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Archivos Españoles de Urología, vol. 66, núm. 6, julio-agosto, 2013, pp. 597-601
Editorial Iniestares S.A.
Madrid, España

Available in: http://www.redalyc.org/articulo.oa?id=181043651009
Case Reports

RENAL CELL CARCINOMA OF THE COLLECTING DUCTS OF BELLINI WITH RETROPERITONEAL RECURRENCE BY ANOTHER HISTOLOGICAL SUBTYPE OF RENAL TUMOR

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Summary.- OBJECTIVE: Collecting Duct Carcinoma or Bellini Carcinoma (CDC) is a rare aggressive histological subtype. We present a case of CDC with retroperitoneal recurrence by another histological subtype of renal tumor and review of the literature.

METHODS: A 59-year-old man with no relevant clinical history presented gross hematuria. At the time of diagnosis, a computed tomography (CT) showed a tumor mass occupying the left renal pelvis. Left Laparoscopic radical nephroureterectomy was performed with endoscopic intramural ipsilateral ureter disinsertion.

RESULTS: The pathological diagnosis was CDC with negative surgical margins. A CT scan control was performed 10 months later, showed a left retroperitoneal tumor compatible with a local recurrence. We performed a left subcostal laparotomy with complete resection of the mass. Histological diagnosis was large cell carcinoma with components of granular cells and clear cell.

CONCLUSIONS: The CDC is a rare subtype of renal cell carcinoma (RCC) and has an aggressive behavior that is associated with poor prognosis. Surgical resection remains the treatment of choice. We present the first reported case of CDC with retroperitoneal recurrence by another histological subtype of renal tumor.

Keywords: Renal cell carcinoma. Collecting Duct Renal Carcinoma. Bellini Tumor. Radical nephroureterectomy. Laparoscopy.

Resumen.- OBJETIVO: El Carcinoma Renal de los Conductos Colectores (CRCC) o Carcinoma de Bellini es un subtipo histológico raro y agresivo. Presentamos un caso de CRCC con recidiva retroperitoneal por otro subtipo histológico de tumor renal y revisión de la literatura.

MÉTODOS: Paciente masculino de 59 años, sin antecedentes médicos de importancia, quien consultó por presentar hematuria macroscópica. Se realizó Tomografía Computada de abdomen (TAC) la cual mostró una masa tumoral que ocupa la pelvis renal izquierda. Se practicó nefroureterectomía radical izquierda laparoscópica con desinserción endoscópica de uréter intramural ipsilateral.

RESULTADOS: El diagnóstico anatomopatológico fue CRCC con márgenes quirúrgicos negativos. Se realizó una TAC control 10 meses después, la cual reveló una masa tumoral retroperitoneal izquierda, compatible con una recidiva local. Se realizó una laparatomía subcostal izquierda con resección completa de la masa. El diagnóstico histológico fue un carcinoma renal de células grandes con componentes de células granulares y células claras.

CONCLUSIONES: El CRCC es una forma poco frecuente de todos los carcinomas renales y presenta un comportamiento agresivo que se asocia a mal pronóstico. La resección quirúrgica sigue siendo el tratamiento de elección. Presentamos el primer caso descrito de CRCC con recidiva retroperitoneal por otro subtipo histológico de tumor renal.

INTRODUCTION

Collecting Duct Carcinoma (CDC) or Bellini Carcinoma is a rare, highly aggressive malignant neoplasm that arises from the collecting duct epithelium of the kidney, representing 0.4% to 1.8% of all renal cell carcinomas (RCC) (1). Unlike most of RCC that occur in the epithelial cells of proximal tubules, the CDC is originate in the distal collecting ducts. Macroscopically located near the central or pelvic region (2).

Most CDC show tubular or papillary patterns surrounded by desmoplastic stroma. CDC papillary appearance can sometimes make it difficult to distinguish from papillary renal cell carcinoma, immunohistochemical studies are needed to make the definitive diagnosis (3,4). Typically CDC react positively to the high molecular weight cytokeratins that are expressed frequently in distal tubule cells.

Clinical data suggest that the CDC has a tendency to affect younger patients, occur in advanced stages and associated with poor prognosis. The CDC has been reported to coexist in the kidney with other tumors such as RCC, transitional cell carcinoma and oncocitoma (5). To our knowledge, we present the first reported case of CDC with retroperitoneal recurrence by another histological subtype.

CASE REPORT

A 59-year-old man with no relevant clinical history presented gross hematuria. At the time of diagnosis, a computed tomography (CT) showed a tumor mass occupying the left renal pelvis (Figure 1).

Left Laparoscopic radical nephroureterectomy was performed with endoscopic intramural ipsilateral ureter desinsertion for the suspicion of an upper urothelial tumour, with 120 minutes operating time and minimal bleeding. The patient was discharged 48 hours after surgery.

The biopsy showed a kidney weighs 170 grams and measuring 11 x 6 x 5 cm. It showed a tumor of 2 x 2 cm on the lower calyx, which was occupied and extends into the renal parenchyma and perirenal adipose tissue forming an exophytic mass with negative surgical margins for tumor (Figure 2).

Histological examination of the tumor showed papillary and tubular patterns with highly atypical cells and nuclei with “hobnail” appearance (Figure 3). Immunohistochemically, nearly 100% of tumor cells were positive for high-molecular weight 19-cytokeratin with staining of renal tubules, with focal positivity for 20-cytokeratin and 7-cytokeratin. These findings were compatible with CDC.

