CASUISTRY

Collecting duct renal cell carcinoma


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KEYWORDS
Kidney; Tumor; Collecting duct; Bellini

Abstract
Introduction: Bellini’s renal cell collecting duct carcinoma is a rarely prevalent renal tumor, with low cancer-specific survival, although its rate of response to antiangiogenic therapies is unknown.
Objectives: We retrospectively revise a series of collecting duct tumors, with special emphasis on the indication of target therapies and on their results.
Materials and methods: Retrospective analysis of renal cell collecting duct carcinoma treated at our institution from January 2000 to June 2010, taking into account the patient’s age, sex, reason for the consultation, oncological background, side of the affection, surgical treatment, other anatomopathological characteristics, tumor size, TNM clinical staging (2009), adjuvant treatment and survival time.
Results: Six patients are described, five men and one woman, with a mean age of 75 (±7.7) years. Four of them (66.6%) presented disseminated disease upon diagnosis. Five (83%) were treated with radical nephrectomy and three (50%) received systemic adjuvant treatment, without response. The means survival was 5.5 months (4.75-14.75). Only 2 patients (33.3%), both with localized disease upon diagnosis, are in complete remission.
Conclusion: Renal cell collecting duct carcinoma is a disease with a bad prognosis, little survival and bad response to target therapies.

PALABRAS CLAVE
Rinón; Tumor; Conductos colectores; Bellini

Carcinoma de células renales del túubo colector

Resumen
Introducción: El carcinoma renal de los conductos colectores de Bellini es un tumor renal escasamente prevalente, con baja supervivencia cáncer-específica, aunque realmente se desconoce su tasa de respuesta a terapias antiangiogénicas.
Objetivos: Se revisa de manera retrospectiva una serie de tumores de túubo colector con especial énfasis en la indicación de terapias diana y en los resultados de la misma.

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Material y métodos: Análisis retrospectivo del carcinoma renal de túbulo colector tratados en nuestra institución desde enero 2000 a junio 2010, teniendo en cuenta la edad del paciente, el sexo, el motivo de consulta, los antecedentes oncológicos, el lado de afectación, el tratamiento quirúrgico, otras características anatomopatológicas, el tamaño tumoral, la estadificación TNM (2009), el tratamiento adyuvante y el tiempo de supervivencia.

Resultados: Se describen 6 pacientes, 5 varones y una mujer; con una media de edad de 75 (± 7,7) años. Cuatro de ellos (66,6%) presentaban enfermedad diseminada al diagnóstico. Cinco (83,3%) fueron tratados mediante nefrectomía radical y tres (50%) recibieron tratamiento sistémico adyuvante, sin respuesta. La mediana de supervivencia fue 5,5 meses (4,75-14,75). Sólo dos pacientes (33,3%), ambos con enfermedad localizada al diagnóstico, se encuentran en remisión completa.

Conclusión: El carcinoma renal de túbulo colector es una enfermedad con mal pronóstico, escasa supervivencia y mala respuesta a terapias diana.

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Introduction

Collecting duct renal carcinoma, also called Bellini’s tumor, is the least prevalent subtype of renal carcinoma. Except for three major series published in recent years, only isolated cases have been reported. The collecting duct carcinoma is characterized by being an aggressive entity with an unfortunate outcome in most patients. In our country, only isolated cases or case series have been published. The aim of this study is to evaluate our own case series, with emphasis on the analysis of the response to systemic therapy and, in particular, to new target therapies.

Materials and methods

We revised all renal tumors diagnosed in our center from January 2000 to June 2010, and identified cases of collecting duct renal carcinoma. The variables analyzed were: age, sex, reason for consultation, oncological background, side of the affection, surgical treatment, pathological characteristics, tumor size, TNM staging (rating 2009), adjuvant treatment and survival time.

A descriptive statistical analysis was performed using Microsoft Office Excel 2003®. For the analysis of quantitative variables the mean with the standard deviation and the median with the interquartile range were used. In the analysis of qualitative variables, we used the percentage. An inferential statistical analysis was not performed due to low sample size.

Results

Of a total of 670 renal tumors, 6 (0.89%) were collecting duct renal cell carcinoma. Table 1 summarizes the characteristics of these patients. The mean age was 75 (±7.71) years. The male:female ratio was 5:1. The average size of tumor, measured as the maximum diameter of the macroscopic piece for tumors that were operated, or with computerized axial tomography (CT)/nuclear magnetic resonance (NMR) for the rest, was 65.5 mm (±39.6) (Figs. 1 and 2). Five patients (83%) were treated with radical nephrectomy. In one patient (16.6%) diagnosis was made by percutaneous biopsy. Four of the 6 cases (66.6%) had lymphatic and/or hematogenous dissemination at diagnosis. Only two (33.3%) presented localized disease, which are those that are free of recurrence and alive today, with a follow-up of 120 and 17 months, respectively. The remaining patients died of progression of their disease, with a median survival of 5.5 (4.75–14.75) months.

Of the 4 patients with lymphatic and/or hematogenous dissemination, three (75%) received adjuvant systemic therapy to surgery. Two received immunotherapy (sunitinib and temsirolimus) and a third patient received a conventional chemotherapy scheme based on cisplatin and gemcitabine. None of them presented remission of the adenopathy and/or metastasis in the follow-up performed with CT. Surgery on the metastasis was not carried out either.

