Atypical renal cell carcinoma in an adolescent

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ABSTRACT

Background: Kidney tumors are more common in adult patients and the most frequent type is clear cell renal cell carcinoma. Clinical symptoms are pain, hematuria and palpable mass in up to 15% of cases.

Objective: To present the case of an adolescent patient with a histologically rare type of kidney tumor.

Methods: The patient is a 17-year-old adolescent boy presenting with pain in the left costovertebral angle and with macroscopic hematuria. Physical examination revealed pain upon left costovertebral angle percussion. Diagnosis was established through kidney ultrasound and computerized tomography.

Results: Left radical nephrectomy was performed. Histological report stated papillary renal cell carcinoma with microscopic clear cell component. Surgical margins were tumor-free.

Discussion: Papillary kidney tumors are rare in adolescents. The presence of kidney tumor was determined through computerized tomography and radiological studies and radical nephrectomy was indicated.

Conclusions: Kidney tumors are rare in adolescent patients. Adequate study protocol is important in order to make opportune diagnosis and determine adequate treatment of these patients.

Keywords: kidney tumor, papillary renal cell carcinoma, Mexico.

RESUMEN

Antecedentes: Los tumores renales se presentan con mayor frecuencia en pacientes adultos; el más frecuente de ellos es el adenocarcinoma de células claras. El cuadro clínico se manifiesta por dolor, hematuria y masa palpable hasta en 15% de los casos.

Objetivo: Presentación de un tumor renal en paciente adolescente y de estirpe histológica poco frecuente.

Métodos: Presentamos el caso de un paciente masculino de 17 años quien presentó dolor en ángulo costo vertebral izquierdo y hematuria macroscópica. A la exploración física, se encontró Giordano izquierdo. Con el estudio de ultrasonido renal y tomografía computarizada, se estableció su diagnóstico.

Resultados: Se realizó nefrectomía radical izquierda. El reporte histopatológico informó adenocarcinoma renal papilar, con componente microscópico de células claras; los márgenes, libres de neoplasia.

Discusión: Los tumores renales papilares son tumores poco frecuentes en pacientes en edad adolescente. La tomografía computarizada y los exámenes de gabinete permitieron en este caso, determinar la presencia de tumor renal, en el que estaba indicada la nefrectomía radical.

Conclusión: Los tumores renales son poco frecuentes en pacientes adolescentes. Es importante realizar un adecuado protocolo de estudio para poder llegar al diagnóstico oportunamente y así determinar el tratamiento más adecuado en estos pacientes.

Palabras Clave: Tumor renal, adenocarcinoma renal papilar, México.
INTRODUCTION

Renal cell carcinoma (RCC) represents 2-5% of pediatric kidney tumors and 0-2% of all renal cell cancers that occur before 21 years of age. Mean RCC presentation age in pediatric patients is 9-15 years.1,2

The majority of patients present with macroscopic hematuria, lumbar pain or palpable renal mass. Twenty-five percent of patients are asymptomatic and tumor is diagnosed through imaging studies.3-5

In 20% of RCC cases, metastasis to lung, bone, liver or central nervous system is identified at the time of diagnosis. There are cases of bilateralness that are associated with underlying conditions such as von Hippel-Lindau disease.1,2

Pediatric papillary RCC has the classic histological architecture of its counterpart in adults. This variety is frequent and in different series represents 20-50% of pediatric RCC - a higher incidence than in the adult population.5-7

An attempt has been made to establish individual prognostic factors for RCC. In a revision of 619 patients, Eun-Jung et al stratified patients as older or younger than 55 years of age. Their results stated that the below 55-year-olds presented with lower stage, lower nuclear grade and smaller size than the above 55-year-olds.

