

Early Recurrence of Chromophobic Renal Cell Carcinoma with Sarcomatoid and Rhabdoid Differentiation: About a Case

Ângelo João Peti^{1*}, Larbi Hamedoun¹, Gogui Douru Akim¹, Conde Fadama¹, Jamali Mounir¹, Hassan Ilias¹, Younes Bourkhlifi¹, Mohammed Mrabti¹, Jendouzi Omar¹, Alami Mohamed¹, Ahmed Ameur¹

¹Department of Urology of the Military Instruction Hospital Mohammed V- RABAT, Morocco

DOI: [10.36347/sasjs.2022.v08i03.015](https://doi.org/10.36347/sasjs.2022.v08i03.015)

| Received: 13.02.2022 | Accepted: 16.03.2022 | Published: 25.03.2022

*Corresponding author: Ângelo João Peti

Department of Urology of the Military Instruction Hospital Mohammed V- RABAT, Morocco

Abstract

Case Report

Chromophobic renal cell carcinoma with sarcomatoid differentiation is a rare tumor characterized by a biphasic tumor with both classic epithelial components and sarcomatoid components with poor prognostic factors, usually in association with the rhabdoid contingent which has the potential to behave aggressively and metastasize. We report a particular case of recurrence of this entity in a male patient in which the diagnosis was suspicious preoperatively for the tumor evolution time and the size of the mass and the imaging data performed. The patient underwent extensive left total nephrectomy and lymph node dissection. The postoperative follow up was marked by early appearance of recurrence within 6 weeks after surgery.

Keywords: Renal carcinoma; chromophobe carcinoma; sarcomatoid differentiation; rhabdoid differentiation; Nephrectomy, lymph node dissection.

Copyright © 2022 The Author(s): This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 International License (CC BY-NC 4.0) which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use provided the original author and source are credited.

INTRODUCTION

Sarcomatoid differentiation features in chromophobe RCC (ChRCC) is quite rare [1]. Renal cell carcinoma (RCC) with rhabdoid features has recently been established as a morphological subtype of renal neoplasm [2]. It is rare and exhibits aggressive behavior [2-4]. Rhabdoid features have been described in conventional cells (clear cells), papillary and collector RCCs [3, 4].

Most ChRCCs have a good prognosis, but the change by sarcomatoid type associated with rhabdoid is a factor of very poor prognosis with progression at high risk of malignancy and mortality, which is observed in our case.

We report clinicopathological particularity of an exceptional case of early recurrence of sarcomatoid and rhabdoid chromophobe renal cell carcinoma with aggressive behavior in a young patient.

CASE REPORT

Mr L.D, 38 years old, with no particular history and having consulted for left low back pain with a history of weight loss of 10 kg for 6 months and macroscopic hematuria without urinary disorder. On

examination, the patient presents deterioration of the general condition (PS at 1), presence of bloating at the level of the left lumbar fossa with presence of a palpable mass. The thoraco-abdomino-pelvic computed tomography showed a large mass measuring 130x120x155mm, developed depending on the left mid-renal anterior lip, pseudo-encapsulated in appearance, heterogeneous, without venous invasion with arterial enhancement and driving back neighboring structures, absence secondary location, (Figure 1, 2, and 3). Biologically, the patient had a hemoglobin level of 9.5 g/dl, leukocytes at 12.5/mm³, normal renal function with serum creatinine at 10.22 mg/L.

A radical left total nephrectomy with lymph node dissection was performed and revealed a solid tumor of 24x30x28 cm, weighing 1587g (Figure 4, 5), with infiltration of the renal capsule and extended into the perirenal fatty tissue with invasion left colic. The cut surface was grey-white-beige in appearance with necrosis.

The postoperative course was simple without incident, leaving on day 5. Under the microscope, the surgical specimen reveals a chromophobic renal carcinoma associated with the presence of foci of tumor necrosis with sarcomatoid and rhabdoid differentiation

with the presence of vascular emboli and regional lymph nodes (Figures 7, 8 and 9). The surgical resection margins were healthy.

