

## CASE REPORT

# GIANT RETROPERITONEUM SCHWANNOMA CAUSING IPSILATERAL HYDRONEPHROSIS: A RARE CASE REPORT

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**ABSTRACT:** Retroperitoneal schwannomas are rare benign tumors originating from Schwann cells and are seldom diagnosed preoperatively due to their anatomical location and nonspecific clinical-radiological features. We report a case of 25-year-old woman with progressive right upper abdominal pain for eight months and a palpable right subcostal mass. Imaging showed a large, well-circumscribed, heterogeneously enhancing retroperitoneal lesion displacing right kidney with secondary hydronephrosis. With a provisional diagnosis of ganglioneuroma, laparoscopic complete excision was performed with repair of iatrogenic renal vein rents. Histopathology confirmed benign schwannoma with Antoni A and B areas. Recovery was uneventful, with no recurrence at six months follow-up period.

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**Impact statement:** Current case highlights the diagnostic challenge of retroperitoneal schwannoma, while illustrating the feasibility of laparoscopic complete excision for large lesions with favorable postoperative and short-term oncological outcomes.

**Key words:** Antoni; hydronephrosis; retroperitoneal; schwannoma; Verocay.

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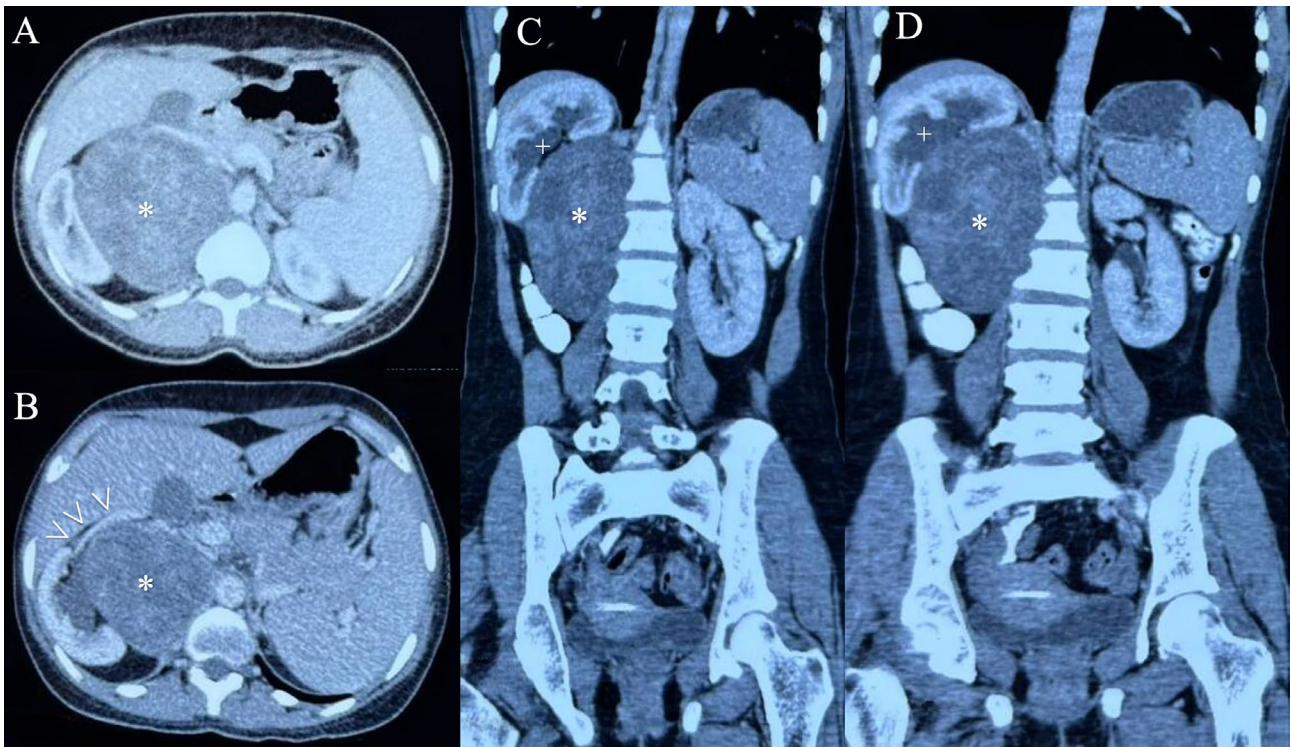
## INTRODUCTION

Schwannomas, also known as neurilemmomas, are slow-growing, benign, encapsulated tumors arising from Schwann cells of neural crest origin (1). Although they are typically benign, malignant transformation has been reported, particularly in association with neurofibromatosis type 1 (NF1) (2). These tumors most commonly occur in the head and neck region and along the flexor surfaces of the extremities. In contrast, retroperitoneal schwannomas (RPS) are exceedingly rare, accounting for approximately 4% of all retroperitoneal tumors and nearly 3% of all schwannomas (3). Owing to their deep anatomical location and nonspecific clinical presentation, diagnosis is often delayed and challenging, with definitive diagnosis usually established only after imag-

ing and surgical excision (1). We report a rare case of RPS managed by laparoscopic excision in a 25-year-old female who presented with right flank and right hypochondrial pain.

