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Renal malacoplakia: Case report of a differential diagnosis for renal cell carcinoma

Jorge Ayllon[1], Virginie Verkarre[2], Florian Scotté[1], Laure Fournier[3], Jean Michel Corréas[4], Arnaud Mejean[5], Corine Teghom[1], Stéphane Oudard[1]

[1] Service de Cancérologie Médicale, Hôpital Européen Georges Pompidou, Paris, France
[2] Service d'Anatomie Pathologique, Hôpital Necker Enfants Malades, Paris, France
[3] Service de Radiologie, Hôpital Européen Georges Pompidou, Paris, France
[4] Service de Radiologie, Hôpital Necker Enfants Malades, Paris, France
[5] Service d'Urologie, Hôpital Européen Georges Pompidou, Paris, France

Summary

Background: Renal malacoplakia is a very rare chronic inflammatory disorder characterized by specific infiltration of tissue by inflammatory cells, and presents similar radiological characteristics to those of renal cell carcinoma.

Case Report: A 54-year old woman, with a 37-year history of smoking, weight loss, anorexia, asthenia, and night sweats, was included in an antiangiogenesis clinical trial. Clinical signs of inflammation were apparent in the right lumbar region without functional limitations. Previous imagery identified a mass infiltrating the lower pole of the right kidney, extending to the psoas, perinephretic region and ganglia. Biological testing revealed inflammation and a urinary tract infection, treated with ciprofloxacin. Based on histology of a renal puncture biopsy, clear cell carcinoma with oxyphilic cells was suspected but not confirmed by immunohistochemistry.

Urine analysis was positive for *Escherichia Coli*. Computed tomodensitometry revealed three masses (right kidney, between right psoas and the inferior vena cava, and right psoas) and a second puncture biopsy confirmed malacoplakia. After successful antibiotherapy, a right-sided nephrectomy was performed. The patient now shows no evidence of disease.

Conclusions: This case underscores the importance of excluding the differential diagnosis of renal malacoplakia before undertaking partial or total nephrectomy and/or initiating neoadjuvant treatment for renal cell carcinoma.

key words: **malacoplakia • kidney • diagnosis**

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Author's address: Stéphane Oudard, Service de Cancérologie Médicale, Hôpital Européen Georges Pompidou, 20, rue Leblanc, 75015 Paris, France, e-mail: stephane.oudard@egp.aphp.fr

Background

Renal malacoplakia is a very rare chronic inflammatory disorder, which is characterized by specific infiltration of tissue by inflammatory cells as well as unifocal or multifocal lesions [1,2]. As malacoplakia presents radiological characteristics similar to those found in renal cell carcinoma, careful differential diagnostic is warranted to avoid misdiagnosis and unnecessary medical and surgical treatments. Here, we present a case of renal malacoplakia initially misdiagnosed as clear cell carcinoma.

Case Report

A 54-year old woman was referred to us by a urologist in January 2009 for the treatment of a clear cell renal cell carcinoma and for possible inclusion in the Biological, Pathological and Imagery Markers testing in the First-Line Treatment of Metastatic Clear-Cell Renal Cell Carcinoma (PREINSUT) clinical trial, evaluating antiangiogenic treatment of metastatic renal cell carcinoma prior to radical nephrectomy. Patient presented with a 17 kg weight loss (7 kg between September 2008 and January 2009), anorexia, asthenia, and night sweats. The patient had a 37-year history of smoking. Her ECOG performance score was 1 and her Karnofsky score was 80%. Hardening with signs of inflammation in the right lumbar region was associated with pain radiating through the right lower limb upon palpation. The pain was not associated with any functional limitations.

Prior to being referred to us, a conventional abdomino-pelvic computed tomodensitometry exam had been performed in November 2008, in response to hematuria, lumbar pain, and weight loss. A 75 mm diameter mass infiltrating the lower pole of the right kidney was identified. Infiltration of the psoas, perinephretic extension, and non-obstructive dilatation of the pyelocaliceal diverticulum were noted, but no infiltration of the renal vein or the inferior vena cava was apparent. A positron emission tomography scan revealed an additional infiltration of the ganglia with limited dissemination. Biological testing revealed a urinary tract infection, an inflammatory syndrome with an erythrocyte sedimentation rate >100 mm in the first hour, and a C-reactive protein level of 130 mg/L. The urinary tract infection was treated with ciprofloxacin. In December 2008, a renal puncture biopsy was performed. Clear cell carcinoma with oxyphilic cells was suspected based on morphological analysis of histology; however, this diagnosis could not be confirmed by immunohistochemistry. The patient developed fever up to 38.5°C after the biopsy.

Our January 2009 evaluation was performed in the context of inclusion in the PREINSUT clinical trial. Biological testing revealed an inflammatory syndrome with a C-reactive protein concentration of 191 mg/L, a hemoglobin concentration of 9.3 g/dL, leucocytosis (20,000 leucocytes/mm^3), thrombocytosis (756,000 platelets/mm^3), a lactate dehydrogenase level of 115 UI/L, and a calcium level of 2.28 mg/dL. Urine analysis was positive for *Escherichia Coli*. Computed tomodensitometry revealed three masses: one tumor in the lower pole of the right kidney measuring 83 mm along its long axis, a second tumor located between the right psoas muscle and the inferior vena cava and measuring 41 mm in diameter, and a third heterogeneous mass involving the right psoas, measuring 52 mm in diameter, and extending to the posterior right soft tissue.

