

# Renal Schwannoma: A Rare Pathology

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## ABSTRACT

Schwannomas are benign neoplasms that originate from Schwann cells of the peripheral nerves sheath. They are most commonly found in the head, neck, and extremities, although up to 3% occur in the retroperitoneum, with renal involvement being exceptional. The renal hilum is the most frequent site of origin; however, many renal schwannomas are located in the parenchyma, mimicking renal cell carcinoma, or in the perirenal space, broadening the range of diagnostic possibilities. We present a case in which a mass in the right anterior perirenal space was identified on imaging and confirmed by percutaneous biopsy.

**Keywords:** schwannoma, kidney, retroperitoneal tumor, percutaneous biopsy.

## Schwannoma renal: una patología infrecuente

### RESUMEN

Los schwannomas son neoplasias benignas originadas en las células de Schwann de la vaina del nervio periférico. Se localizan con mayor frecuencia en cabeza, cuello y extremidades, aunque hasta un 3% se presenta en el retroperitoneo; es una afectación renal excepcional. El hilio renal es el sitio más frecuente de origen; sin embargo, muchos schwannomas renales se ubican en el parénquima simulando carcinoma de células renales o en el espacio perirrenal abriendo el abanico de posibilidades diagnósticas. Presentamos un caso en el que una masa en el espacio perirrenal anterior derecho fue hallada por imágenes y confirmada mediante biopsia percutánea.

**Palabras clave:** schwannoma, riñón, tumor retroperitoneal, biopsia percutánea.

## INTRODUCTION

Schwannomas were described as benign neoplasms arising from Schwann cells within the peripheral nerve sheath<sup>1-3</sup>. These lesions most commonly occur sporadically; however, in some cases they have been found to be associated with conditions such as neurofibromatosis type 1 and type 2<sup>4-5</sup>. Schwannomas most frequently arise in the head, neck, and extremities; nevertheless, up to 3% have been reported to occur in the retroperitoneum, with

renal involvement being an extremely rare condition<sup>5</sup>, with fewer than 50 cases reported in the literature<sup>1,2</sup>.

This entity may present in patients of any age, although a slight predilection for adults and for the female sex has been observed<sup>8,1</sup>. The renal hilum and the parasympathetic nerve fibers accompanying the renal artery constitute the most frequent site of origin (51%)<sup>1-6</sup>. However, a substantial proportion of renal schwannomas have been found within the renal parenchyma (43%),

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mimicking renal cell carcinoma, or in the perirenal parenchyma, thereby broadening the spectrum of differential diagnoses<sup>8</sup>.

In many cases, these lesions were identified incidentally, which posed a significant challenge when attempting to differentiate them from other renal and perirenal masses<sup>2</sup>. The diagnosis of this condition has been established primarily through histopathological examination, obtained either from surgical resection or percutaneous biopsy, with the aim of preserving as much functional renal parenchyma as possible<sup>2</sup>.

## CLINICAL CASE

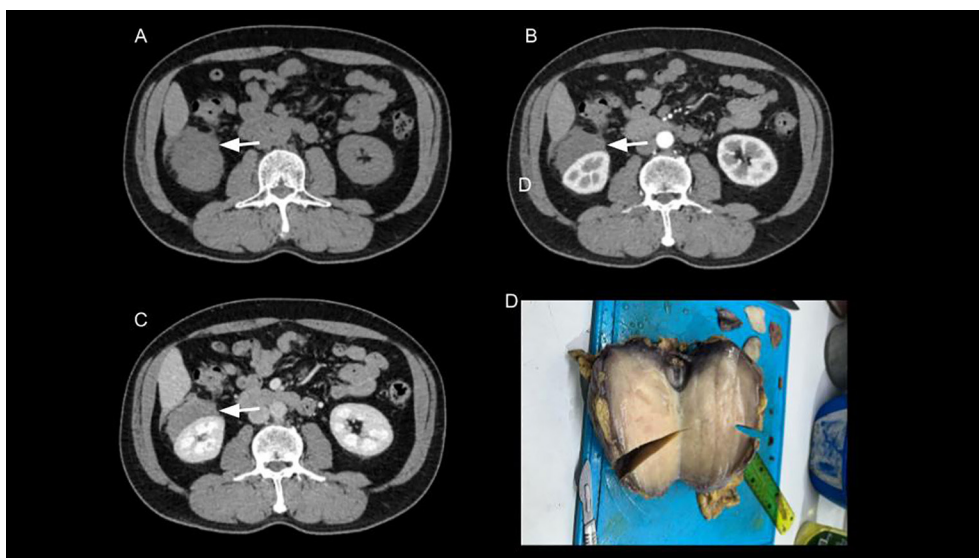
We present the case of a 59-year-old male patient with a medical history of HIV infection under antiretroviral therapy, light tobacco use, and arterial hypertension under pharmacological treatment and adequate control. He presented for an outpatient consultation due to an umbilical hernia. The patient denied abdominal pain, hematuria, and weight loss, and reported no relevant family history. Vital signs were within normal limits. Laboratory tests and an abdominal ultrasound were requested.