## CASE PRESENTATION

A 25-year-old female with no known comorbidities presented to our outpatient department with progressive right upper abdominal pain of 8 months' duration, which had been managed with over-the-counter analgesics. Physical examination revealed a non-tender, globular mass measuring approximately 6 × 6 cm below the right subcostal margin, which did not move with respiration. In view of the uncertain origin of the mass, an ultrasonography of



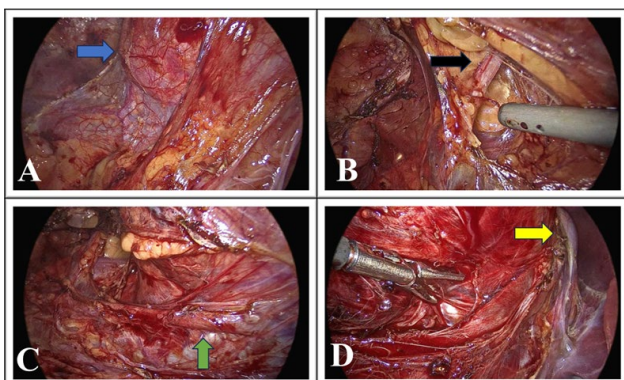
**Figure 1.** Contrast-enhanced CT images depicting the retroperitoneal mass.

**(A, B)** CT Axial sections revealing a heterogeneously enhancing clearly encapsulated solid retroperitoneal mass (denoted by \*) with displacing effects on the surrounding structures like IVC, Liver, Kidney and right renal vessels (denoted by arrow heads). **(C, D)** Coronal reconstruction section of the CT abdomen revealing a retroperitoneal large mass of size  $10 \times 9$  cm (denoted by \*) with displacing effects on the right kidney and hydronephrosis owing to proximal ureteral extrinsic compression (denoted by +).

the whole abdomen was performed, which demonstrated a well-defined heterogeneous hypoechoic lesion in the right retroperitoneum, displacing the right kidney and associated with moderate hydronephrosis. Further evaluation with contrast-enhanced computed tomography (CECT) of the abdomen revealed a well-circumscribed, smoothly margined, heterogeneously enhancing solid mass measuring  $104 \times 92$  mm in the right retroperitoneum, with central non-enhancing necrotic areas. The right kidney and adrenal gland were visualized separately, confirming the retroperitoneal origin of the lesion. The right kidney was compressed and displaced superolaterally with secondary hydronephrosis. The mass was seen abutting the inferior vena cava, duodenum, and ascending colon without evidence of invasion, and no significant lymphadenopathy was noted (**Figure 1**). Given the radiological suspicion of a paraganglioma, a metabolic workup including serum metanephrines, cortisol, and dehydroepiandrosterone was performed and found to be within normal limits. With a provisional diagnosis of ganglioneuroma, the patient was planned for laparoscopic excision of the retroperitoneal mass.

Intraoperatively, a large retroperitoneal mass measuring approximately  $10 \times 10$  cm was identified, abutting the liver and displacing the right kidney superolaterally. The ascending colon, including the hepatic flexure, and the duodenum were mobilized to expose the mass (**Figure 2A**). The right ureter and gonadal vessels were displaced inferolaterally (**Figure 2B**). The right ureter was identified, carefully mobilized, and preserved. The mass was dissected off the underlying psoas muscle, proceeding medially toward the inferior vena cava and superolaterally toward the right kidney after identification of the right renal artery and vein (**Figure 2C,D**). The right renal vessels were found to be closely adherent to the mass, and during dissection, three rents occurred in the renal vein, which were successfully repaired using prolene sutures. After complete mobilization, the specimen was extracted through a right iliac fossa incision. Post-operative course of the patient was uneventful. Drain was clamped on 3<sup>rd</sup> day and removed next day, with the patient being discharged on day 4.