After 6 weeks of postoperative monitoring, the patient was readmitted to the emergency room for a picture of left low back pain associated with a deterioration in general condition. Control CT scan in favor of tumor recurrence in the left renal compartment measuring 193 x 117 mm with multiple intra and retro peritoneal masses, presence of secondary lesion in hepatic segment VIII of 24 x 23 mm, without lung lesion (Figures 10, 11 and 12). Physical examination

revealed a pale appearance of integuments with blood pressure of 137/60 mm/Hg, heart rate was 142 bpm. Abdominal examination revealed mild abdominal distention with tenderness. Presence of palpable mass in the region of the left lumbar region. Laboratory tests revealed hemoglobin 7.6 g/dl, uremia was 0.26 g/l, creatinine 12 mg/dl. The white blood cells were at 15900/mm³. The balance sheets were normal. The patient had no curative surgical indication after the recurrence.

The patient died at week 8th before receiving scheduled palliative treatment (immunotherapy).

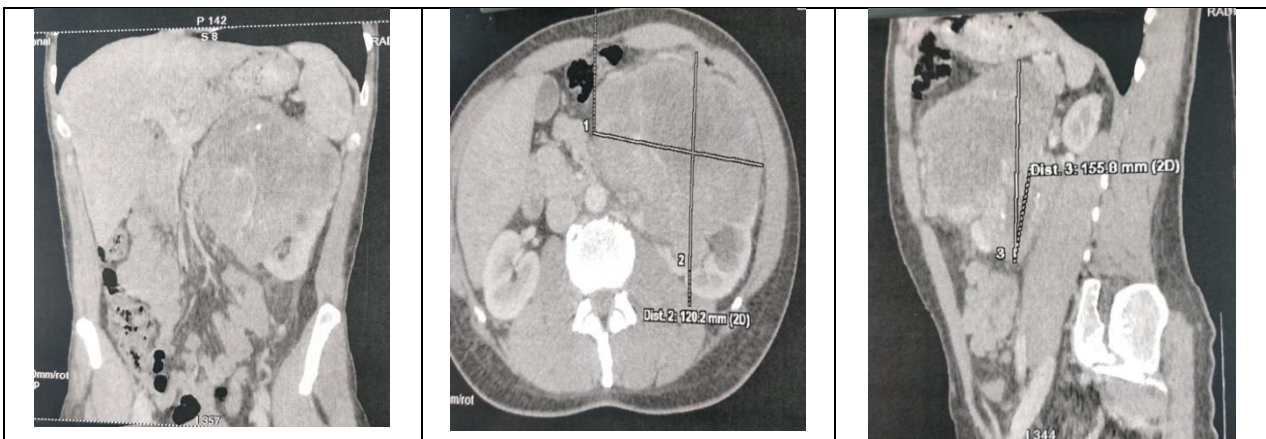


Figure (1, 2 and 3) Thoraco-abdomino-pelvic CT scan of frontal, parasagittal and axial reconstruction: highlighting voluminous left mass measuring 130 x 120x 155 mm in size

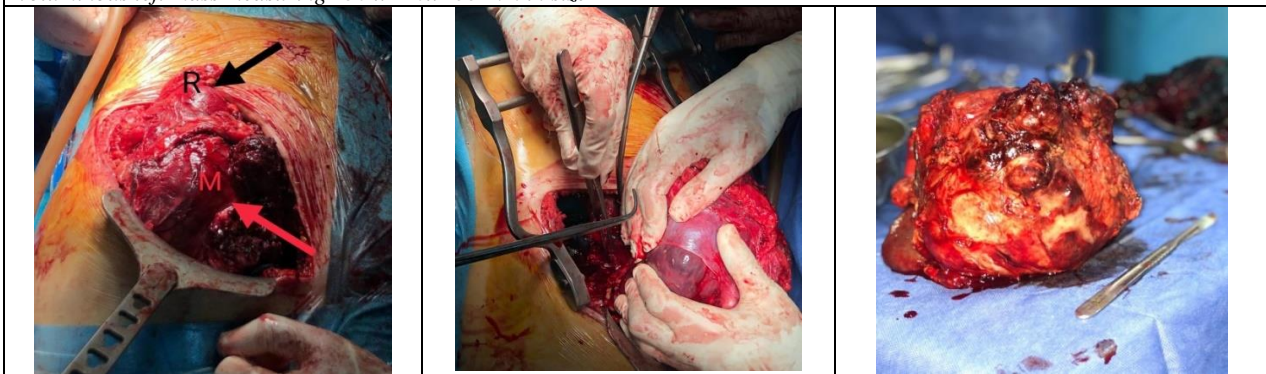


Figure (4, 5 and 6) Intraoperative macroscopic appearance showing mass exposure, retraction and preparation of the vessel before the start of the resection of the renal mass, tumor parts.