A CT scan control was performed 10 months later, showed a 12 cm left retroperitoneal lesion compatible with a local recurrence (Figure 4). We performed a left subcostal laparotomy with complete resection of the mass, there was no infiltration of adjacent organs.

The analysis of the surgical specimen showed an ovoid tumor mass of 13.5 x 9.5 x 8.5 cm, weight of 639 grams. Histological diagnosis was large cell carcinoma with components of granular cells and clear cell, grade III - Fuhrman IV.

Immunohistochemistry was positive for vimentin and CD-10 in tumor cells and negative for CK-7, CK-20, inhibin and carciñoembryonic antigen, excluding tubular origin. Surgical margins were negative. The patient is asymptomatic and free of disease at 3 years of follow-up.

Figure 1. CT Scan showing a central tumor in the left kidney.

Figure 2. Macroscopic view of the tumor after opening of the renal pelvis
DISCUSSION

The CDC is a rare type of RCC, representing 0.4% to 1.8% of all RCC. The CDC generally affects young people and has an aggressive behavior; is also a highly rare tumor occurrence. The clinical presentation is usually in the form of hematuria and lumbar pain as the most frequent manifestations (1). On the other side, patients with RCC are typically in the range of age 60, males predominate over females with a ratio 3:1, about 75% of cases occur in pathological stage pT2-pT3 and over 80% have nuclear grade III-IV (1,6).

Unlike most renal cell carcinomas that occur in the epithelial cells of proximal tubules, the CDC originates in the distal collecting duct (2). The embryological origin of these tumors is different from other renal parenchymal tumors. The ureters, pelvis, calyces and collecting tubules originate from the ureteric bud. Moreover, the metanephric blastema is responsible for the development of convoluted tubules. This explains the clinical, radiographic, macroscopic, microscopic, immunohistochemical and cytogenetic characteristics of the CDC (7).

The CRCC has distinctive macroscopic and microscopic features that make it different to any of the common renal carcinomas. They have established criteria for diagnosis (Table I) (2). The low incidence of CDC has made it difficult to characterize the disease. Although the macroscopic characteristics of the tumor are well established, always diagnostic confusion can occur (2-4). Immunohistochemistry is a widely used tool for the diagnosis of the CDC, because the cells of the distal and proximal nephron react differently to lectins and antibodies (8).

However despite the fact that most of the expression of high molecular weight cytokeratins occurs in the distal nephron cells and expression of cytokeratins of low molecular weight occurs in the proximal nephron, the results of immunohistochemistry for CDC not are always consistent (3,9).

Cytogenetic studies have shown that the tumor contains a diverse range of chromosomal abnormalities, unrelated to other variants of renal cell carcinoma (10). The CDC has not shown a consistent association with loss of heterozygosity on chromosome 3p, typical of renal cell carcinoma or trisomy of chromosomes 7, 12, 16, 17 and 20, typical of papillary renal carcinoma (9). Currently accumulating data on genetic analysis of CDC, only serves to demonstrate that it is a distinct entity in the categorization of renal cell carcinomas. Perhaps a better understanding of the genetic alterations associated with this tumor will help explain why most of the CDC act as biological aggressiveness and only a small subset of patients appears to follow a relatively benign course. CDC management has been limited primarily to surgical resection.

In reviewing Tokuda 44.2% of patients had metastatic lymph nodes, systemic dissemination 32%, 17% lung metastases and 16% bone. Karakiewicz et al. described 41 cases in which 48.8% had metastatic lymph nodes and 19.5% metastases distance (1,6).

These tumors are fast-growing invasive systemic dissemination. In the revised series have used multiple chemotherapy regimens and / or immunotherapy for the management of the CDC and most of these studies shows that these therapies only provide very limited benefit in a select group of patients (9).
There are few cases where the CDC coexist in the kidney with other tumors. To our knowledge, we present the first reported case of retroperitoneal recurrence of CDC by other histological subtype of renal tumor.

CONCLUSIONS

The CDC is a rare subtype of renal cell carcinoma (RCC), it represents 0.4% to 0.8% of them and has an aggressive behavior that is associated with poor prognosis. The diagnosis is based on major and minor histological criteria, the most important are: hobnail cells, marked desmoplasia and reacts positively to antibodies against high molecular weight keratin and Ulex europaeus. Surgical resection remains the treatment of choice. We present the first reported case of CDC with retroperitoneal recurrence by another histological subtype of renal tumor.

REFERENCE AND RECOMMENDED READINGS

(*of special interest, **of outstanding interest)

RENAL ALLOGRAFT RUPTURE: CASE REPORT AND BIBLIOGRAPHIC REVIEW

Juan Francisco Galiano Baena, Enrique Herrero Polo, Juan Pablo Caballero Romeu, Carla Perez Tomas, Antonio Miguel Pelluch Auladell and Juan Jose Lobato Encinas.


Summary.- OBJECTIVE: To improve the knowledge about complications of renal transplantation and, in particular, graft rupture.

METHODS: Case report and literature review.

OUTCOME: We present the case of a 37 year-old patient receiving a second renal transplant. In the third postoperative day, he suffered an abrupt change from the correct evolution, with intense pain in the left iliac fossa (the side of the implant) and hemodynamic instability. Imaging