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Author(s)	Chung, HY; Lau, WH; Chu, SM; Collins, RJ; Tam, PC
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Carcinoid tumour of the kidney in a Chinese woman presenting with loin pain

HY Chung 鍾浩宇
WH Lau 劉泳雄
SM Chu 朱世文
RJ Collins 高 嶺
PC Tam 談寶鏵

Renal carcinoid tumours are uncommon. The aetiology is not yet fully understood and there is still no useful diagnostic tool for detecting them. We report our experience managing a Chinese woman with a primary renal carcinoid tumour.

Introduction

Carcinoid tumours of the kidney are rare. The first case was reported by Resnick et al in 1966.¹ A review of the literature shows that 56 cases of primary renal carcinoid have been reported²; most of them in Caucasians. To our knowledge, the disease has never been reported in a Chinese population although Asian countries, including Japan, have reported a few cases. We report our recent experience managing a Chinese woman with a carcinoid tumour of the kidney who presented with acute loin pain. All the relevant clinical, radiological, operative, and pathological findings are presented.

Case report

A 29-year-old woman with a history of good past health presented in October 2006 with a first episode of right loin pain, associated with urinary frequency. Physical examination was unremarkable and no abdominal masses were palpable. A computed tomographic (CT) scan with contrast showed a 5.8 x 7.7 cm well-circumscribed heterogenous mass at the middle and lower poles of the right kidney (Fig 1). The mass had no fat components but mild arterial enhancement with enlarged feeding arteries was seen. There was also no lymphadenopathy. The overall features suggested a renal cell carcinoma.

In view of the suspected renal cell carcinoma, a radical nephrectomy was performed in early November 2006. At operation, a 9-cm centrally located tumour arising from the posterior part of the right kidney near the renal hilum and stretching the hilum anteriorly was observed. She recovered well after the operation.

Macroscopically, the resected kidney (Fig 2a) contained a large, smooth-surfaced, haemorrhagic, tan-coloured tumour mass. The tumour measured 8 x 7 x 6 cm. It arose from the middle and lower poles of the kidney but protruded into the hilar region and was surrounded by a thin capsule. The renal pelvis and proximal ureter were also displaced. Histologically, the tumour was separated from the adjacent parenchyma by a well-defined thick fibrous capsule. It was composed of cords, ribbons, and tubules of relatively uniform cells with little intervening fibrous stroma. The cells were cubo-columnar and medium-sized with moderate eosinophilic cytoplasm. The nuclei showed minimal variation, and were round to oval with coarse chromatin and small nucleoli. Extensive haemorrhage and congestion were noted. There was no evidence of vascular invasion and the resection margin was clear.

Because the tumour did not resemble a usual renal carcinoma, immunohistochemical studies were performed. It was negative for renal cell marker CD-10 (CALLA), cytokeratin CK7 and CK20, pulmonary marker TTF-1, calcitonin and GCDFP. It did show diffuse immunoreactivity to CD56 (N-CAM), but was negative for neuroendocrine markers chromogranin and synaptophysin. A low proliferative index was illustrated by the proliferative marker mib-1. An ultrastructural study showed that most cells possessed many membrane-bounded dense core granules ranging in size from 140 to 300 nm (Fig 2b). These overall features were typical of a renal carcinoid tumour.

Our patient was followed up regularly after the operation and a CT scan of the abdomen and thorax performed 1 year later showed no evidence of secondaries. She was also offered an octreotide scan to detect recurrence or metastases but this was not carried out for financial reasons. She has never had any symptoms suggestive of carcinoid syndrome and her urinary 5-hydroxyindole acetic acid level was within the normal range.