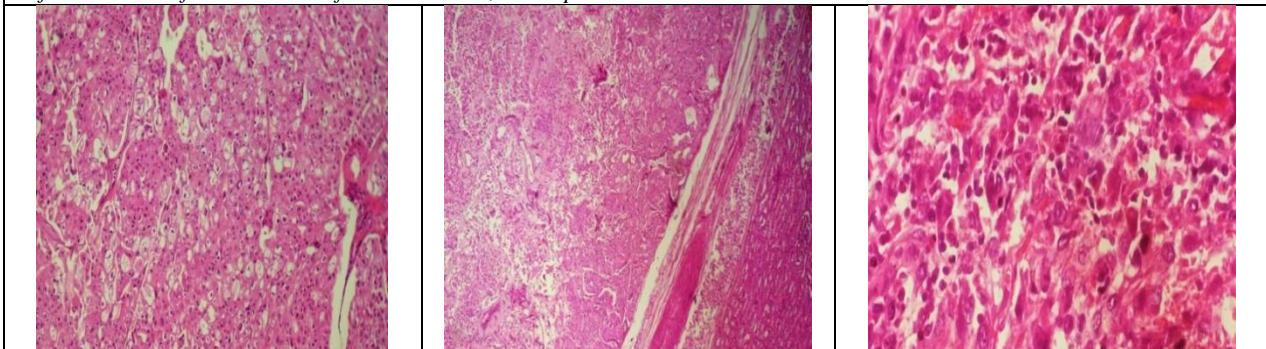


Figure (7, 8 and 9) chromophobe carcinoma with sarcomatoid and rhabdoid differentiation
 7. The sections analyzed show tumor proliferation made of cord and clusters at 4x100 magnification
 8. The sections analyzed show tumor proliferation made of cord and clusters at 4x40 magnification
 9. Tumor cells have enlarged and moderately atypical nuclei, clear perinuclear halos are also present at 4x400 magnification.

Thoraco-abdominopelvic computed tomography after six weeks of postoperative monitoring

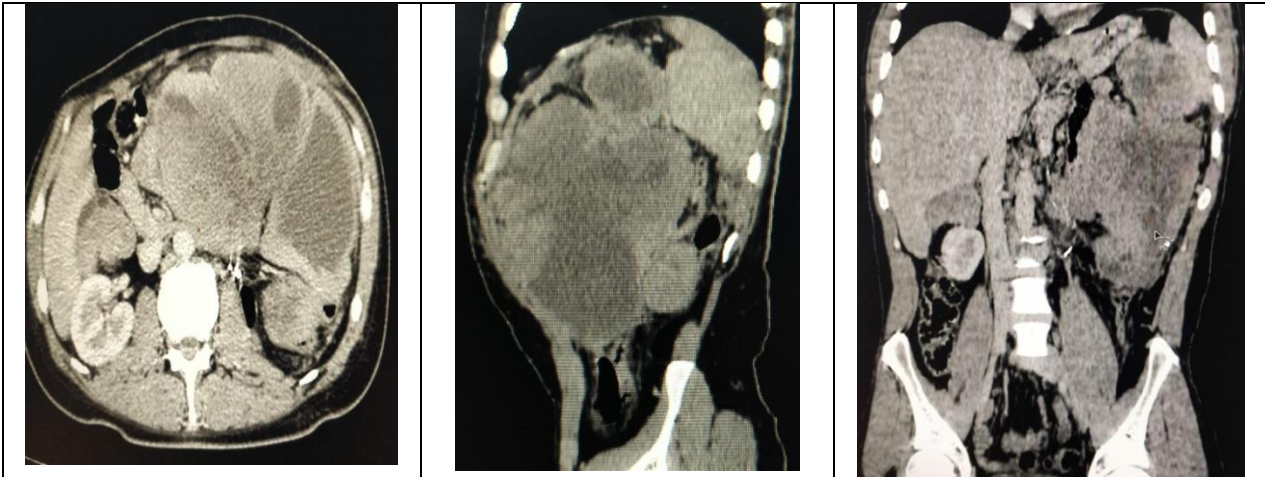


Figure (10, 11 and 12) CT. Tumor recurrence measuring 193 x 117 mm, multiple intra and retro peritoneal masses, hepatic VIII lesion measuring 24 x 23 mm.