McClellan et al evaluated 52 patients with von Hippel-Lindau disease that presented with tumors smaller than 3 cm and concluded that none of them presented with metastatic disease in a 60-month follow-up after tumor resection.8-10

Image 1. Computed tomography showing contrast-enhanced 10 cm x 8.5 cm left kidney tumor.
CASE PRESENTATION

The patient is a 17-year-old adolescent student with a medical history of CHARGE syndrome in three direct relatives, adequate dietary and hygienic habits, occasional smoking for a one-year period and appendectomy at the age of 13 with no complications. Patient came to the emergency room for pain of 12-hour progression in the left costovertebral angle accompanied by total macroscopic hematuria with no clots, by dysuria and by urinary frequency. Patient said there had been no previous hematuria. At physical examination, vital signs were within normal parameters, there was adequate tegument coloration and hydration and no signs of cardiopulmonary compromise. There was left costovertebral percussion, no palpable masses or visceromegaly, peritoneal irritation or ureteral points. Genitals had masculine aspect and penis and both testes had normal characteristics. Laboratory work-up reported: Hb 10.5 g/dL, Hct 30%, PT 16 sec, PTT 45.8 sec, INR 1.3, creatinine 1.0 mg/dL, urea 23.5 mg/dL, BUN 11 mg/dL, Na 145 mmol/L, K 4.5 mmol/L, Cl 112 mmol/L, calcium 8.4 mg/dL, alkaline phosphatase 120 U/L, total bilirubin 1.4 mg/dL, erythrocyte sedimentation rate 25 mm/hour. Urinalysis: straw color, clear, density 1.000, pH 5.0, Hb ++++, leukocytes 5-10 x field, innumerable erythrocytes and scarce bacteria. Radiology and imaging studies ordered: chest X-ray, simple abdominal X-ray, computed tomography (CT) of abdomen and pelvis in which a 10 cm x 8.5 cm lobulated image dependent on the left kidney was seen and was partially enhanced with intravenous contrast application (Image 1). Left kidney tumor diagnosis was made with this finding.

Patient was hospitalized to complete study protocol and decide on treatment. Kidney scintigram with Tc99m-DTPA showed total glomerular filtration of 62.18 mL/minute (15.88 mL/minute and 46.3 mL/minute for left and right kidneys, respectively) but left kidney function was not seen. Based on these results, left ascending pyelography was done in which contrast medium showed communication from left collector system to tumor mass (Image 2). Left radical nephrectomy was carried out in which there were no intraoperative or postoperative complications (Image 3). Histopathological study reported papillary renal adenocarcinoma with extensive necrosis and microscopic component of papillary clear cell adenocarcinoma, Fuhrman grade III, with no vascular or capsular invasion and no invasion of the pyelocaliceal system (Image 4). Margins were also negative. Adrenal gland was tumor free and there were no metastatic lymph nodes in hilar fat. Patient

Image 2. Left ascending pyelography in which communication of the contrast medium from collector system to tumor is seen.

Image 3. Left radical nephrectomy produced 13 cm x 8.3 cm x 6.5 cm left kidney weighing 360 g with 9.5 cm x 8 cm x 5 cm tumor having hemorrhagic areas that do not involve the renal capsule.
postoperative progression was good and he was released from hospital to continue under outpatient surveillance.

■ DISCUSSION

Renal cell carcinoma (RCC) is a rare oncological process in pediatric patients.

Once the finding of kidney tumor is made it is difficult, and at times impossible, to differentiate RCC from other solid tumors such as Wilms’ tumor, among others. There is evidence of greater incidence of papillary RCC in children and genetic and familiar patterns have been observed in these cases.6,7

Tumor resection is the most important factor in regard to prognosis and survival in pediatric RCC patients and, as in adult patients, there is not a favorable response to chemotherapy or radiotherapy. 7,8

Different studies have tried to establish prognostic factors for this type of patient that can influence survival, even though in the majority of cases patients seek medical attention when there is multiple symptomatology and tumors have become large.7,8,10

In the present case, the patient presented with costovertebral angle pain accompanied by macroscopic hematuria, which were the reason for beginning diagnostic approach. Abdominopelvic CT, renal scintigram and ascending pyelography were done in order to carry out the most conservative procedure possible. However, given the results of the imaging studies and intraoperative findings, it was not possible to perform partial nephrectomy.

■ CONCLUSIONS

RCC in the second decade of life is a rare neoplasia when compared with adult age and its diagnostic and therapeutic approach is similar to that of RCC in adults. The papillary variety is particularly frequent and hereditary patterns are evident in pediatric RCC. This diagnosis should be considered in patients presenting with macroscopic hematuria so that early surgical resection can be offered, improving therapeutic possibilities for these patients.

BIBLIOGRAPHY