CASE REPORT

Atypical carcinoid presenting as dumb-bell-shaped tumour in the normal kidney

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SUMMARY

Carcinoid tumours are low-grade malignant neoplasms with neuroendocrine differentiation and occur frequently in the gastrointestinal and respiratory tracts. Primary carcinoid tumours of the kidney are rare and a majority of these tumours occur in anomalous kidney and exhibit typical renal carcinoid morphology. We reported a middle-aged man with primary atypical carcinoid tumour occurring in a normal kidney. The patient was diagnosed as having renal cell carcinoma owing to a lack of neuroendocrinal clinical features. Immunohistochemical staining of the nephrectomy specimen helped in the diagnosis of atypical renal carcinoid.

BACKGROUND

Carcinoid tumours are slow growing malignancies of neuroendocrinal origin with distant biological and clinical effects. Their frequency corresponds to the site-density of the neuroendocrinal cells in the different organs. The intestine (including rectum), being the largest endocrine organ in humans, represents the most common site of occurrence of these tumours. Other organ systems in order of frequency include the bronchopulmonary system, hepatobiliary system, urogenital system, skin and soft tissues. Among the urogenital system, carcinoid tumours involve ovaries and testes. A primary carcinoid tumour very rarely involves the kidneys and a majority of the reported cases belong to the class of typical renal carcinoids. Atypical renal carcinoids are extremely rare. We reported a case of a primary atypical carcinoid tumour involving a normal kidney in a middle-aged man.

CASE PRESENTATION

A 37-year-old man presented with a progressive swelling and heaviness in the right flank for 20 days. There was no history of fever, loss of weight/appetite, lithuria, haematuria, headache, flushing, palpitations or diarrhoea. He was hypertensive and diabetic for the past 5 years. A general examination revealed mild pallor. Perabdominal palpation revealed a large, firm, non-tender mass in the right hypochondrium extending up to the right lumbar region. Upper and lower pole of the mass was reachable.

INVESTIGATIONS

Laboratory test results revealed normal blood counts, serum chemistry and urinalysis. Contrast-enhanced CT of the abdomen showed a heterogeneously enhancing right renal mass with compression of renal vascular pedicle. There was no evidence of local infiltration or distant metastasis.

Figure 1 Gross photograph of the renal tumour showing dumb-bell appearance.

DIFFERENTIAL DIAGNOSIS

Clinical diagnosis of the renal cell carcinoma was suspected.

TREATMENT

The right open radical nephrectomy was performed.

OUTCOME AND FOLLOW-UP

Gross examination of the resected specimen showed a dumb-bell-shaped mass replacing the entire renal parenchyma (figure 1). One half of the mass measured $14 \times 10 \times 7$ cm and the other was $12 \times 10 \times 7$ cm in size. Outer surface of the tumour was bosselated. The cut surface of the tumour showed variegated areas of necrosis and haemorrhage.

On microscopy, tumour cells were arranged predominantly in tubules, ribbons and in sheets (figure 2). These cells had round, mildly anisomorphic nuclei, inconspicuous nucleoli and moderate amount of cytoplasm. The tumour cells showed stippled nuclear chromatin and mitotic activity $>2/10$ high-power field. Necrosis was evident at the periphery of the tumour. Ureter and renal vein did not show any tumour infiltration. Immunohistochemistry showed tumour cells positive for cytokeratin, neuron-specific enolase, synaptophysin and chromogranin (figure 3).

On the basis of the microscopical and immunohistochemical findings, a final diagnosis of atypical carcinoid of the kidney was made. Upper and lower gastrointestinal endoscopy and chest CT did not show any evidence of primary site. At 6 months of

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Figure 2 Tumour is composed of tubules lined by cells with round nuclei and pale granular eosinophilic cytoplasm and frequent mitosis (H&E, ×400).

follow-up, there was no clinical evidence of metastasis. However, because of the financial constraint, nuclear scan could not be performed to rule out if there was any evidence of intestinal carcinoid which could have been missed by endoscopy.

DISCUSSION

Renal carcinoids were considered as distinct entity in the 2004 WHO classification.¹ Primary renal carcinoids are extremely rare: first documented in literature by Resnick *et al*² in 1966. The true incidence of renal carcinoid tumours is not known owing to under-reporting of this carcinoma due to the lack of familiarity with this rare entity.³

Pathogenesis of this renal carcinoid tumour is uncertain as neuroendocrine cells are not found in renal parenchyma, pelvis or ureter. Primary renal carcinoid tumours are probably derived from interspersed neuroendocrine cells as a result of acquired/congenital abnormalities such as misplaced or entrapped neural crests or pancreatic tissue in the kidney during embryogenesis, activation of gene sequences common to neuroendocrine programmed cells in multipotent stem cells or metaplasia of pelvicalyceal urothelium induced by chronic inflammation. That is why these tumours are often associated with other renal anomalies such as horseshoe kidney (18%), renal teratoma (14%) and polycystic kidney disease (2%). Approximately 30% cases are incidentally discovered.³ Krishnan *et al*⁴ reported a calculated relative risk of 62 for occurrence of renal carcinoid tumour in a horseshoe kidney.

Figure 3 On immunohistochemistry, tumour cells show positive cytoplasmic staining for synaptophysin (×400).

The mean age of the occurrence for renal carcinoid (49 years) is lower than that of renal cell carcinoma. The most common presentation is either with abdominal mass, abdominal pain or with haematuria.¹ Neuroendocrine syndromes occur only in about 12% cases.² Since, there are no definite radiological features of these tumours, preoperative diagnosis is difficult. Nuclear scans may diagnose neuroendocrine tumours elsewhere like in the gastrointestinal tract, but normal renal uptake of tracer obscure the primary renal carcinoid tumour. Even on histopathological examination, about 14.5% of primary renal carcinoid tumours are diagnosed only on retrospective analysis due to its rarity and absence of neuroendocrine clinical features.⁵

The differential diagnosis of primary renal carcinoid tumour includes primitive neuroectodermal tumour, small cell carcinoma of kidney metanephric adenoma, neuroblastoma, oncocytoma and papillary renal cell carcinoma.

Histologically majority of the renal carcinoids reported in the world literature are typical carcinoids with low mitotic activity and absence of necrosis.¹ Atypical renal carcinoid tumours having mitotic activity of more than two and presence of necrosis are extremely rare.^{3 6–8}

The renal carcinoids usually have less aggressive behaviour than other histologically similar renal tumours including renal cell carcinomas and primitive neuroectodermal tumours.⁹ Therefore, initial recognition and diagnosis is important. A low tumour stage at diagnosis is the most favourable prognostic factor and radical nephrectomy with surveillance is the current recommended management of these tumours.

Learning points

- ▶ Atypical renal carcinoid tumours are extremely rare.
- ▶ Most reported cases occur in an anomalous kidney and rarely affect a normal kidney.
- ▶ Lack of neuroendocrinal clinical features, lack of specific radiological picture and normal tracer uptake on nuclear scans often obscure the diagnosis.
- ▶ Use of appropriate immunohistochemical markers help in differentiating primary renal carcinoid tumours from histologically similar looking tumours-like renal cell carcinoma and primary neuroectodermal tumours.
- ▶ Renal carcinoid tumours have better prognosis than other malignant renal tumours.

Contributors RV conceived and wrote the manuscript. PG collected clinical/pathological data

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