

## Oncology

## Sarcomatoid chromophobe renal cell carcinoma revealed by acute pyelonephritis in a diabetic patient

Raja Jouini<sup>a</sup>, Ayoub Saidi<sup>a,\*</sup>, Fatma Khanchel<sup>a</sup>, Imen Helal<sup>a</sup>, Maher Chtourou<sup>b</sup>, Aschraf Chadli-Debbiche<sup>a</sup><sup>a</sup> Department of Pathology, Habib Thameur Hospital, Faculty of Medicine of Tunis, Tunis El Manar University, Tunis, Tunisia, 8, Ali Ben Ayed Street, 1008, Tunis, Tunisia<sup>b</sup> Department of Urology, Habib Thameur Hospital, Faculty of Medicine of Tunis, Tunis El Manar University, Tunis, Tunisia, 8, Ali Ben Ayed Street, 1008, Tunis, Tunisia

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## ABSTRACT

Chromophobe renal cell carcinoma (CRCC) is a rare tumor comprising 2 types of cells (eosinophilic and clear cells), coexisting in varying proportions.

CRCC has an overall good prognosis. However, the rarely encountered sarcomatoid differentiation is associated with a pejorative outcome and must, therefore, be rigorously ruled out.

Acute pyelonephritis and other infectious complications rarely occur.

We report a case of CRCC with sarcomatoid differentiation in a woman in her sixties which was revealed by acute pyelonephritis and had a lethal outcome.

## Introduction

Chromophobe renal cell carcinoma (CRCC) is a rare tumor described for the first time by Thoenes in 1985.<sup>1</sup> It usually comprises 2 types of cells (eosinophilic and clear cells), coexisting in varying proportions.

CRCC has an excellent prognosis, the best among all types of renal cell carcinoma. Nevertheless, a pejorative outcome has been noted in cases with large tumors, necrosis or sarcomatoid differentiation.<sup>1</sup>

CRCC is often incidentally discovered on imaging performed for another purpose. However, if present, clinical signs usually include abdominal pain, hematuria and abdominal mass syndrome.<sup>1,2</sup> Acute pyelonephritis and other infectious complications rarely occur.

We report a case of CRCC with sarcomatoid differentiation in a woman in her sixties which was revealed by an acute pyelonephritis with a lethal outcome.

## Case presentation

We report the case of a 64-year-old woman with a history of diabetes mellitus admitted to the intensive care unit for septic shock caused by an acute left pyelonephritis.

CT scan showed a lower mass syndrome of the left kidney measuring 84 mm with necrosis and calcifications (Fig. 1). A wide nephrectomy

with adrenalectomy was performed urgently. Gross examination of the nephrectomy specimen showed a kidney occupied at its inferior pole by a fleshy well-defined tumor measuring 7.5 × 7 × 7 cm, solid on cutting with patchy hemorrhagic and necrotic areas. On histological examination, the sections analyzed showed a dual carcinomatous proliferation composed of clear and eosinophilic cells, adopting predominantly lobular, otherwise tubulo-cystic growth pattern.

There was no vascular invasion. Renal capsule and adrenal gland were spared.

In addition, the carcinomatous proliferation showed scattered foci of sarcomatoid differentiation made of large overlapping bundles of spindle-shaped cells (Figs. 2 and 3). On immunohistochemical study, tumor cells were negative for CD117, CD10, racemase and vimentin.

The septic shock couldn't be controlled, and the patient state deteriorated. She died a few days after her admission.

## Discussion

CRCC is a rare kidney tumor accounting for about 6%–8% of renal neoplasms in adults, and 4%–10% of renal cell carcinomas. The mean age at diagnosis is 59 years with no gender predominance.<sup>1</sup> The term chromophobe has been used as opposed to classical renal cell cancer formerly known as chromophilic.<sup>3</sup> CRCC is often incidentally

\* Corresponding author. Department of Pathology, Habib Thameur Hospital, 8, Ali Ben Ayed Street, 1008, Tunis, Tunisia.

E-mail address: [saidiayoub.md@gmail.com](mailto:saidiayoub.md@gmail.com) (A. Saidi).

