Spontaneous rupture of the kidney affected by multifocal papillary renal cell carcinoma

Lucio Dell'Atti

Department of Urology, University Hospital S. Anna, Ferrara, Italy

Abstract

Papillary renal cell carcinoma (pRCC) represents the second most common type of malignant renal epithelial tumor (represents the 10% of the kidney's carcinoma) and can be subclassified in the basophile type I and eosinophile type II. We report a clinical case of spontaneous rupture of the kidney affected by multifocal (42 tumors foci) pRCC in a young man 53 years old, without showing earlier specific cancer signs and symptoms. Prognosis for type I pRCC is better than type II pRCC, but it is anyway related to the tumoral grade, to the tumoral stage and to the diagnostic precocity. Signs and symptoms are very similar to those characterizing the more frequent clear cell carcinoma. Nevertheless in the 40% of the cases the lesion is asymptomatic. To our knowledge, this is the first case of spontaneous rupture of the kidney affected by multifocal pRCC in literature without showing earlier specific cancer signs and symptoms.

Introduction

Papillary renal cell carcinoma (pRCC) represents 10% of the kidney's carcinoma.[1] This disease affects more frequently male gender (M/F:3/1) during the fifth-sixth decade, showing usually like a sporadic disease, even though it could arise even as an hereditary form; in the first case the kidney presents a single tumor, while in the second case one or both kidneys are involved by bilateral neoplasms.[2,3] pRCC is more frequently multifocal than other types of renal cell carcinoma.[2] The Heidelberg classification system is used to classify renal cell carcinoma (RCC) subtypes as clear cell renal cell carcinoma (ccRCC), papillary, chromophobe, collecting duct, or as unclassified RCC. pRCC is subclassified in the basophile type I and eosinophile type II.[3] Signs and symptoms are very similar to those characterizing the more frequent clear cell carcinoma: hematuria, pain, palpable mass are the three classic manifestations. Nevertheless in the 40% of the cases the lesion is asymptomatic.[4] We report here the only clinical case of spontaneous rupture of the kidney affected by multifocal pRCC in a young man 53 years old.

Case Report

A Caucasian 53-years-old man arrived at our emergency room in October 2013 reporting intense pain to the left side aroused about a couple of hours before, after some gardening. His past medical history showed obesity (BMI>30 kgm^{-2}), diabetes, hypertension, chronic renal insufficiency with the right atrophic kidney since his birth. His past surgical history: heart transplant at the age of 40 years due to a dilated cardiomyopathy. Soon after his arrival the patient showed drowsiness and unconsciousness. On physical examination, oral temperature was 36.0°C, blood pressure 80/50 mmHg, heart rate 110 beats/min and pulse oximetry 96% on room air. Laboratory tests shown hemoglobin: 6.9 g/dL, white blood cells: 13.04×10^{3}µL, hematocrit: 30% and creatinine 4.7 mg/dL. After an emergent fast ultrasound was performed CT with contrast-enhanced and it was possible to appreciate the presence of a great amount of blood in the peri and para-renal left kidney zone, generated by a lesion of the kidney's parenchyma (Figure 1). The patient underwent a surgical exploration and a left nephrectomy. Macroscopically the kidney showed a prominent cystic component. The cysts were filled with serosanguinous fluid. Hemorrhages were also noted and no necrosis was present. The histology exam of the surgical frame came out on the side interested by the multifocal papillary renal cell carcinoma type I (with 42 foci, which largest diameter measured circa 7 mm). Microscopically the tumors were composed of a mixture of cysts and papillae. Irrespective of the architecture, the tumors were composed entirely of cuboidal or columnar cells with clear cytoplasm, small hyperchromatic, round or oval nuclei, and inconspicuous nucleoli (Figure 2). Marked stromal lymphoplasmacytic infiltrates, non-necrotizing epithelioid cell granulomas, organizing hemorrhage with hemosiderin deposits and capsular calcifications were also noted. The stromal component formed bands of fibrous and leiomyomatous tissue inside the tumors. After 3 days of intensive care unit hospitalization the patient died because of cardio circulatory complication.