DISCUSSION

Our case report provides further evidence that chromophobe renal cell carcinoma with sarcomatoid and rhabdoid differentiation has a propensity to progress to malignancy with high risk of mortality. And while a chromophobe CRC usually clinically presents with hematuria, flank pain, or abdominal pain, weight loss, and its altered general condition. Other patients may be symptomatic until the disease is advanced. Our patient is a 38-year-old man with a history of weight loss and anemia who presented with an advanced stage of the disease. Sarcomatoid differentiation is of poor prognosis, but association with rhabdoid component presents a very high risk of mortality as in our case is exceptionally rare or never reported in any case published in the literature.

Chromophobe renal cell carcinoma (CCRC) affects men more than women. The average age at presentation is 50 to 70 years old. CCRCS usually present as a unilateral renal mass. This tumor can also be, but rarely, bilateral. Chromophobic CRC may present with hematuria, lumbar pain or a palpable abdominal mass. It is grossly well circumscribed, solitary, with a homogeneous gray to brown cut surface, without hemorrhage or necrosis. Under the microscope, this tumor shows a characteristic nest organization of tumor cells. Tumor cells have abundant cytoplasm and well-defined boundaries. There is often a clear perinuclear region which is secondary to the presence of numerous cytoplasmic vesicles. Immunohistochemically, tumor cells express EMA, cytokeratin 7, CD82, CD9, claudin 7, claudin 8, E-cadherin and often CD10. These tumors do not express N-cadherin or vimentin. Mitotic activity is generally lower than that of clear cell CRC.

The loss of chromosomes 1, 2, 6, 10, 17 and 21 was observed by comparative genomic hybridization technique and by use of microsatellite markers. Sarcomatoid transformation in chromophobe renal cell carcinoma is extremely rare compared to other

types of RCC [5, 6]. In his series, Akhtar *et al.*, [6] reported six cases of sarcomatoid transformation in chromophobe renal cell carcinoma. In this series, all patients with chromophobe sarcomatoid carcinoma were in the fifth to seventh decades of life, which is higher than that of conventional chromophobe renal cell carcinoma in our case, suggesting that sarcomatoid transformation occurs after a relatively long [7, 8]. In this same series [6], all the tumors with sarcomatoid differentiation presented a sarcomatoid component extending beyond the renal capsule, which is the case of our patient with invasive regional lymph nodes and the presence of vascular emboli. In all cases, atypia and pleomorphism were clear in the sarcomatoid component.

So far, it is widely accepted that the development of rhabdoid morphology appears to represent a common dedifferentiation pathway for malignant renal parenchymal tumors, with clonal progression to an aggressive high-grade biological state [9].

Pure rhabdoid tumors of the kidney are mainly encountered in the pediatric population, with very early metastatic extension and a poor prognosis [10].

Our case is an example of ChRCC with rhabdoid differentiation, a rare tumor with only one case report published in the literature [11]. Rhabdoid areas were associated with high histological grade and were surrounded by abundant positive fibrovascular stromal immunohistochemistry for SMA. Metastases were also found on CT scan and indicated aggressive behavior of this malignancy.

Sarcomatoid transformation of chromophobe renal cell carcinoma is less studied than that of clear cell carcinoma which has been extensively studied [12]. In all these cases, the primary renal tumor was a clear cell type renal cell carcinoma with bleeding as the predominant clinical feature, whereas the primary renal

tumor in our case was a chromophobe RCC of sarcomatoid and rhabdoid differentiation with secondary colonic invasion as the presentation. Clinic making our patient a unique case presenting these two characteristics.