Key words

Carcinoid tumor; Kidney neoplasms

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Queen Mary Hospital, Pokfulam Road,
Hong Kong:
Department of Surgery
HY Chung, MB, BS
WH Lau, MB, BS
SM Chu, FRCS (Edin)
PC Tam, FRCS (Urology) (Edin)
Department of Pathology
Robert J Collins, FRCPA

Correspondence to: Dr HY Chung
E-mail: phychung@graduate.hku.hk



FIG 1. Computed tomographic scan of the abdomen showing the lesion before operation

Discussion

The pathogenesis of renal carcinoid tumours is still debatable. Krishnan et al³ reported a frequent association with a horseshoe kidney, and related it to the presence of teratomatous epithelium. It is believed the tumour cells are derived from multipotential primitive stem cells capable of neuroendocrine differentiation.^{4,5} Unlike carcinoid disease of other organs, renal involvement tends to occur in the younger age-group with a mean age of 45.2 years and an equal incidence in males and females.⁶

In most reported cases, renal carcinoid tumours were diagnosed after pathological examination of resected specimens that were initially thought to be renal cell carcinoma. In symptomatic patients, abdominal masses and acute loin pain are the most common presenting symptoms. An associated carcinoid syndrome is uncommon.² There are still no definite radiological features able to indicate the diagnosis before operation. A carcinoid tumour is often indistinguishable radiologically from other renal tumours such as renal cell carcinomas, making it a diagnostic challenge for most clinicians managing the disease. Although nuclear medicine scans have been used extensively to detect neuroendocrine tumours of the gastro-intestinal tract, the drawback when evaluating primary renal carcinoid tumours is that normal renal uptake of the tracer material may obscure the lesion.² Nevertheless, some authors believe it may help to detect residual disease in patients who are found to be disease-free by a postoperative CT scan.^{7,8} We usually recommend such a scan to our patients as a self-financed item. There is still no standard protocol for follow-up of patients with renal carcinoid tumours. Measurement of hormone production is not useful for making the diagnosis as few patients with reported renal

一名患有腎類癌並呈現腰痛的華裔女性

腎類癌臨床比較少見。到目前為止，未能完全理解其病理，亦未有有效的方法診斷此病。本報告描述我們治理一位患有原發性腎類癌的華裔女性的經驗。

carcinoid tumours showed an elevation of 5-hydroxyindole acetic acid.⁷ Nonetheless, it is a useful means of monitoring disease progress.

Since renal carcinoid tumours have a reported metastatic incidence of approximately 25.7%,⁶ most surgeons believe radical nephrectomy should be offered. The size of the tumour reportedly correlates well with the incidence of metastasis.⁹ The most common sites for metastasis are the pre hilar lymph nodes and liver, followed by the bones and lungs.² Radiolabelled somatostatin analogues are being used by endocrine surgeons to manage metastatic unresectable neuroendocrine tumours although no conclusion about their effectiveness has been reached. In general, the prognosis is good for renal carcinoid tumours, with reported survival rates reaching 88.6%.⁶

Renal carcinoid tumours are uncommon in the Chinese population. This case illustrates management of a renal mass with special reference to renal carcinoid tumours. Although it is difficult to diagnose this disease without an operation, it is potentially curable and regular follow-up is important for detecting metastases if present.

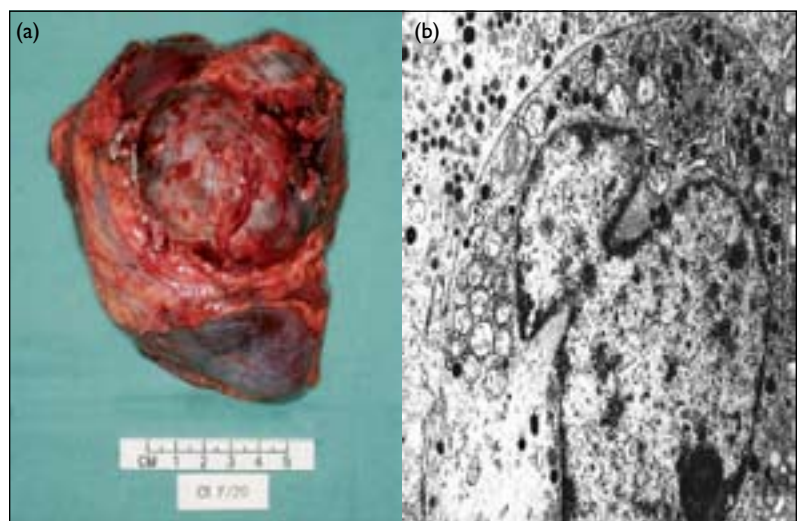


FIG 2. (a) Macroscopic and (b) ultrastructural views of the resected specimen

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