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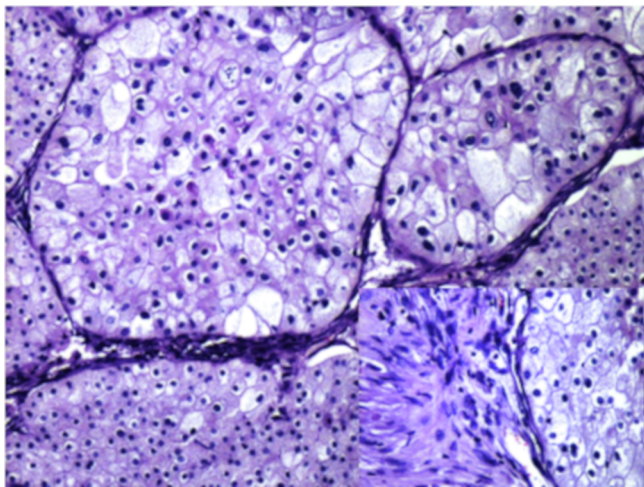
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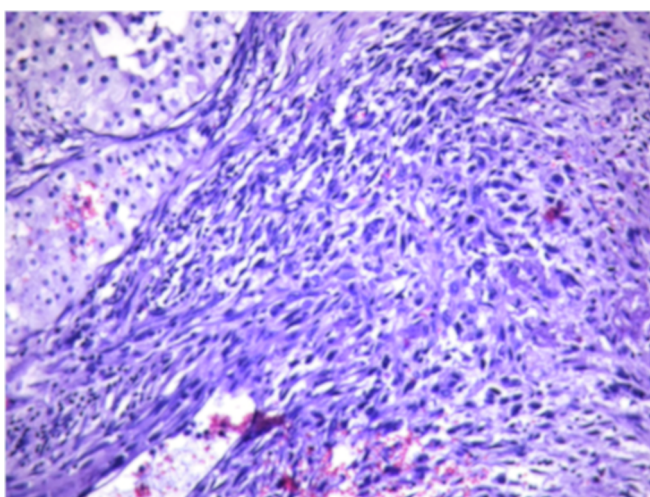
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**Fig. 1.** CT scan: a mass syndrome of the left kidney measuring 84 mm with images of necrosis and calcifications (arrow).



**Fig. 2.** A tumoral growth composed of classic chromophobe renal cell carcinoma along with a sarcomatoid spindle cell component (lower right corner image).



**Fig. 3.** The sarcomatoid component in chromophobe renal cell carcinoma is of fibrosarcoma-like type composed of intersected bundles of malignant spindle-shaped cells.

discovered. The association of CRCC to acute inflammatory conditions like acute pyelonephritis, pyonephrosis or renal abscess is an uncommon event. When this occurs, it may lead to a delayed diagnosis due to misinterpretation of radiological images. In our case, acute pyelonephritis was the clinical revealing event. This unusual presentation could be partly explained by the increased risk and the more aggressive course of acute pyelonephritis in diabetic patients. The presence of clinical symptoms is usually the evidence of an advanced disease.<sup>2</sup> These symptoms may include hematuria, abdominal mass syndrome, more rarely a decreased renal function, proteinuria and metastasis related pain.<sup>1,2</sup> On imaging, CRCCs contrast uniformly with CT injected, unlike other types of renal cell carcinoma in which enhancement is peripheral and heterogeneous. Grossly, CRCC is a uniform, well limited, non encapsulated tumor with beige to brown or yellow cutting surface.<sup>2</sup> Hemorrhagic or necrotic changes are rarely observed. Microscopically, the proliferation is variably composed of large lobules, solid nests, trabeculae, tubules or papillae. The two types of cells encountered in this tumor, chromophobe (clear) cells and eosinophilic cells, often coexist but in varying proportions. Subtyping these tumors according to the predominant cell component has no likely prognostic impact.<sup>1</sup> Fuhrman nuclear grade was applied to CRCCs, but the International Society of Urological Pathology (ISUP) Consensus Conference (2013) recommended not applying it for this type of carcinoma any longer.<sup>4</sup>

Sarcomatoid differentiation is seen in 2–9% of CRCC cases, a significantly higher proportion compared to other types of renal cell carcinoma.<sup>4</sup> The sarcomatoid component is most often of fibrosarcoma-like type showing intersected bundles of malignant spindle-shaped cells like in our case. Less frequently, the appearance is that of undifferentiated pleomorphic sarcoma (malignant fibrous histiocytoma).<sup>4</sup> A study of 101 cases proposed to categorize CRCC as sarcomatoid if there is at least one distinct spindle-cell foci in one high-magnification field.<sup>5</sup>

The overall CRCC prognosis is excellent with a 5-year and 10-year survival rate of 78%–100% and 80%–90%, respectively.<sup>1</sup> However, a high pT stage, tumor necrosis, and sarcomatoid differentiation are associated with a more aggressive behavior. These three parameters are independent predictors of poor prognosis.<sup>1</sup> In our case, the patient died few days after surgery and this could be explained by the acute pyelonephritis.

## Conclusion

Acute pyelonephritis complicating renal neoplasms has been only a few times reported in the literature and is often due to urinary tract obstruction or compression related to these tumors. Even though sarcomatoid differentiation in CRCC is rare, it must be rigorously ruled out given its negative prognostic impact.

## Declaration of competing interest

None.

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