Laboratory results were within normal ranges, with undetectable viral load, normal white blood cell and CD4 counts, no evidence of anemia, and preserved renal function. Abdominal ultrasound revealed a retroperitoneal mass in the right anterior perirenal space, well circumscribed, in close contact with the renal cortex, isoechoic relative to it, with negative Doppler vascular signal. The lesion did not deform or infiltrate the kidney, showed no involvement of the renal hilum, no urinary tract dilatation, and no retroperitoneal lymphadenopathy.

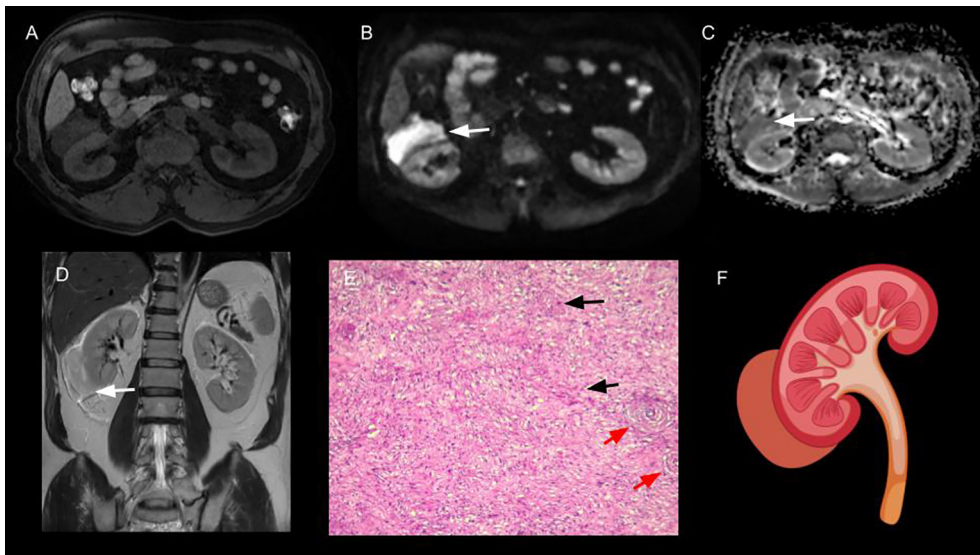
The evaluation was complemented with contrast-enhanced computed tomography (CT) of the abdomen and pelvis (Fig. 1), which confirmed the presence of a homogeneous soft-tissue –density lesion with irregular margins, located in the right anterior perirenal retroperitoneal space, measuring 52 × 28 mm. Following intravenous contrast administration during the nephrogenic phase, the interface between the renal cortex and the lesion could be clearly delineated; the lesion appeared hypovascular with mild delayed enhancement.

Magnetic resonance imaging (MRI) of the abdomen and pelvis with intravenous contrast was performed for further characterization (Figs. 2 and 3). This demonstrated a homogeneous right anterior perirenal mass, hyperintense on T2-weighted and STIR sequences, hypointense on T1-weighted sequences, with marked diffusion restriction and progressive/delayed enhancement after contrast administration. No signs of renal capsular invasion or distant lesions were observed.