Discussion

Papillary renal cell carcinoma represents the second most common type of malignant renal epithelial tumor.[5] Two are the histological types of multifocal papillary renal cell carcinoma, and it is fundamental to distinguish them since they have a different prognosis: Type I is more frequently multifocal, it is made of papillae covered by small cells with modest cytoplasm, with a low nuclear grade, and dislocated in one single layer upon the papillae basement membrane; Type II: is made of papillae covered by cells with eosinophilyc cytoplasm.[6] Chromosomal and cytogenetic analyses have revealed gain of chromosomes 7 and 17, loss of Y chromosome, and additional gains (chromosome 3q,8p,12q,16q, and 20q) in type I pRCC, but the chromosomal aberrations of type II pRCC seems to be more heterogeneous.[7] Prognosis is better for type I pRCC than type II pRCC, but it is anyway related to the tumoral grade, to the tumoral stage and to the diagnostic precocity.[4,8] Our case confirms that pRCC is a tumor with a predominance during the fifth-sixth decade and the tumor is associated with chronic failure in 40% of cases.[9] Multifocal pRCC continues to be an unresolved clinical problem from diagnostic, prognostic and therapeutic perspectives. In fact, the clinical presentation is non-specific, and in most cases the tumor is discovered incidentally, as in our case where the diagnosis was accidental after spontaneous rupture of the kidney. Macroscopically, most of these tumors were cystic and well-delimited; moreover, the tumor architecture was composed predominantly of short and aborted papillary structures but, in some cases, a prominent tubular or solid pattern was observed, potentially causing confusion with RCC, even if it does not have the characteristic yellow-gold color of conventional RCC. Microscopically, pRCCs were predominantly papillary or tubopapillary, often with foam cells, necrosis, hemorrhage and multifocality.[10] Mancilla-Jimenez *et al.*[11] in 1976 have been two of the first to recognize pRCC as a distinct RCC type with unique clinicopathological features. Furthermore, two subtypes of pRCC show different clinicopathologic behaviors: type II tumors are associated with a poorer prognosis than type

Correspondence: Lucio Dell'Atti, Department of Urology, University Hospital "S.Anna", 8 A. Moro Street, 44124 Cona, Ferrara, Italy.
Tel.:+39.0532.236078 - Fax: +39.0532.236547.
E-mail: dellatti@hotmail.com

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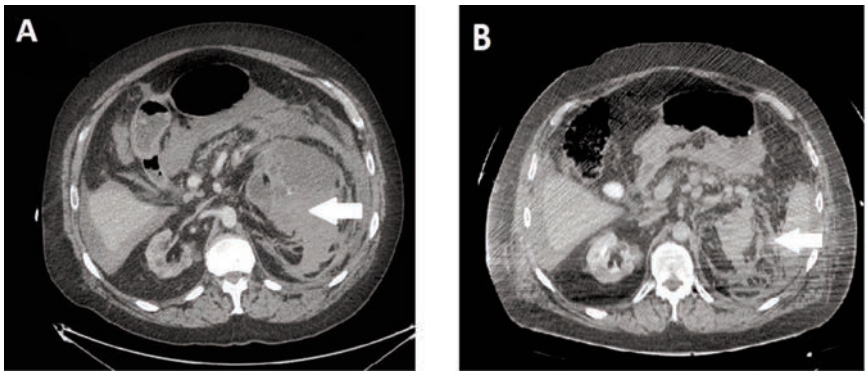

Figure 1. Unenhanced (A), corticomedullary phase CT scans showing a great amount of blood in the peri and para-renal left kidney zone, generated by a lesion of the kidney's parenchyma (B).

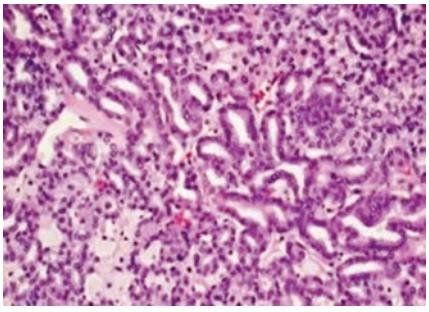

Figure 2. Type I papillary renal cell carcinoma: basophilic cuboidal or columnar cells with clear cytoplasm, small hyperchromatic, round or oval nuclei, and inconspicuous nucleoli.

I tumors and even than clear cell RCC.[12-15] Four studies reported a significant difference,[16-19] showing less aggressive histopathological variables for type I pRCC, whereas the study conducted by Yamanaka *et al.*[20] showed no significant difference between type I and II pRCC in almost all clinicopathological variables, including age, gender, clinical stage, metastasis, pathological stage, tumor grade and vascular invasion. Only in the study of Delahunt and Elbe,[16] multifocality was significantly associated with type I pRCC whereas Mejean *et al.*[19] and Pignot *et al.*[17] found no significant difference between the two types. This case was consistent with previous reports regarding his age and number of foci in multifocal tumors. Multifocality of type I pRCC is frequently reported in these tumors as in our present case.

Conclusions

Case reports in the literature, also bilateral, of multifocal pRCC have been described in patients with autosomal dominant polycystic kidney disease, but they were not so multiple (in a number of 42 tumor foci) in order to cause the spontaneous rupture of the renal parenchyma. To our knowledge, this is the first case of spontaneous rupture of the kidney affected by multifocal pRCC in literature without showing earlier specific cancer signs and symptoms.

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