Clear cell carcinoma in horseshoe kidney: a case report and literature review

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ABSTRACT

Horseshoe kidney is the most frequent kidney fusion alteration. The majority of patients presenting with this pathology are asymptomatic. When symptoms are present they are associated with hydronephrosis, lithiasis, infections and a small percentage are associated with neoplasia. The case of a patient with macroscopic hematuria, palpable mass and pain is presented. Tumor in a horseshoe kidney was detected and patient underwent preoperative embolization and nephrectomy with isthmectomy. Follow-up progression has been satisfactory.

Key words: horseshoe kidney, clear cell carcinoma, Mexico.

RESUMEN

El riñón en herradura es la alteración de la fusión renal más frecuente. La mayor parte de los pacientes portadores de ésta patología son asintomáticos. Aquellos pacientes que presentan síntomas, están relacionados con hidronefrosis, litiasis, infecciones y en un menor porcentaje por neoplasias. Se presenta el caso de una paciente quien debió con hematuria macroscópica, masa palpable y dolor. Se demostró la presencia de una tumoración presente en un riñón en herradura, por lo cual fue sometida a embolización preoperatoria y a nefrectomía con istmectomía. Evolucionó de forma satisfactoria en su seguimiento.

Palabras clave: Riñón en herradura, carcinoma de células claras, México.
INTRODUCTION

Described for the first time in 1521 by Jacopo Berengario da Carpi, horseshoe kidney is perhaps the most frequent alteration of kidney fusion. It consists of two kidneys joined at their lower poles by parenchymatous or fibrous tissue called isthmus. It occurs in 0.25% of the population. It is more frequent in men with a 2:1 male/female ratio. It occurs in the embryo between the 4th and 6th weeks of gestation after the ureteral yolk has entered into the renal blastema. This usually occurs before rotation and the renal pelvises are facing forward. The cause has not been completely identified but it has been suggested that alterations in the position of the umbilical or common iliac artery is the cause, altering the ascent and rotation of the kidneys which end up situated in the lower part of the abdomen. The position of the superior mesenteric artery has also been implicated as a cause of ascent obstruction once the kidneys are fused. The calyces are normal in number but atypical in orientation and blood supply varies widely.

Almost a third of patients presenting with this congenital malformation remain asymptomatic. Clinical manifestations become apparent as a consequence of hydronephrosis, lithiasis, infection and less frequently, tumor.

Different abnormalities are associated with horseshoe kidney but carcinoma has been reported in only 123 patients. Forty-seven percent of these cases correspond to clear cell carcinoma, 20% to urothelial carcinoma, 20% to Wilms’ tumor and 5% to sarcomas.

Carcinoma incidence in horseshoe kidney has not been described in the literature but the observation has been made that it is higher than that of the rest of the population.

Survival in patients with this type of tumor is related to stage and histopathological grade.

Knowledge of preoperative neoplastic localization, extension and vasculature is indispensable as part of the diagnostic approach to horseshoe kidney tumors so that complete resection of the tumor can be carried out without unnecessarily removing functional tissue. Angiography or helical computed tomography (CT) angiography is essential for planning surgical approach due to the great variability of blood vessels.

CLINICAL CASE

The patient is a 69-year-old woman with no important pathologies in her medical history or drug addictions. Her disease began 6 months before diagnosis and was characterized by pain, a palpable mass at the left flank, macroscopic hematuria and an approximate 5 kg weight loss. Based on these data diagnostic approach was made at a regional general hospital, documenting left kidney tumor, and she was referred to the Centro Médico Nacional 20 de Noviembre for continued study.

Upon admittance the patient had hemoglobin 13.3 g/dL, platelets 200,000, prothrombin time 11.3", glucose 74 mg/dL, BUN 12 mg/dL and creatinine 0.7 mg/dL. Liver function tests were normal and chest X-ray revealed no metastasis.

Simple contrasted abdominopelvic computed axial tomography (CAT) was done that showed a heterogeneous lesion with calcifications that was dependent on the left kidney with fusion of both kidneys at the lower pole (Images 1 and 2). Preoperative planning included angiogram that showed the left renal artery with multiple ramifications towards the tumor site (Images 3 and 3a). Embolization was carried out on the left renal artery using platinum peripheral coils.

Afterwards the patient underwent left radical nephrectomy and isthmectomy by way of midline incision finding a 17 x 10 x 6.5 cm tumor that was dependent on the left kidney and was joined to the right kidney by the isthmus (Image 4).

The patient’s postoperative progression was satisfactory and she was released 72 hours after surgery.

Histopathological study reported a light yellow multinodular tumor with fibrous septae and scant necrotic areas in 80% of the renal parenchyma. Microscopic diagnosis was clear cell carcinoma with 10% necrosis, Fuhrman II, no invasion of the capsule, no lesions at the surgical margins and renal hilum vessels were free from malignant neoplasia. There was no tumor activity in 9 lymph nodes.

DISCUSSION

Renal cell carcinoma is the most frequent malignant neoplasia associated with horseshoe kidney. The tumor can be localized at any part of the kidney, however, its most common site is the isthmus.

In any type of kidney tumor the use of abdominopelvic CAT is essential to determine the presence of contralateral neoplasia, the degree of tumor infiltration into the perirenal fat, adjacent organs and vena cava and/or renal vein involvement. Recently helical tomography with 3D reconstruction has provided important information as to anatomical detail and it is recommended as the only diagnostic imaging study.
In patients with tumors in ectopic kidneys, routine use of angiography for embolization is not recommended except in patients with important hematuria who are not candidates for radical surgery or in patients who will undergo resection of paravertebral metastatic activity. Nevertheless, in patients with horseshoe kidney tumor this study is recommended because vascularity is widely variable. Angiograph facilitates the preservation of the non-affected renal unit and follows the principle of preserving as many nephrons as possible. For these reasons angiograph and selective embolization using platinum peripheral coils were carried out on this patient. Surgical approach was through the midline in order to have better vascular control and to have better exposure of the isthmus for isthmectomy.

The objective of kidney tumor postoperative surveillance is the early identification of metastatic activity. This activity is related to positive surgical
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The fact that a third of patients remain asymptomatic and clinical manifestation is a result of hydronephrosis, lithiasis, infection and less frequently, tumor, as in the present case.

There are fewer than 150 cases of malignant tumor in horseshoe kidney reported in the literature making this an important report for the Mexican literature.

The use of current imaging studies such as helical tomography with 3D reconstruction is an important diagnostic tool because it greatly facilitates diagnostic approach and the making of surgical decisions.

It is important to stress the use of angiographic studies and to evaluate the use of embolization especially in patients with low cardiovascular reserve who do not tolerate hemodynamic changes secondary to hemorrhage.

BIBLIOGRAPHY


Follow-up of the present case was carried out through chest and abdominal CAT since histological grade was Fuhrman II and tumor size was important. No tumor activity was detected at 7-month follow-up.

CONCLUSIONS

Horseshoe kidney is the most frequent fusion abnormality and is predominant in men. It is important to underline the fact that a third of patients remain asymptomatic and clinical manifestation is a result of hydronephrosis, lithiasis, infection and less frequently, tumor, as in the present case.

There are fewer than 150 cases of malignant tumor in horseshoe kidney reported in the literature making this an important report for the Mexican literature.

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