CONCLUSION

Our case is characterized by its diversity histological subtype of ChRCC mixed by sarcomatoid contingent and rhabdoid areas, are relatively rare variants with an unfavorable clinical and prognostic course, it is consistent with a high risk of metastasis and greater relapse. Symptomatic discovery at an advanced stage, requiring rapid and appropriate multidisciplinary care. Metastatic or recurrent forms are linearly related with tumor size and grade. Total nephrectomy remains the gold standard treatment for localized kidney cancer and medical treatment for metastatic cancer.

Declaration of Interests: The authors declare that they have no conflicts of interest in relation to this article.

REFERENCES

1. Przybycin, C. G., Cronin, A. M., Darvishian, F., Gopalan, A., Al-Ahmadie, H. A., Fine, S. W., ... & Tickoo, S. K. (2011). Chromophobe renal cell carcinoma: a clinicopathologic study of 203 tumors in 200 patients with primary resection at a single institution. *The American journal of surgical pathology*, 35(7), 962-970.
2. Eble, J. N., Sauter, G., Epstein, J. I., & Sesterhenn, I. A. (2004). World Health Organisation classification of tumours. Pathology and genetics of tumours of the urinary system and male genital organs. Lyon: IARC Press; pp. 58–59. (Chapter 1, Rhabdoid tumour).
3. Shannon, B., Wisniewski, Z. S., Bentel, J., & Cohen, R. J. (2002). Adult rhabdoid renal cell carcinoma: divergent differentiation of conventional (clear cell) carcinoma. *Archives of pathology & laboratory medicine*, 126(12), 1506-1510.
4. Leroy, X., Zini, L., Buob, D., Ballereau, C., Villers, A., & Aubert, S. (2007). Renal cell carcinoma with rhabdoid features: an aggressive neoplasm with overexpression of p53. *Archives of pathology & laboratory medicine*, 131(1), 102-106.
5. Gomez-Roman, J. J., Mayorga-Fernandez, M., Fernandez-Fernandez, F., & Val-Bernal, J. F. (1997). Sarcomatoid chromophobe cell renal carcinoma: immunohistochemical and lectin study in one case. *General & diagnostic pathology*, 143(1), 63-69.
6. Akhtar, M., Tulbah, A., Kardar, A. H., & Ali, M. A. (1997). Sarcomatoid renal cell carcinoma: the chromophobe connection. *The American journal of surgical pathology*, 21(10), 1188-1195.
7. Taki, A., Nakatani, Y., Misugi, K., Yao, M., & Nagashima, Y. (1999). Chromophobe renal cell carcinoma: an immunohistochemical study of 21 Japanese cases. *Modern pathology: an official journal of the United States and Canadian Academy of Pathology, Inc.*, 12(3), 310-317.
8. Latham, B., Dickersin, G. R., & Oliva, E. (1999). Subtypes of chromophobe cell renal carcinoma: an ultrastructural and histochemical study of 13 cases. *The American journal of surgical pathology*, 23(5), 530-535.
9. Leroy, X., Zini, L., Buob, D., Ballereau, C., Villers, A., & Aubert, S. (2007). Renal cell carcinoma with rhabdoid features: an aggressive neoplasm with overexpression of p53. *Archives of pathology & laboratory medicine*, 131(1), 102-106.
10. Eble, J. N., Sauter, G., Epstein, J. I., Sesterhenn, I. A. (2004). World Health Organisation classification of tumours. Pathology and genetics of tumours of the urinary system and male genital organs. Lyon: IARC Press, pp. 58–9. (Chapter 1, Rhabdoid tumour).
11. Shannon, B., Wisniewski, Z. S., Bentel, J., & Cohen, R. J. (2002). Adult rhabdoid renal cell carcinoma: divergent differentiation of conventional (clear cell) carcinoma. *Archives of pathology & laboratory medicine*, 126(12), 1506-1510.
12. de Peralta-Venturina, M., Moch, H., Amin, M., Tamboli, P., Hailemariam, S., Mihatsch, M., ... & Amin, M. B. (2001). Sarcomatoid differentiation in renal cell carcinoma: a study of 101 cases. *The American journal of surgical pathology*, 25(3), 275-284.