CLINICAL CASE

Chromophobe type kidney tumor and uterine myomatosis in a solitary kidney patient


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KEYWORDS
Renal cell carcinoma; Chromophobe renal cell carcinoma; Partial nephrectomy; Outcome; Live donor; Kidney transplant donor care; Mexico.

Abstract  Kidney cancer continues to be a surgical disease. Chromophobe type kidney tumors have a better outcome than other histologic variants and usually present sporadically or are associated with familial syndromes. Partial nephrectomy is indicated when the total loss of kidney function is foreseen. A 45-year-old woman had a past history of left nephrectomy due to kidney donation and she had no surveillance of the remaining kidney. The patient sought medical attention for an abdominal mass and predominantly nocturnal pruritus. An abdominal tomography scan revealed tumors in the right kidney and uterus. Laboratory tests reported normal kidney function, negative tumor markers for ovarian cancer, and altered liver function. Hysterectomy and right heminephrectomy with 19 minutes of warm ischemia were performed. The kidney parenchyma was closed with an adipose tissue patch and a ureteral stent was placed. Liver function normalized after the nephrectomy. The histopathologic study reported chromophobe renal cell carcinoma and uterine leiomyomatosis. A long-term follow-up period is recommended for patients with a solitary kidney. This patient did not present with a hereditary kidney cancer syndrome, but nevertheless, treatment should strive to spare the greatest number of nephrons, regardless of the histologic strain.

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Chromophobe tumor in a solitary kidney

Introduction

Clear cell carcinoma represents more than 80% of the renal tumors, followed by papillary carcinoma and chromophobe cell carcinoma. The majority of chromophobe type kidney tumors are sporadic, but occasionally they are associated with Birt-Hogg-Dubé syndrome and they are considered to have a better outcome than clear cell carcinoma. The differential diagnosis of chromophobe type tumors is not easy, and can require the use of diagnostic aids such as immunohistochemistry studies.

The same as tumors of the prostate, colon, or breast, kidney cancer can present sporadically or hereditarily. Various types of hereditary kidney cancer have been described. In 2001 a kidney cancer familial syndrome was described in which the patients developed cutaneous and uterine leiomyomas and type 2 papillary renal cell carcinoma.

Stauffer syndrome is a paraneoplastic syndrome characterized by non-metastatic liver dysfunction that normalizes after nephrectomy and has been reported in 3%-20% of the patients with kidney cancer.

Treatment of renal tumors is primarily surgical. The goal is to remove the entire tumor with sufficient surgical margin. When the patient is at a high risk for dialysis or when nephrectomy implies leaving the patient anephric, partial nephrectomy is indicated.

There are no convincing data indicating that living donors are at greater long-term risk for developing chronic nephropathy due to kidney donation. Nevertheless, long-term periodic follow-up evaluation is recommended for donors. This can be carried out by the donor's personal physician.

The aim of this study was to show the therapeutic behavior adopted for treating a patient with kidney cancer in a solitary kidney and its complete accordance with that reported in the literature. There were not enough clinical or paraclinical data to support a hereditary kidney cancer syndrome. Rather, this was a patient with a sporadic chromophobe type renal tumor associated incidentally with uterine myomatosis that presented with Stauffer syndrome.

Case presentation

A 45-year-old woman, whose mother presented with breast cancer and synchronous kidneys, had a past history of left nephrectomy due to kidney donation 6 years before, 1 pregnancy, and 1 birth. She had never had a Papanicolaou test done nor had she received follow-up or monitoring of her solitary kidney.

She was referred to our service because of an incidental finding of an abdominal tumor and a right kidney tumor in an abdominal ultrasound that had been ordered to resume surveillance of the solitary kidney. In addition, the patient had presented with predominantly nocturnal, generalized pruritus and a painless, palpable mass in the lower abdomen with no accompanying symptoms 2 years before. During physical examination a mobile, painless, abdominal mass was palpated in the hypogastrum, with no other alterations. An abdominal computerized tomography (CT) scan was done that revealed a TNM classification stage T2b right kidney tumor and an abdominal tumor that was dependent on the uterus (figs. 1 and 2). The chest x-ray was normal. The laboratory tests reported urea 24 mg/dL, creatinine 0.82 mg/dL, liver profile with total bilirubin 0.4 mg/dL, alkaline phosphatase 1140 U/L, AST 210 U/L, ALT 185 U/L, GGT 751 U/L, Ca 1-25 46.25 U/mL, Ca 19-9 7.34 U/mL, carcinoembryonic antigen 1.16 ng/mL, and alpha fetoprotein 3.08 U/mL.

