There was no evidence of tumour necrosis and no increase in mitotic activity (Fig. 3).

Immunohistochemical staining was positive for neuroendocrine markers including chromogranian, synaptophysin and neuron-specific enolase, confirming the diagnosis of carcinoid. The cells stained negative for TTF-1, WT-1, and CK-20, ruling out lung and bowel primary sources (Fig. 4).

Discussion

Carcinoid tumours are a well described, but rare entity arising from neuroendocrine cells. First described over 100 years ago by Lubarsch, they tend to behave in a more indolent progression than adenocarcinomas. 

![CT abdomen](image1.png)

**Fig. 1.** CT abdomen. The tumour arises within the horseshoe kidney and is heterogeneous in nature.

![Renal isthmus-containing tumour](image2.png)

**Fig. 2.** Renal isthmus-containing tumour. The black ink and blue ink display left and right surgical margin, respectively. A well-circumscribed pale tumour can be seen superiorly with normal renal parenchyma inferiorly.
Primary carcinoid arising in the kidney is rare (<1%), being first described in 1966 by Resnick and colleagues. Lane et al. recently studied primary neuroendocrine tumours of the kidney and found only 56 published case reports of primary renal carcinoid, 10 of which arose within horseshoe kidneys. Our literature review revealed 12 articles (including the above) identifying 17 patients with a primary renal carcinoid arising in the setting of a horseshoe kidney. Carcinoid appear over-represented in horseshoe kidneys compared with normal renal anatomy.

Carcinoid tumours arise from neuroendocrine cells but because neuroendocrine cells are not identified within the kidney or renal pelvis, the precise pathogenesis of these lesions remains unclear. Current hypotheses suggest that these tumours derive from interspersed neuroendocrine cells associated with acquired and/or congenital abnormalities. Hypotheses currently include

1. Metaplasia of the urothelium induced by chronic inflammation
2. Metastasis from an unknown primary
3. Abnormal migration of neural crest cells
4. Concurrent congenital renal abnormalities, most commonly arising in the setting of a teratomatous respiratory or gastrointestinal epithelium within the kidney.

The final hypothesis receives the most support, given that there has been coexistence of congenital renal abnormalities (horseshoe kidney) and teratoma in several of the previously reported case studies, suggesting a role for predisposing embryological factors.

The diagnosis of carcinoid tumours of the kidney is often made incidentally and patients are usually asymptomatic at time of diagnosis. Common presenting symptoms are similar to that of other renal tumours, including abdominal, back or flank pain, hematuria, constipation and fever. Up to a quarter of patients have a palpable abdominal mass. The carcinoid syndrome,
characterised by skin flushing and telangiectasia, diarrhoea and abdominal pain, cardiac valvular lesions, and bronchoconstriction, has not been reported to occur within a tumour arising in the setting of a horseshoe kidney\textsuperscript{[2]}. Carcinoids display slow growth and remain clinically undetected despite being present for many years. Commonly observed at a mean age of 50 years, they appear to have no predilection for gender. Their size at diagnosis is large; the retroperitoneum is able to expand with relatively little impedance before becoming symptomatic and thus the average size at diagnosis of primary renal carcinoid has been reported to be 8 cm. Fourteen of 17 horseshoe kidney carcinoids were greater than 5 cm at diagnosis\textsuperscript{[1]}. The most common finding at radiologic imaging are solid, circumscribed lesions, often appearing heterogeneous on CT and ultrasound with minimal enhancement on contrast CT, thus making differentiating renal carcinoid from renal cell carcinoma unreliable on imaging alone\textsuperscript{[7]}. Newer methods for characterising carcinoid include the use of octreotide scintigraphy. Around 80\% of carcinoid tumours express somatostatin receptors, and somatostatin receptor scintigraphy with radiolabelled octreotide shows an 85\% sensitivity for disease. This method of imaging has advantages in detecting carcinoid lesions before CT and magnetic resonance imaging detection. Its disadvantage occurs within the kidneys; octreotide’s main elimination is via the renal system, and this may obscure suspicious lesions within the kidney\textsuperscript{[8]}. All patients should receive somatostatin scintigraphy as part of their disease staging and follow-up process.

Microscopically, renal carcinoids are characterised by tightly packed cords and trabeculae of neoplastic cells appearing eosinophilic with a granular cytoplasm and nuclei containing fine stippled chromatin. These impart a ribbon-like appearance, with minimal stroma often composed of fibrous tissue. Calcifications are present in around 25\% of cases, and frequent mitosis (>2 per 10 HPFs) are characteristically absent\textsuperscript{[2,9]}. Immunohistochemical stains are utilised in the diagnosis and exclude other sites as a primary lesion. Nearly all lesions reported are positive for synaptophysin and chromogranin. Staining for vimentin and CAM 5.2 has been more variable. These stains detect the intracytoplasmic secretory granules of neuroendocrine tumours. Other important immunochemical markers are TTF-1 and WT-1, markers for lung carcinoid and Wilms tumour, respectively. These are common differentials in a carcinoid arising within the kidney\textsuperscript{[11]}. The mainstay of treatment for primary renal carcinoid is resection, because it is the only treatment known to cure patients of their primary disease. Procedures include partial nephrectomy, surgical excision of isthmus tumours to total nephroureterectomy. There is no consensus on which technique is favoured, and patients should be assessed on a case by case basis.

Chemotherapeutic regimes in the management of primary renal carcinoid have been utilised, but little evidence exists that this improves survival. To date, there is no published material specifically dealing with chemotherapy for renal carcinoids. Somatostatin analogues, most notably octreotide, are used in reducing the symptoms of the carcinoid syndrome but where this is absent, they have no role in therapy\textsuperscript{[10]}. Because extended follow-up periods have been previously observed, and during this time new metastatic disease has been identified, it is important to continue to monitor patients for evidence of recurrence\textsuperscript{[8]}. Octreotide scintigraphy is the most valuable imaging investigation and the monitoring of serum chromogranin and urinary 5-hydroxyindoleacetic acid provides a valuable biochemical marker (when present) for detecting disease progression or recurrence after treatment. The frequency of imaging and biochemical monitoring should be tailored to the individual patient; there are few data to suggest exactly within which time frame this should occur; previous authors have suggested 6-monthly and 12-monthly biochemical and imaging screening, respectively. It is important that patients are reviewed over the long-term, and new disease presentations are managed accordingly.

**Teaching point**

Although rare, carcinoid tumour arising within the kidney is a well-defined entity. The diagnosis is confirmed by immunohistochemistry, and surgical excision provides the only definitive treatment modality to date. Metastases are described but are infrequent and the incidence of carcinoid syndrome accompanying a renal carcinoid is very rare. Imaging of choice involves CT scanning and octreotide scintigraphy. Follow-up is essential in the long-term, as these lesions have recurred.
References


