Sarcomatoid chromophobe renal cell carcinoma: a rare entity


ABSTRACT

Introduction: Chromophobe renal cell carcinoma is a subtype of renal cell carcinoma that is known for its good prognosis. Sarcomatoid transformation in this subtype is rare. Of all renal cell carcinomas, chromophobe renal cell carcinoma has an overall incidence of 4-5%. It arises from the collecting tubule and characteristically stains positive for Hale’s colloidal iron.

Clinical case: Patient is a 73-year-old man with past history of high blood pressure. He presented with two-month progression of abdominal pain predominantly in right hypochondrium and did not complain of urinary symptomatology. Patient was diagnosed with chronic lithic cholecystitis for which laparoscopic cholecystectomy was carried out that revealed increased retroperitoneal volume. Computed tomography scan showed right kidney tumor with probable thrombus in inferior vena cava. Nuclear magnetic resonance image revealed adenopathy compressing inferior vena cava with no thrombus. Open right radical nephrectomy and lymphadenectomy were performed, obtaining 24 x 13 x 5 cm tumor that weighed 1080 g with 17 x 17 x 2.5 cm adenopathies.

RESUMEN

Introducción: El carcinoma de células renales cromóbico (CCRC) es un subtipo del carcinoma de células renales (CCR), conocido por su buen pronóstico. La transformación sarcomatoide en este subtipo de tumor es rara. El CCRC tiene una incidencia global de 4-5%, de todos los CCR. Se origina del túbulo colector y se caracteriza por tener positivo al hierro coloidal de Hale.

Caso clínico: Masculino de 73 años, con historia de hipertensión arterial. Cuadro clínico de dos meses de evolución, consistente en dolor abdominal de predominio en el hi pocondrio derecho, negando sintomatología urinaria, diagnosticado con colecistitis crónica, por lo que se realizó colecistectomía laparoscópica y luego se encontró aumento de volumen del retroperitoneo. Se realizó tomografía axial computada (TAC), que mostró tumor renal derecho con probable trombo en vena cava inferior (VCI). En la resonancia magnética (RM) se encontraron adenopatías, que comprimían la VCI sin trombo. Se realizó nefrectomía derecha abierta y linfadenectomía. Se obtuvo tumor de 1080 g de 24 x 13 x 5 cm, con adenopatías de 17 x 17 x 2.5 cm con tres nódulos.
cm adenopathy with three nodules. Chromophobe renal cell carcinoma was reported with 80% sarcomatoid differentiation and lymph node invasion. Chest computed tomography scan revealed pulmonary metastases. Tumor was classified as clinical stage IV, pT4N1M1.

**Discussion:** Sarcomatoid transformation in chromophobe renal cell carcinoma is extremely rare. Sarcomatoid differentiation, microscopic necrosis, small vessel invasion, and tumor size have been established as poor prognosis factors. When there is sarcomatoid histology, survival is less than one year.

**Keywords:** Sarcomatoid differentiation, chromophobe renal cell carcinoma, Mexico.

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**INTRODUCTION**

Renal cell carcinoma (RCC) is a relatively rare neoplasia that corresponds to approximately 3% of malignant tumors. Close to 200,000 new cases of RCC are diagnosed per year worldwide and the number of annual deaths reaches 100,000. Between 70-90% of RCC cases can be cured in patients with clinical stage I of the Tumor, Regional Lymph Nodes, Distant Metastasis (TNM) staging system, 55-70% in stage II, 20-30% in stage III, and less than 10% in stage IV. The World Health Organization (WHO) recognizes numerous RCC subtypes, the most common of which are: clear cell RCC (70%), papillary RCC (10-15%), chromophobe RCC (4-6%), and collecting duct carcinoma (1%).

Chromophobe renal cell carcinoma (CRCC) is generally diagnosed in the sixth decade of life. Its incidence is the same in men and in women and 86% of CRCC is diagnosed in stage I and stage II. Renal vein invasion presents alone in fewer than 5% of RCC cases. Metastases are found in 6% of patients and the most frequent sites are the liver (39%) and the lung (36%).

Chromophobe renal cell carcinoma (CRCC) is a tumor with a 5-year survival rate above 83%. Mean time from nephrectomy to metastasis is double that of other types of RCC. Sarcomatoid differentiation is found in 1.9% of CRCC. Tumor size, vascular invasion, sarcomatoid histology, and necrosis are associated with poorer survival, and is less than 1 year in the majority of series.

