CLINICAL CASE

Synchronous retroperitoneal schwannoma with clear cell renal cell carcinoma


Urology Division, Hospital General «Dr. Manuel Gea González», Mexico City, Mexico

Received 1 November 2015; accepted 25 February 2016
Available online 14 April 2016

KEYWORDS
Schwannoma; Renal cell carcinoma; Tumor

Abstract
Background: Schwannomas are well-encapsulated tumors arising from the peripheral nerve sheaths and they appear in young or middle-aged adults. These tumors make up 0.5-2.7% of retroperitoneal tumors. The majority of schwannomas are benign and single, although multiple tumors have been described. There are reports of malignant cases associated with von Recklinghausen’s disease and melanocytic nevus in children, with hypercalcemia of parathyroid origin.

Clinical case: A 54-year-old woman had a past history of tubular adenoma of the colon. Her current illness began with lower urinary tract symptoms, mainly of storage, along with total gross hematuria with no clots. An abdominal tomography scan revealed a 21mm contrast-enhanced left kidney upper pole lesion and a 53×48×32mm solid lesion adjacent to the left adrenal gland. The latter had well-defined regular edges that anteriorly displaced the pancreas and splenic artery, with punctiform intraparenchymal calcifications. Left partial nephrectomy of the upper pole was performed and the histopathology study reported clear cell carcinoma limited to the kidney, Fuhrman 2, with no lymphovascular invasion.

Discussion: Schwannomas in the retroperitoneum are rare, making up approximately 1% of retroperitoneal tumors. Patient age at the time of diagnosis varies from 20 to 50 years. Incidence has been observed to be slightly more frequent in women than in men. When symptoms present, the most frequent are abdominal pain, abdominal distension, and lumbalgia.

Conclusions: Retroperitoneal schwannoma is a rare disease. Its presentation is nonspecific and it is generally an incidental finding. Computed tomography and magnetic resonance are useful for the preoperative approach.

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*Corresponding author at: División de Urología, Hospital General «Dr. Manuel Gea González», Calzada de Tlalpan 4600, México D.F., México.
Email: ulisescsa@gmail.com (U. Sánchez-Aquino).
**PALABRAS CLAVE**
Schwannoma; Carcinoma de células renales; Tumor

**Resumen**

**Introducción:** Los schwannomas son tumores bien capsulados, que se originan en las vainas de los nervios periféricos, y que aparecen en jóvenes o adultos de edad media. Constituyen entre el 0.5-2.7% de los tumores retroperitoneales. La mayoría de los schwannomas son benignos y únicos, aunque hay descritos casos múltiples. Se han informado casos malignos asociados a enfermedad de von Recklinghausen, nevus melanocítico en niños y con hipercalcemia de origen paratiroideo.

**Caso clínico:** Se trata de una mujer de 54 años con antecedente de adenoma tubular de colon. Comienza con síntomas urinarios en el tracto inferior, predominio de almacenamiento, agregándose hematuria macroscópica total, no formadora de coágulos. Tomografía abdominal con presencia de lesión en polo superior en riñón izquierdo de 21 mm, que refuerza tras la aplicación de medio de contraste, lesión sólida adyacente a la glándula suprarrenal izquierda de bordes regulares bien definidos, que desplaza en sentido anterior el páncreas y la arteria esplénica de 53×48×32 mm, con calcificaciones intraparenquimatosas puntiformes. Se realiza nefrectomía parcial izquierda de polo superior. Reporte histopatológico: carcinoma de células claras limitado al riñón Fuhrman tipo II, sin invasión linfvascular.

**Discusión:** En el retroperitoneo, los schwannomas son rares, constituyendo aproximadamente el 1% de los tumores retroperitoneales. La edad del diagnóstico suele estar entre los 20-50 años. La incidencia observada es levemente más frecuente en las mujeres que en los varones. Cuando presenta síntomas, los más frecuentes son el dolor abdominal, la distensión abdominal y la lumbalgia.

**Conclusiones:** El schwannoma retroperitoneal es una enfermedad poco frecuente. Su forma de presentación es inespecífica y generalmente es un hallazgo. La TC y la RM son útiles para el enfrentamiento preoperatorio.