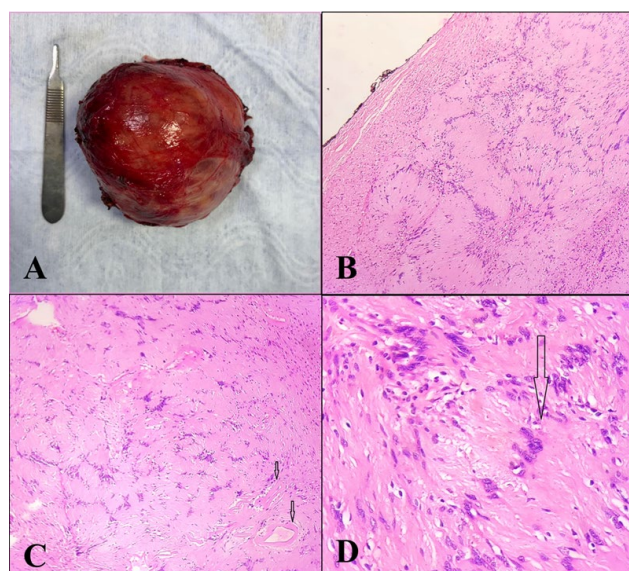
On gross examination, the tumor was greyish white to yellow bosselated in appearance, measuring  $10 \times$



**Figure 2.** Intraoperative anatomical relationships of the retroperitoneal mass.

(A) Inferior aspect of the retroperitoneal mass (blue arrow). (B) Right Ureter identified coursing inferolaterally to the mass (black arrow). (C) Inferior vena cava located medially in relation to the mass (green arrow). (D) Relationship of the mass with the right renal vein (yellow arrow) and right kidney.

9 cm in size, being covered by capsule (**Figure 3A**). The cut surface was predominantly solid, lobulated and firm in consistency, and was whitish to tan-yellow with mucoid to myxoid texture. Areas of necrosis and focal hemorrhage were seen. On microscopic examination, findings were consistent with



**Figure 3.** Gross and histopathological features of the resected specimen.

(A) Gross appearance of the excised encapsulated tumor. (B) Microscopic examination revealing an encapsulated lesion composed of biphasic areas (H & E; 100X). (C) Presence of compact hypercellular Antoni A areas and myxoid hypocellular Antoni B areas along with thickened and hyalinized blood vessels (H & E; 100X). (D) High power view demonstrating Verocay bodies characterized by nuclear palisading around fibrillary process (H & E; 400X).

schwannoma with Antoni A and B areas. Section showed an encapsulated neoplasm composed of cytologically bland spindle cells arranged predominantly in densely cellular areas with nuclear palisading (Verocay bodies) and alternating hypocellular areas. Focal areas showed thick walled hyalinized blood vessels and microcystic change, with no mitosis/atypia seen. The pathological findings were consistent with schwannoma without any signs of malignancy (**Figure 3B-D**). At the 6-month follow-up visit, the patient was asymptomatic with no clinical or radiological evidence of recurrence.

## DISCUSSION

Schwannomas are usually solitary benign tumors, with malignant transformation being exceptional. However, patients with NF1, particularly those harboring plexiform neurofibromas, are at increased risk of developing malignant peripheral nerve sheath tumors, with the reported lifetime risk reaching up to 60% in selected high-risk subgroups (4). Retroperitoneal involvement is rare, accounting for approximately 0.7% of benign schwannomas and 1.7% of malignant schwannomas (5, 6). The age spectrum of RPS is wide, ranging from 6 months to 70 years; however, they are most frequently reported in women between the second and fifth decades of life (7). Although extremely uncommon, schwannomas should be considered in the differential diagnosis of retroperitoneal masses in young patients, with only two cases reported in the adolescent age group to date (1).

Most RPS are detected incidentally, particularly smaller lesions identified during evaluation for unrelated conditions. When symptomatic, clinical presentation is often nonspecific, including low back pain or gastrointestinal and urinary complaints due to mass effect on adjacent organs, which contributes to diagnostic delay (8). Owing to the capacious nature of the retroperitoneum, these tumors frequently attain a large size before detection, typically ranging from 4 to 15 cm, with a reported median size of 8.7 cm (4). Preoperative imaging plays a central role in identifying the origin of retroperitoneal masses despite diagnostic challenges posed by nonspecific symptoms. Ultrasonography and computed tomography are indispensable initial modalities, allowing assessment of tumor location, density, and relationship to surrounding structures. These typically demonstrate a well-circumscribed, heterogeneous mass with peripheral enhancement, suggestive of

a benign lesion. Magnetic resonance imaging (MRI) remains the gold standard due to its superior diagnostic accuracy, characteristically showing hypo- to isointense signals on T1-weighted images and hyperintensity on T2-weighted images. MRI also facilitates better evaluation of origin, vascular involvement and possible extension into the intervertebral foramina, although such extension is rare (9).

Preoperative percutaneous biopsy remains controversial and is generally not recommended because of the risks of hemorrhage and infection, as well as the rare but serious possibility of neoplastic dissemination in the event of a malignant tumor (9). The differential diagnosis of RPS includes several mesenchymal and neuroendocrine neoplasms, such as pheochromocytoma, paraganglioma, liposarcoma, leiomyosarcoma, fibrosarcoma, and ganglioneuroma; therefore, a high index of suspicion and meticulous clinical evaluation are essential (**Table 1**).

