Metanephric adenofibroma: presentation of an atypical case and literature review

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ABSTRACT

Metanephric adenofibroma is a rare kidney tumor. Clinically, it usually presents with hematuria and some cases are associated with polycythemia and high blood pressure. It is indistinguishable from other solid tumors, particularly Wilms’ tumor. Optimum treatment has yet to be established. The case of a patient with atypical symptoms who underwent percutaneous biopsy that reported metanephric adenofibroma is presented. Right heminephrectomy was performed leaving tumor-free surgical margins. There was no metastasis at postoperative follow-up.

Key words: metanephric adenofibroma, kidney tumor, Mexico.

RESUMEN

El adenofibroma metanéfrico es una neoplasia renal poco frecuente. Suele presentarse clínicamente con hematuria, algunos casos están relacionados con policitemia e hipertensión. Es indistinguible de otras neoplasias sólidas, en particular del tumor de Wilms. Su óptimo tratamiento aún no ha sido establecido. Presentamos el caso de un paciente quien debuta con síntomas atípicos, sometido a biopsia percutánea, la cual reportó un adenofibroma metanéfrico. Se realizó heminefrectomía derecha, obteniendo márgenes quirúrgicos libres de tumor. El seguimiento posoperatorio no demostró la presencia de metástasis.

Palabras clave: Adenofibroma metanéfrico, tumor renal, México.
INTRODUCTION

Initially called nephrogenic adenofibroma, metanephric adenofibroma is a rare kidney tumor first described by Hennigar and Beckwith. It is a tumor composed of a mixture of embryonic, stromal and epithelial elements. Since 2004 it appears in the World Health Organization (WHO) classification of kidney tumors. Due to its stromal component and the presence of embryonic epithelium it has been proposed that this neoplasia can arise from the maturation of intralobular nephrogenic remnants. An association between this neoplasia and Wilms’ tumor has been described. Some authors consider it to be a mature and hyperdifferentiated form of Wilms’ tumor.

Only a few cases have been reported in the literature and the National Wilms’ Tumor Study Group (NWTSG) Pathology Center is the group with the largest series of cases of this neoplasia.

Metanephric adenofibroma usually presents in individuals between the first and third decade of life and is more frequent in men with a 2:1 man/woman ratio.

The most common symptom is hematuria. However, a number of patients remain asymptomatic while some patients can present with polycythemia which is resolved through tumor resection.

The tomographic image of this tumor is indistinguishable from other solid tumors, particularly Wilms’ tumor. There are no specific data about its appearance to guide the physician towards its diagnosis.

Optimum metanephric adenofibroma treatment has not yet been established due to its low frequency and to the fact that the majority of pathologists believe that it can be a benign lesion since up to the present there have been no reports of its causing metastasis.

CLINICAL CASE

The patient is a 16-year-old male with no previous pathology in his medical history. His disease began 3 weeks prior to his admittance to the Centro Médico Nacional 20 de Noviembre, with symptoms of asthenia, adynamia, general malaise, hyporexia, fever up to 40º and chills predominantly at night along with an approximate 7 kg weight loss in one month. He went to his corresponding general hospital where he underwent study protocol. Abdominopelvic computerized axial tomography (CAT) revealed the presence of a right kidney tumor for which he was referred to the authors’ urology service.

Upon admittance, patient vital signs were within normal parameters, except for fever of 39º. There was no cardiac or pulmonary involvement. Two left inguinal lymph nodes of approximately 3 cm each were palpated. Laboratory work-up was normal. Repeat abdominopelvic CAT was done revealing the presence of a renal mass located at the lower pole of the right kidney of approximately 4.0 x 3.9 cm (Image 1).

During his hospital stay the patient continued with symptomatology suggestive of lymphoproliferative neoplasia and therefore excisional biopsy of the inguinal lymph nodes was performed. Biopsy report stated mixed hyperplasia (follicular and sinusoidal) with no evidence of malignancy. Percutaneous biopsy of the kidney tumor guided by tomography was carried out and its report stated the presence of metanephric adenofibroma (Image 2).

The patient then underwent right heminephrectomy, obtaining a surgical specimen with a 3 x 2.5 x 2 cm lesion with irregular but well-defined margins, hemorrhagic appearance that did not invade the kidney capsule and surgical margins that were tumor-free (Image 3).

Histological observation revealed epithelial and stromal components suggestive of metanephric adenofibroma, along with abundant microcalcifications and psammomatous bodies.

The patient remained hospitalized for 3 days for postoperative recuperation during which he did not present with fever and his general state of health improved. At 5-month follow-up, imaging studies have shown no signs of metastasis.

DISCUSSION

Metanephric neoplasias are a group of lesions related to Wilms’ tumor. These tumors include purely stromal lesions, purely epithelial lesions (metanephric adenoma) and mixed epithelial-stromal lesions (metanephric adenofibroma).

Metanephric adenofibroma was initially described by Beckwith and Hennigar in 1992. It is a rare benign kidney tumor and very few cases have been reported in the international literature. Its origin has been proposed to arise from nephrogenic remnants. These remnants, classified by Beckwith, are residual elements from embryological kidney development. This tumor is characterized by the proliferation of mesenchymatous cells that surround nodules of immature epithelial cells. The epithelial component is histologically identical to metanephric adenoma and is similar to Wilms’ tumor. Differential diagnosis is very difficult to make but it is crucial to distinguish between them for treatment purposes since metanephric adenofibroma is a benign tumor and does not require chemotherapy while Wilms’ tumor is malignant and thus requires it.
The management of these tumors has not yet been defined and the majority of these tumors are benign. However, metanephric adenofibroma may be associated with non-embryonic malignant elements (tubulopapillary carcinoma). In published series, patients presenting with this variety have been treated with nephrectomy only and no metastasis has been reported in their follow-up. Other series recommend the use of adjuvant chemotherapy and have demonstrated no recurrence. When epithelial components are almost indistinguishable from Wilms’ tumor, adjuvant chemotherapy (dactinomycin and vincristine) is recommended.

**CONCLUSIONS**

Metanephric adenofibroma is a benign kidney tumor and only a few cases have been reported in the international literature. It usually presents in young people and hematuria is the common clinical presentation. It is indistinguishable in imaging studies from other solid tumors. Its diagnosis requires histopathological confirmation and the demonstration of stromal and epithelial components characteristic of this lesion. Ideal treatment has not yet been defined but the majority of patients are treated with nephrectomy with good results. Adjuvant chemotherapy may be reserved for disease that is associated with malignant elements.
BIBLIOGRAPHY