Exploratory laparotomy revealed a tumor that was dependent on the lower pole of the right kidney that measured approximately 10 x 10 x 6 cm and a multinodular uterine tumor with an irregular surface of approximately 20 x 20 x 9 cm. Right heminephrectomy with normothermic ischemia of 19 minutes was performed, the kidney parenchyma was closed with a pedunculated fatty tissue patch ligament of the liver and a double-J catheter was placed (figs. 3 and 4). Afterwards, hysterectomy and a right salpingo-oophorectomy were performed.

The patient had adequate progression during the first postoperative days. The liver function tests normalized after the nephrectomy. On the third day, she presented with increased drainage output, anuria, leukocytosis (22.9 x10³/µL with 93% neutrophilia) and elevated serum creatinine (3.06 mg/dL) and urea (70 mg/dL). It was necessary to...
replace the double-J catheter after cystoscopy, cystography, and retrograde pyelography ruled out ureteral or bladder injury. This increased diuresis, the leukocyte figure and kidney function normalized, and the drainage production diminished.

The patient was released from the hospital and the drain was removed after 7 days. The ureteral stent was removed 8 weeks after surgery.

The histopathologic study reported a Fuhrman 2, pT2bNxMx, classic renal cell chromophobe carcinoma. Immunohistochemistry studies were positive for the CK7 and S100 markers, uterine leiomyomatosis, and the peritoneal fluid was positive for chromophobe tumor (figs. 5 and 6).

Discussion

In Mexico, kidney cancer represents 1%-5% of all neoplasias. More than 80% of the cases are clear cell carcinoma, followed by papillary carcinoma and chromophobe cell carcinoma. The latter represents 3%-5% of the neoplasias arising from the renal tubular epithelium, and appears to derive from the cortical portion of the collecting tubule. It is mainly diagnosed in the 6th decade of life and its incidence is similar in men and women. Chromophobe cell carcinoma makes up a heterogeneous group that includes the classic, eosinophilic, and mixed types. The principal diagnostic criterion is its morphology associated with an immunophenotypical characteristic (diffuse CK7 and positive KiT). Genetic analysis shows a chromosome 1, 2, 6, 10, 13, 17, and 21 deletion. The differential diagnosis between chromophobe cell carcinoma, clear cell carcinoma, and oncocytoma is a challenge for the anatomopathologist, because they share macroscopic and microscopic characteristics that are very difficult to differentiate. Therefore the use of diagnostic aids such as immunohistochemistry is very helpful in clarifying the diagnosis in complex cases.

The majority of chromophobe cell carcinomas are sporadic, but occasionally they are associated with the Birt-Hogg-Dubé syndrome, in which the patients develop cutaneous fibrofolliculomas, pulmonary cysts, spontaneous pneumothorax, and kidney tumors. Hereditary kidney cancer makes up 3%-5% of the renal tumors. In 2001 a familial kidney cancer syndrome was described in which the patients commonly developed cutaneous and uterine leiomyomas and type 2 papillary renal cell carcinoma. The affected gene in the uterine leiomyomatosis and hereditary kidney cancer syndrome is the fumarate hydratase gene on the 1q42-44 locus with autosomal dominant inheritance. Penetration for renal cell carcinoma is less than for cutaneous and uterine manifestations. Cutaneous leiomyomas appear in 76% of

Figure 1  Computerized axial tomography scan showing the presence of a tumor in the lower pole of the right kidney and absence of the left kidney. The upper part of the uterine tumor can be seen.

Figure 2  Computerized axial tomography scan showing the dimensions of the uterine tumor, 156 mm in the transversal direction and 96 mm in the anteroposterior direction.