Discussion

The collecting duct renal carcinoma is a rare entity that has traditionally presented an ominous oncologic prognosis. Our
Table 1
Clinicopathologic features of patients with collecting duct renal cell carcinoma.

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Sex</th>
<th>Size (maximum diameter)</th>
<th>TNM</th>
<th>Pathological anatomy</th>
<th>Adjuvant treatment</th>
<th>Surgical treatment</th>
<th>Reason for consultation</th>
<th>Survival (months)</th>
<th>Adjuvant treatment status</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>77</td>
<td>Man</td>
<td>4.5 cm</td>
<td>T1N0M0</td>
<td>Pure Bellini</td>
<td>None</td>
<td>Radical nephrectomy</td>
<td>Incidental finding</td>
<td>Temsirolimus</td>
<td>17 months - CR</td>
</tr>
<tr>
<td>2</td>
<td>72</td>
<td>Woman</td>
<td>3.5 cm</td>
<td>T1N0M2</td>
<td>Sarcomatoid type Bellini associated with clear cell carcinoma (Fuhrman grade 3)</td>
<td>Temsirolimus</td>
<td>Radical nephrectomy</td>
<td>Study of cerebellar metastases</td>
<td>8 months - CR</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>68</td>
<td>Man</td>
<td>13 cm</td>
<td>T3bN2M1</td>
<td>Tubulopapillary Bellini</td>
<td>Carboplatin and gemcitabine</td>
<td>Radical nephrectomy</td>
<td>Macroscopic hemorrhata Constitutional syndrome</td>
<td>6 months - DD</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>86</td>
<td>Man</td>
<td>11.5 cm</td>
<td>T4hN1M2</td>
<td>Microscopic hemorrhata</td>
<td>None</td>
<td>Radical nephrectomy</td>
<td>None</td>
<td>2 months - DD</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>79</td>
<td>Man</td>
<td>5 cm</td>
<td>T3bN2M1</td>
<td>Microscopic hemorrhata</td>
<td>Sunitinib</td>
<td>Radical nephrectomy</td>
<td>Incidental finding</td>
<td>7 months - DD</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>69</td>
<td>Man</td>
<td>5 cm</td>
<td>T2N0M0</td>
<td>Pure Bellini</td>
<td>None</td>
<td>Radical nephrectomy</td>
<td>None</td>
<td>120 months - CR</td>
<td></td>
</tr>
</tbody>
</table>

DD: died of disease; CR: complete remission.

Figure 2 NMR radiological imaging of localized collecting duct renal carcinoma.

experience with target therapies and chemotherapy is very limited, but it also confirms its ominous prognosis without clinical response.

This type of renal tumor develops from the collecting tubules of the renal medulla, thus, presenting an unusual and distinctive histology (Fig. 3). There are no specific radiological features of this tumor. From the pathological point of view, there are a number of microscopic features that point towards its diagnosis: commitment of the medullary pyramids, irregular tubular architecture, marked desmoplasia and the presence of high degree tack cells. Immunohistochemistry is usually positive for Ulex europeus, high molecular weight cytokeratin (CK19, CK7, CK8/18) Fez1, mucin, lysozyme and lectins/peanut agglutinina.

There are three series published with a significant number of cases. They all observed a predominance of males. The age at diagnosis of this neoplasm is usually from the fifth decade of life, although there are also cases in young patients. The mean size at diagnosis varies depending on the series, but it ranges between 6 and 8 cm. At diagnosis most patients have locally advanced disease with lymph node
and/or metastatic disease. All these data are consistent with our cases, although the age at diagnosis we observed tends to be higher.

The treatment that was performed in most patients described above was radical nephrectomy, even in those with disseminated disease. However, the benefit of cytoreductive nephrectomy has not been objectified in patients with metastatic collecting duct renal carcinoma either. Survival described long term is low and is around 0–58% at three years. In our experience, only patients with localized disease at diagnosis, lack of lymph node or hematogenous disease are free of recurrence. Two studies compared cancer-specific survival of this neoplasm with clear cell renal carcinoma. In the series of Karakiewicz et al. no differences in survival are found if adjusted for stage. Nevertheless, this finding may be due to a low number of cases (47 patients) and the inclusion of slants in choosing the comparison cohort because it includes cases treated in the 80s. In the series of Wright et al. a comparison between cases diagnosed in the same period is performed, all from 2000, finding statistically significant difference in survival in favor of conventional renal cell carcinoma.

None of the large series evaluates the response to adjuvant treatments with chemotherapy, immunotherapy or antiangiogenic therapy. From a pathological point of view, there is some histopathogenetic proximity between collecting duct carcinoma and carcinoma of the upper urothelium. That is why, some authors have proposed therapeutic regimens based on carboplatin-gemcitabine, doxorubicin, gemcitabine and paclitaxel-carboplatin. In our series, patients treated systemically have not shown a good response to adjuvant treatment with temsirolimus, sunitinib or cisplatin and gemcitabine.

In summary, collecting duct renal cell carcinoma is a rare renal tumor. In our series, most patients are in a disseminated or locally advanced stage at diagnosis. The rate of long-term survival is low because the only potentially curative treatment appears to be surgery if it is considered in patients with localized tumors.

Conflict of interest

The authors declare that they have no conflict of interest.

References