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**Introduction**

Benign soft tissue tumors originating in the nerve sheath of the peripheral nerves are a subtype of neoplasias that include neurofibromas, solitary circumscribed neuromas, perineuromas, and schwannomas. Schwannomas represent 4% of all retroperitoneal tumors. They usually present as a single, slow-growing, nonaggressive tumor discovered incidentally in an imaging study carried out for a different indication. The classic finding in a tomography scan is a solid, well-circumscribed, contrast-enhanced lesion that can have cystic degeneration and displace local structures. They can develop in any nerve of the body, but they have a predilection for peripheral nerves of the upper limbs and cranium (except cranial nerve pairs I and II), and from 0.3-3.2% present in the retroperitoneum. The risk for malignization is extremely low and recurrences are rare. Preoperative diagnosis is important because retroperitoneal tumors have a 4-fold higher risk for being malignant than for being benign. The treatment of choice is surveillance and surgical resection is indicated only when there are severe symptoms. Synchronous retroperitoneal schwannomas with clear cell renal cell carcinoma have not been previously reported in Mexico.

**Aims**

To present in the Mexican national literature a case of synchronous retroperitoneal schwannoma with clear cell renal cancer managed at the Hospital General “Dr. Manuel Gea González”.

**Case presentation**

A 54-year-old woman had a past history of tubular adenoma of the colon. Her current illness began one month prior to her admission with lower urinary tract symptoms and self-limited gross hematuria with no clots. Physical examination results were normal. Computed tomography urography showed a 21 mm contrast-enhanced tumor in the upper pole of the left kidney that was observable in the nephrogenic phase and a second 6.0 x 4.0 cm solid retroperitoneal tumor adjacent to the left adrenal gland, displacing adjacent structures. It had a density of 23 HU that was measurable at 53 HU after contrast enhancement (fig. 1). Serum laboratory results were unaltered. Free cortisol in urine was 60 µg and metanephrines in urine at 24 h was 145 µg, both findings consistent with adrenal tumor. A left open partial nephrectomy of the upper pole was performed and the histopathology study reported clear cell carcinoma pT1a, Fuhrman II (fig. 2) and the definitive histopathology study of the retroperitoneal tumorectomy stated retroperitoneal schwannoma. The patient had a satisfactory postoperative period and was released from the hospital on day three. Three months after the procedure she was asymptomatic and in a good state of health. The patient is currently undergoing outpatient follow-up every 6 months.
Synchronous retroperitoneal schwannoma with clear cell renal cell carcinoma

Discussion

Preoperative diagnosis of a retroperitoneal schwannoma is important due to its risk for malignancy. Having tomographic findings with clinical symptoms is sufficient for making the diagnosis. In our patient, the diagnostic evaluation was centered on defining the etiology of the gross hematuria that she first presented with. The abdominal tomography scan revealed a left kidney tumor and the retroperitoneal tumor was an incidental finding. The guidelines recommend making the diagnosis based on the imaging characteristics and clinical data, and with today’s imaging techniques this can be done in a large number of patients. Fine needle aspiration biopsy is used when there is diagnostic doubt. Anamnesis and complete physical examination with a contrast-enhanced tomography scan and fine-needle biopsy produce the diagnosis in nearly 100% of the patients. In our case, the presence of a kidney tumor suggested an adrenal tumor, and in this context, with a high risk for malignancy. The treatment of choice is surveillance due to the risk for injuring neighboring structures during surgery. If resection is feasible, then it is adequate treatment. Our patient underwent a radical procedure due to kidney cancer and there was adequate access to completely resect the tumor.

The definitive diagnosis was not confirmed until the histopathologic result. Recurrence is rare, but the patient was kept under surveillance because of the kidney cancer. In our opinion, the preoperative diagnosis of retroperitoneal tumors should be made based on imaging findings and confirmed through fine needle aspiration biopsy when there is diagnostic doubt. The synchronous clear cell renal cancer present in this case is the first to be reported in the Mexican medical literature.

Conclusions

Retroperitoneal schwannomas are rare, benign, asymptomatic tumors discovered incidentally. Preoperative diagnosis is key for treatment decision. If feasible, total resection is adequate treatment. The synchronous clear cell renal cancer presented herein is the first case to be reported in the Mexican medical literature.

Ethical responsibilities

Protection of persons and animals. The authors declare that no experiments were performed on humans or animals for this study.
Data confidentiality. The authors declare that no patient data appear in this article.

Right to privacy and informed consent. The authors declare that no patient data appear in this article.

Financial disclosure
No financial support was received in relation to this article.

Conflict of interest
The authors declare that there is no conflict of interest.

References