Intrarenal schwannoma: two case reports and a literature review

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ABSTRACT

Retroperitoneal tumors are relatively rare and tend to be classified according to the tissue in which they arise. Of the nerve sheath tumors, schwannomas have a 1-10% prevalence among total retroperitoneal primary neoplasms and a 0.2-0.6% prevalence among all neoplasms. Two cases of intrarenal schwannomas that were incidentally diagnosed after performing radical nephrectomy are presented.

Key words: Renal schwannoma

RESUMEN

Los tumores del retroperitoneo suelen ser poco frecuentes y se clasifican según el tejido del que se originan. Dentro de los tumores derivados de las vainas nerviosas se encuentra el schwannoma, cuya prevalencia varía entre 1% a 10% del total de las neoplasias primarias retroperitoneales y entre 0.2% a 0.6% de todas las neoplasias. Presentamos dos casos de schwannoma intrarrenal diagnosticados de forma incidental, tras la realización de una nefrectomía radical.

Palabras clave: neoplasias del riñón, neurilemoma, México.

OBJECTIVE

Two cases of intrarenal schwannoma management and progression are presented along with a review of the literature.

CLINICAL CASE

CASE 1

The patient is a 37-year-old woman. Her family medical history included a father with high blood pressure. She smoked 1 to 2 cigarettes a day for 10 years and was an occasional drinker. Recently diagnosed high blood pressure is controlled with captopril. She sought medical attention due to the sensation of a mass in the left back flank of 3-month progression that was not associated with pain, weight loss or hematuria. Physical examination revealed blood pressure of 120/90 and soft abdomen with a palpable non-painful firm mass at the left flank. Laboratory tests reported Hb: 14.3g/dl, creatinine: 0.73mg/dl and normal liver function.

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test (LF) and chest X-ray. Computed tomography (CT) showed a 12 x 12 cm solid mass dependent on the left kidney with areas of necrosis and calcification (Image 1). Left radical nephrectomy was performed and a 20 x 15 x 10 cm tumor dependent on the inferior pole was identified (Image 2). Histopathological study reported renal schwannoma. Special immunohistochemical staining resulted in S-100 (+) and vimentin (+).

CASE 2
This patient is a 71-year-old woman. Family medical history included a diabetic father and a mother with high blood pressure. She sought medical attention due to a painful mass at the right flank of 2-month progression associated with 7 kg weight loss and occasional chills. Physical examination revealed blood pressure of 120/80 and a normal thorax. Her abdomen was soft and a hard painful mass involving the entire right hemi-abdomen was palpated. Laboratory tests reported Hb: 10.7, creatinine: 1.22 and urinalysis: 3-4 erythrocytes per field. LF and chest X-ray were normal. CT and magnetic resonance angiography showed heterogeneous solid mass with areas of necrosis, slight collection of contrast material and no large vessel involvement (Images 3 and 4). Radical nephrectomy was performed revealing a 27 x 15 x 18 cm tumor dependent on the inferior pole (Image 5). Histopathological study reported renal schwannoma and immunohistochemistry resulted in S-100 (+) and Vimentin (+).

**BIBLIOGRAPHIC REVIEW**
Renal parenchymal tumors make up a large group of neo-formations possessing well-defined clinical, histological and therapeutic characteristics for urothelial tumors. Different types of neoplasms may arise from the kidney: epithelial, mesenchymal, neuroendocrinial, hematopoietic, etc. The majority of these neoplasms are malignant epithelial tumors. Unlike renal schwannomas the so-called renal cell carcinomas are neoplasms that are classified as mesenchymatous in origin (1-3).

Retroperitoneal tumors are rare and usually classified according to the tissue in which they arise. Among the nerve sheath tumors, schwannomas represent 1-10% of all primary retroperitoneal tumors and 0.2-0.6% of all neoplasms. Kidney-dependent schwannomas are even rarer and there are only 10 cases reported in the literature (2-4).

In 1954 Ackerman classified primary retroperitoneal tumors according to histological findings and divided them into tumors of nerve origin, of mesodermic origin and tumors arising from embryonary remains (3-4). Malignant schwannoma is a rare tumor made up of Schwann cells that are the supporting tissue of the peripheral nerves (2 - 4). Men and women are equally affected and the age of presentation is from 30 to 60 years old. Schwannoma can be associated with von Recklinghausen disease, giving it a poor prognosis (1-3). Pre-surgical diagnosis is uncommon, ultrasound images show mixed echogenicity and CT shows cystic, necrotic and solid areas with interior calcification (4-7).
Histology studies report fusiform cells with ovalated nuclei (Antoni A) and nuclear pleomorphism (Antoni B). Immunochemical results are positive for vimentin, S-100 protein and neuron-specific enolase (2,4,7).

**DISCUSSION AND CONCLUSIONS**

In the mature adult the majority of renal neoplasms arise in the epithelium and the less frequent tumors arise in the mesenchyma. Leiomyosarcoma is the most common sarcoma and angiomyolipoma is the most common benign mesenchymatous neoplasm. Nerve sheath tumors of the kidney are extremely rare and only 10 such cases have been reported in the literature.

Clinically the majority of schwannomas are asymptomatic with a symptomatology related to the effect of the mass and on rare occasions they present with fever, anemia and weight loss.

In the majority of cases preoperative diagnosis is malignant kidney tumor and treatment is radical nephrectomy. Nephron-preserving surgery should be considered in select cases due to the possible benign nature of the pathology.

In cases not associated with von Recklinghausen disease, radical nephrectomy alone is usually sufficient treatment. However, when the pathology is associated with this disease, chemotherapy has not had an effect on survival and radiotherapy management is not indicated.

Recurrence of retroperitoneal schwannoma by itself is 20 - 30% at 5 years. When associated with von Recklinghausen disease the rate increases to 45 - 75% at 5 years. Follow-up is the same for retroperitoneal schwannoma and clear cell carcinoma patients and is stricter in those patients who also present with von Recklinghausen disease.

**BIBLIOGRAPHY**