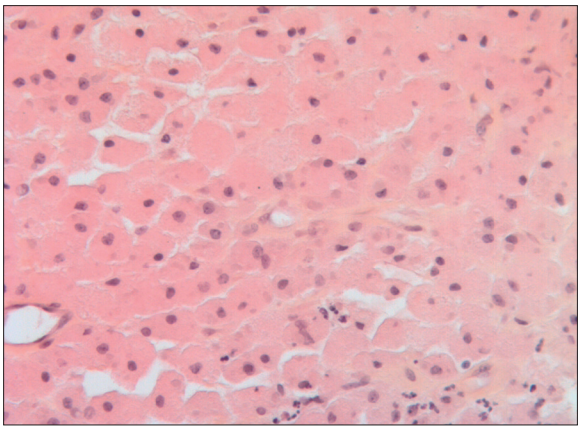

Figure 1. Renal biopsy. Diffuse granulomatous inflammatory infiltrate that includes neutrophils and large numbers of eosinophilic histiocytes containing the typical intracytoplasmic Michaelis-Gutmann bodies (HES ×200).

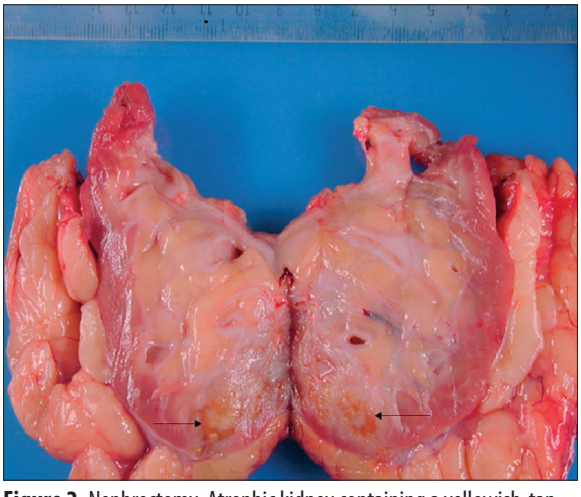

Figure 2. Nephrectomy. Atrophic kidney containing a yellowish-tan tissue nodule (arrows) at the lower pole.

Malacoplakia was diagnosed after a second puncture biopsy of the renal tumor was performed on January 9, 2009 (Figure 1). No tumor-associated proliferation was visible. On January 10, 2009, in response to the persisting fever, back pain, and the presence of an inflammatory placard on the skin, an ultrasound was performed revealing fluid from the kidney infiltrating the posterior muscular wall and creating a fistula. A dilatation of the pyelocaliceal cavities was also noted. Collecting fluid was drained and a double J stent was inserted. Urine analysis was positive for *E. coli*. Fever resolved after treatment with ceftriaxone and ofloxacin [9]. An additional year of treatment with sulfamethoxazole 800 mg/trimethoprim 160 mg bid was prescribed according to international recommendations [3,4], with succesful clinical outcome. In December 2009, renal scintigraphy revealed a non-functioning right kidney. A right-sided nephrectomy was performed on May 28, 2010 without any complications. An atrophic kidney containing a yellowish-tan tissue nodule at the lower pole was removed (Figure 2). At present, the patient is seen regularly for follow up and she has no evidence of disease.

Discussion

Renal malacoplakia, a rare chronic inflammatory disease in which unifocal or multifocal renal masses are often associated with an *E. coli* infection, can easily be confused with renal cell carcinoma as radiologic images are non-specific [1,5]. In most renal malacoplakia cases, imaging reveals enlarged kidneys, poorly defined, hypoechoic, solid masses, or heterogeneous masses with central areas of necrosis. Diagnosis of renal malacoplakia thus depends upon the histological confirmation of massive tissue infiltration by inflammatory cells and in particular on the presence of histiocytic cells containing intracytoplasmic Michaelis-Gutmann bodies [2].

Although the clinical presentation of malacoplakia varies greatly depending on the affected organs, the symptoms presented herein are consistent with those described in the literature [2]. This patient did not have any medical antecedent or condition predisposing to renal malacoplakia. The three renal masses were associated with pain, hematuria, fever, an inflammatory syndrome, and an *E. coli* urinary tract infection. Indeed, *E. coli* is the most frequently (70% of cases) identified bacteria in malacoplakia-associated urinary tract infections, though other infections with *Klebsiella granulomatis*, *Corynebacterium*, mycobacteria, and staphylococci have also been described [2,3,6].

Both pharmacologic and surgical treatments are possible [4,7,8]. For patients with multifocal disease, antibiotic treatment may be favored, whereas as for patients with unifocal disease, surgical treatment is more likely to be indicated. Among antibiotics, quinolones as well as cotrimoxazole, rifampin, doxycycline, trimethoprim, and vancomycin have been shown to be effective [9,10]. In the present case of multifocal renal malacoplakia, short term treatment with ceftriaxone and ofloxacin followed by long term treatment with sulfamethoxazole/trimethoprim resulted in successful resolution. A nephrectomy was nonetheless necessary one year later, due to the massive destruction of the right kidney.

Conclusions

This case report underscores the importance of excluding the differential diagnosis of renal malacoplakia before performing partial or total nephrectomy and/or initiating neoadjuvant treatment for renal cell carcinoma. In the current case, the differential diagnosis of malacoplakia resulted in treatment with antibiotics rather than an unwarranted antiangiogenic therapy.

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Additional statement

The patient signed informed consent for participating in the PREINSUT clinical trial; this consent includes authorizing publication of the case report.

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