Complete surgical excision remains the primary and most effective treatment for schwannoma. However, the necessity of extensive multiorgan resection to achieve negative margins remains debatable. Given that the majority of these tumors are benign, aggressive organ resection may be unwarranted and potentially overtreats the disease. Nevertheless, uncertainty regarding the exact nature of the tumor in the preoperative period often cre-

ates a dilemma, necessitating complete excision to exclude malignancy. To date, no clear consensus has been established. Radical excision of RPS is technically challenging because of the tumor's hypervascularity and its close adherence to major retroperitoneal vessels, which increases the risk of intraoperative bleeding (10).

A definitive diagnosis of schwannoma is established on histopathological examination of the resected specimen. Grossly, RPS are typically solitary, well-circumscribed, encapsulated, and bosselated masses. Histopathological examination reveals Schwann cells arranged in alternating compact hypercellular Antoni A areas and hypocellular myxoid Antoni B areas, with strong immunoreactivity for S-100 protein. Secondary degenerative changes such as hyalinization, calcification, cystic degeneration, and fibrosis characterize the variant known as "ancient schwannoma" (8). The prognosis of RPS is excellent, with minimal risk of recurrence following complete resection. Recurrence rates of 5-10% have been reported after incomplete excision, underscoring the importance of complete tumor removal (1, 3). This highlights the need for a systematic postoperative follow-up protocol, although no standardized guidelines currently exist. Most centers perform imaging at 6-12 months postoperatively; however, follow-up practices vary between institutions.

**Table 1.** Differential diagnosis of retroperitoneal schwannoma: comparative morphological, immunohistochemical, and molecular features of selected retroperitoneal neoplasms.

ENTITY	MORPHOLOGICAL FEATURES	IMMUNOHISTOCHEMISTRY	MOLECULAR FEATURES
Schwannoma	Encapsulated; Antoni A and B areas; Verocay bodies; nuclear palisading	S100+, SOX10+; CD34-; SMA-; Desmin-	NF2 alterations occasionally present
Malignant Peripheral Nerve Sheath Tumor	Hypercellular spindle cells; pleomorphism; necrosis; high mitotic activity	Focal/weak S100, SOX10; loss of H3K27me3	NF1 mutations; CDKN2A, SUZ12, EED alterations
Solitary fibrous tumor	Patternless spindle-cell proliferation; staghorn vessels	STAT6+, CD34+, BCL2+	NAB2-STAT6 fusion
Leiomyosarcoma	Intersecting fascicles of eosinophilic spindle cells with atypia	SMA+, Desmin+, h-caldesmon+	Complex karyotype
Dedifferentiated liposarcoma	Adipocytic component with atypical stromal cells and non-lipogenic sarcoma	MDM2+, CDK4+	MDM2 amplification
Fibrosarcoma	Herringbone architecture	Vimentin+; nonspecific	Complex genetic abnormalities
Ganglioneuroma	Mature ganglion cells within Schwannian stroma	S100+, Synaptophysin+, NSE+	No characteristic alteration
Paraganglioma	Zellballen pattern; sustentacular cells	Chromogranin+, Synaptophysin+, S100 in sustentacular cells	SDHB/SDHD, RET, VHL mutations

## CONCLUSIONS

RPS are uncommon tumors that present significant diagnostic challenges due to their variable and non-specific clinico-radiological features. Surgical excision remains the treatment of choice, providing both definitive histopathological diagnosis and cure. While complete resection is desirable, a conservative surgical approach should be considered whenever feasible, given the predominantly benign nature of these lesions. Early recognition and appropriate follow-up are essential to ensure optimal outcomes.

## COMPLIANCE WITH ETHICAL STANDARDS

### Funding

None.

### Conflicts of interest

The authors declare no competing interests.

### Availability of data and materials

The data that support the findings of this study are not publicly available due to institutional ethical regulations but are available from the corresponding author upon request.

### Authors' contributions

BS, SY, HS: conceptualization. BS, HS, TKA: data curation. BS, HS, PP: formal analysis. BS, SY, TKA, HS: writing – original draft. SY, HS, TKA, PP: writing – review & editing. PP: resources.

### Ethical approval

#### Human studies and subjects

Institutional Ethics Committee approval was not required for publication of this single case report. Written informed consent was obtained from patient included in the study. Consent was also obtained for publication of clinical details and images.

### Publications ethics

#### Plagiarism

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### Data falsification and fabrication

The authors declare that no data fabrication, falsification, or manipulation has been carried out in the preparation of this work.

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