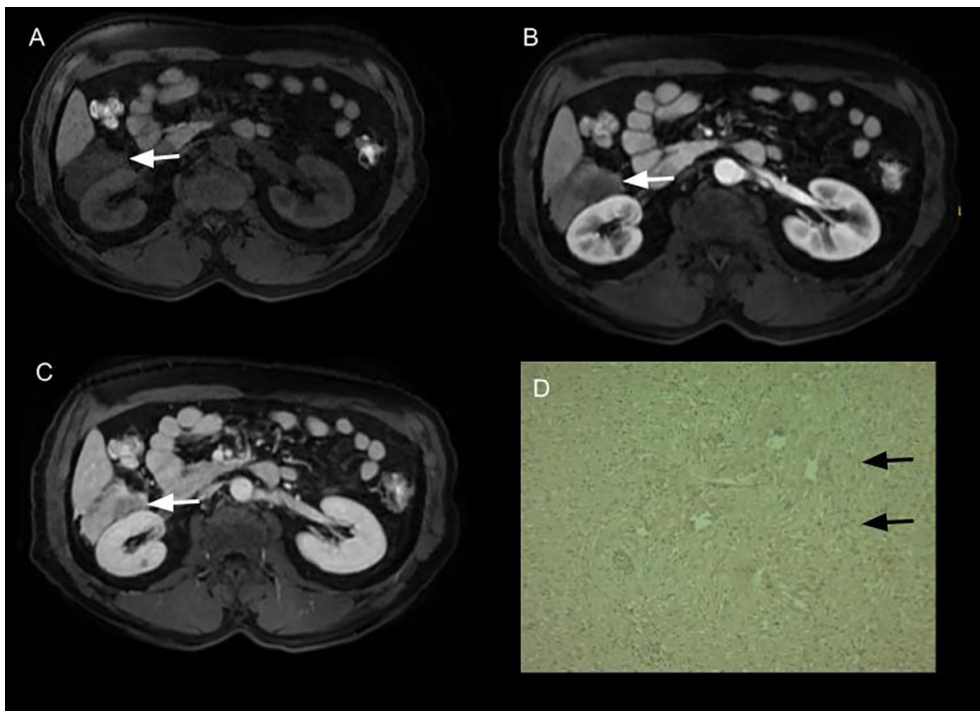
Given the incidental nature of the findings and the imaging characteristics of the lesion, lymphoma was initially suspected due to the homogeneous appearance and delayed enhancement in a patient with HIV. However, other possibilities were considered, including sarcoma, Kaposi sarcoma, or mesenchymal tumors such as leiomyosarcoma. The lesion did not exhibit aggressive behavior, and there was strong suspicion of retroperitoneal fibrosis. Additionally, given the patient's underlying chronic condition –although not in a state of active immunosuppression– chronic granulomatous infections such as mycobacterial infection, histoplasmosis, or fungal disease could not be completely ruled out, although they were considered less likely.



**Figure 1.** (A) Pararenal lesion adjacent to the anterior surface of the right kidney (white arrow), isodense on the non-contrast phase. (B) Arterial phase showing progressive enhancement of the lesion. (C) Delayed phase demonstrating homogenization of the lesion. (D) Gross specimen showing a solid, tan-colored, encapsulated mass.



**Figure 2.** (A) Abdominal MRI shows a well-circumscribed hypointense lesion on axial T1-weighted images. (B, C) Diffusion-weight imaging demonstrates diffusion restriction. (D) The lesion appears hyperintense on coronal T2-weighted images. (E) Histopathological microscopy showing a neoplasm composed of uniform spindle cells arranged in a wavy fascicular pattern (black arrow), with sclerotic-walled blood vessels (red arrow). (F) Schematic representation of the lesion's location in relation to the inferior pole of the right kidney.



**Figure 3.** (A) Abdominal MRI demonstrating a right pararenal lesion (white arrow) that is hypointense on axial T1-weighted images. (B) Arterial phase showing minimal enhancement. (C) Delayed phase demonstrating progressive contrast uptake. (D) Immunohistochemical staining for S100 protein showing diffuse brown staining of the nuclei and cytoplasm of spindle cells (black arrow).