Figure 3  Collecting system closure during the right partial nephrectomy.
the affected individuals, and the uterine leiomyomas are usually multiple, large, and of early onset; renal tumors tend to be unifocal, unilateral, they appear at an early age; they are more aggressive than in other hereditary kidney cancer syndromes, and range from type 2 papillary renal cell carcinoma to collecting duct carcinomas.

Currently more than 50% of the renal cell carcinomas are diagnosed incidentally. The associated symptoms can be due to local tumor growth, metastatic disease, or to the paraneoplastic syndromes that are observed in 20% of the patients and are caused by the anomalous secretion of substances that the kidney normally produces. One of the paraneoplastic syndromes linked with renal cell carcinoma is non-metastatic liver dysfunction or Stauffer syndrome, and has been reported in 3%-20% of the cases. Liver function is normalized after nephrectomy in 60%-70% of the cases.

Renal cell carcinoma continues to be a primarily surgical disease and is still considered the paradigm of the chemoresistant tumor. The goal of surgical therapy is to completely excise the tumor with a sufficient surgical margin. Nephron-sparing surgery consists of complete local resection of the kidney tumor, while leaving the greatest quantity possible of the normal functioning parenchyma of the affected kidney. This surgical modality is indicated in situations in which radical nephrectomy would leave the patient anephric or when there is high risk for dialysis; such is the case in patients with renal masses in a solitary kidney. However, for almost 15 years, some centers have extended the indications to patients with small unilateral tumors (under 7 cm) and a normal contralateral kidney; this is due to a better understanding of renal tumor histology and its oncologic threat, the oncologic equivalence of radical nephrectomy and partial nephrectomy in T1 renal tumors, and the emerging opinion about chronic kidney disease and its potential adverse effect on cardiovascular health, since it has been posited that solitary kidney is the most important risk factor for developing chronic kidney disease in these patients. It is known that the risk for developing chronic kidney disease (defined as a GFR <60mL/
thrombosis, prolonged hospital stay, and late bleeding.15,16,18-21

The main disadvantage of nephron-sparing surgery is the risk for local recurrence in the operated kidney,4 which presents in 10% of the patients with solitary kidney, in contrast to 1%-6% of the patients with a normal contralateral kidney.19 In patients with tumors in a solitary kidney treated with enucleation or partial nephrectomy, with a 5 and 10-year follow-up, overall survival has been 74% and 45%, the cancer-specific survival 80% and 63%, local recurrence-free survival 89% and 80%, and metastasis-free survival of 69% and 50%, respectively.19

The histologic type of renal carcinoma has a certain influence on outcome. In general terms, chromophobe cell carcinoma is considered to have a better outcome than clear cell carcinoma,1,2,5 with a disease-free survival at 5 years of 83.9%, at 10 years of 77.9%,3 and a mortality rate of 10%.11 Nevertheless, tumor stage, size, and nuclear grade continue to be the factors of greatest prognostic value.2,3,5

There are contradictory data about the long-term effects of renal mass reduction in patients that have undergone unilateral nephrectomy.21,24-26 There are no convincing data that indicate that living donors run a greater long-term risk due to kidney donation. However, long-term periodic follow-up that can be carried out by their personal physicians is recommended for donors.10

Conclusions

We consider long-term periodic follow-up advisable in patients with a solitary kidney, so that any pathology affecting kidney function can be detected early and treatments aimed at sparing the largest quantity of nephrons can be given.

During the study of our patient, sufficient clinical or paraclinical data were not found to be considered a uterine leiomyomatosis and hereditary kidney cancer syndrome, in which the histopathologic subtype tends to be type 2 papillary and not chromophobe, rather, the patient presented with sporadic chromophobe renal tumor in a solitary kidney incidentally associated with uterine myomatosis. The management carried out was not different from that reported in the international literature.

The immunohistochemistry findings in this case were also in accordance with that reported in the international literature. The patterns found in the hematoxylin and eosin stains are very significant for the diagnosis of chromophobe tumors, nevertheless, these findings are strengthened by the results of immunohistochemistry studies.

Conflict of interest

The authors state that there is no conflict of interest.

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