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**CASE PRESENTATION**

Patient is a 73-year-old man with genetic burden for high blood pressure and history of high blood pressure of 10-year progression presently treated with nifedipine. Patient came to emergency room with nonradiating, oppressive abdominal pain of 2-month progression located in the right hypochondrium, with an intensity of 9/10 on the Visual Analog Pain Scale (VAS), that was not related to food intake, was not modified with changes of position, and that partially ceded with the use of nonsteroidal anti-inflammatory drugs (NSAIDs). He complained of 10 kg weight loss in two months that was not related to any type of diet. Patient was diagnosed in the emergency room with chronic lithiasic cholecystitis and underwent laparoscopic cholecystectomy that was completed with no complications. Surgery revealed increased retroperitoneal volume behind the liver. During same hospitalization, computed axial tomography (CAT) scan identified a large right kidney tumor dependent on the upper pole and image suggested the presence of tumor thrombus in the inferior vena cava (IVC) and precaval lymph node conglomerate. Nuclear magnetic resonance (NMR) imaging study of the abdomen confirmed right kidney tumor diagnosis, and thrombus in the vena cava was ruled out. A lymph node conglomerate was observed that compressed anterior and posterior sides of the vena cava. Open right radical nephrectomy and lymphadenectomy were performed. Precaval and paracaval lymph node conglomerate was found firmly attached to the vena
cava, along with a 15 cm kidney tumor with firm adherences to the liver and retroperitoneum. Surgery was completed with no complications. There were no complications in the postoperative period and patient was released on the fourth day with no drains or catheters.

A 24 x 13 x 5 cm tumor weighing 1080 g was removed. The upper and middle poles were substituted by 13 x 6 cm tumor with 17 x 17 x 2.5 cm precaval tumor with three nodules, the largest measuring 2 cm and the smallest 0.5 cm. There was CRCC with sarcomatoid differentiation in 80%, Fuhrman grade 4, tumor on the kidney sinus, tumor in ureteral adventitia at surgical margin, extensive lymphatic permeation in the renal parenchyma along with perinephric adipose tissue, and adrenal gland with recent hemorrhage and necrosis. Lymph nodes were invaded by tumor and staging was pT4N1M0 (Figures 3-5).

Chest CAT scan was done during follow-up in which images compatible with metastasis in right pulmonary base and left middle lobe were found. Patient’s final stage was T4N1M1, clinical stage IV (Figure 6).

Figure 1. Abdominal CAT scan in arterial phase showing tumor at upper pole of right kidney. Image suggests thrombus in vena cava.

Figure 2. Phase T2 NMR image showing extrinsic compression of inferior vena cava from tumor with no evidence of tumor thrombus.

Figure 3. Fusiform pattern characteristic of sarcomatoid pattern surrounded by chromophobe cells (Hematoxylin and Eosin [H & E] stain, 100x).

Figure 4. Chromophobe renal cell carcinoma (H&E stain, 100x).
Sarcomatoid differentiation in CRCC is extremely rare. In a published report by Parada et al. in 2006, there were only 16 cases reported worldwide. The medical literature is also controversial, in that some authors report that the most common sarcomatoid transformation is associated with clear cell tumors and others describe a connection between sarcomatoid presentation and chromophobe tumors. In the present review there was a 1.2.2 man-to-woman ratio with mean age of 60.3 years at the time of diagnosis. Principal symptoms were pain and hematuria and 75% of cases involved the right kidney and 25% of cases involved the left. Tumor size varied between 3-18 cm (mean 9.97 cm), a higher mean than that of classic CRCC. Sixteen patients presented with local dissemination (73.3%) and 5 cases presented with distant metastasis (33.3%). In contrast, 96.6% of classic CRCC cases are T1-T2. In several studies the following poor prognosis factors have been established: sarcomatoid differentiation, microscopic necrosis, small vessel vascular invasion, and tumor size. There is no relation between pT and nodal metastasis. The patient described here presented with numerous poor prognosis factors such as sarcomatoid differentiation, vascular invasion, and tumor larger than 13 cm, placing him in a high risk group with survival prognosis of less than 1 year.

REFERENCES