After multidisciplinary discussion, measurement of immunoglobulin levels was performed and found to be normal, and a percutaneous biopsy was indicated to determine the origin of the lesion. CT-guided percutaneous biopsy was performed using a core needle, with cytological

evaluation showing no evidence of infectious granulomatous disease. The biopsy specimens were therefore submitted for deferred histopathological analysis.

The pathological report revealed a spindle-cell neoplasm with fibromyxoid stroma and sclerotic vessels.

No cellular atypia, mitotic activity, or necrosis was observed. Immunohistochemistry showed diffuse S100 positivity, negative desmin, and a Ki-67 index of 1%, leading to a final diagnosis of renal schwannoma. Given the rarity of this diagnosis, nephrectomy was decided upon after confirmation.

## DISCUSSION

Renal schwannomas are benign, slow-growing tumors that present with nonspecific symptoms or are asymptomatic and are diagnosed incidentally<sup>1-3</sup>. The most common presenting symptom is flank and/or abdominal pain, followed by hematuria, fever, and a palpable mass<sup>1,5,7</sup>.

Computed tomography (CT) and magnetic resonance imaging (MRI) are the main imaging modalities used for diagnosis; however, in many cases, these lesions cannot be easily distinguished from other renal masses based solely on imaging findings<sup>1-3</sup>. On CT, benign schwannomas appear as well-circumscribed soft-tissue lesions with homogeneous or heterogeneous enhancement after contrast administration<sup>9</sup>. On MRI, they typically present as solitary, well-defined, rounded lesions with lobulated contours, which are isointense on T1-weighted images and hyperintense on T2-weighted images<sup>6,7,9</sup>. Following intravenous contrast administration, homogeneous and progressive enhancement is usually observed<sup>6</sup>. Larger tumors may exhibit cystic or hemorrhagic areas that contribute to tumor heterogeneity<sup>6</sup>. However, these imaging findings are nonspecific, and renal schwannomas are often misdiagnosed as renal cell carcinoma, making pathological examination necessary to confirm the diagnosis, either through surgical resection or percutaneous biopsy<sup>3</sup>.

Malignant degeneration is extremely rare, with only four cases reported in the literature<sup>1</sup>. Malignant schwannomas behave as high-grade sarcomas, with a high likelihood of local recurrence and distant metastasis. The presence of intratumoral hemorrhage or necrosis on imaging findings suggests malignancy<sup>1</sup>.

Although histopathological examination by fine-needle aspiration or renal biopsy would be the appropriate preoperative tool to differentiate renal masses<sup>7</sup>, these procedures are not always used because of their limited diagnostic accuracy<sup>9</sup>. For this reason, surgical resection remains both the definitive diagnostic method and the treatment of choice for renal schwannomas<sup>5</sup>.

Histologically, these tumors are composed of cells that typically exhibit elongated or spindle-shaped morphology with wavy contours. They are arranged in two distinct patterns of organization known as Antoni A and Antoni B areas, which may be present in varying proportions; in some cases, the Antoni A pattern is exclusive (cellular schwannoma)<sup>3</sup>. The Antoni A pattern is hypercellular, consisting of spindle cells arranged in fascicles, frequently showing nuclear palisading and Verocay bodies<sup>3</sup>. Antoni B areas are less cellular

and display a greater amount of myxoid extracellular matrix<sup>1,3</sup>. All schwannomas exhibit an immunophenotype characterized by strong and diffuse expression of S100 protein<sup>5</sup>, which is considered specific. Other proteins expressed by this neoplasm include SOX10, vimentin, and GFAP<sup>1,5</sup>.

Partial or radical nephrectomy is the treatment of choice for these tumors<sup>6</sup>. Surgical excision is usually performed because of an initial suspicion of renal cell carcinoma, given the solid appearance of the mass and its enhancement on different imaging modalities<sup>3</sup>.

Percutaneous biopsy played a fundamental role by allowing accurate diagnostic characterization and appropriately guiding therapeutic decision-making for optimal patient management, particularly given the unusual renal location of this lesion and the imaging findings that raised multiple differential diagnoses.

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