Case Report
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A Rare Case of Retroperitoneal Schwannoma in an Adult Male

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Abstract

Objective. This study aims to illustrate a rare case of retroperitoneal schwannoma by presenting the clinical, imaging, and histological parameters. **Case Report.** A 36-year-old patient visited the outpatient clinic because of back pain experienced over the previous two months. There were no complaints regarding the nervous system or urinary system. Thorough imaging evaluation, including magnetic resonance for the lumbar spine, abdominal computed tomography, and positron emission tomography was conducted. An encapsulated mass was found in the retroperitoneal area, positioned in front of the O4 vertebra and in close proximity to the left psoas muscle, the left common iliac artery, and the left ureter. The lesion exhibited FDG radioisotope uptake, and a CT-guided biopsy confirmed a benign peripheral nerve tumor. The patient underwent laparotomy surgery, where the tumor was removed. The histological investigation, along with immunohistochemistry, confirmed the presence of a retroperitoneal schwannoma. **Conclusion.** Schwannoma is a rare type of retroperitoneal tumor, with nonspecific clinical and radiological characteristics that make diagnosis difficult. Surgical resection is the primary treatment for symptomatic patients, with a favorable prognosis. Long-term follow-up is advised to reduce the chance of late recurrence.

Key Words: Retroperitoneal Schwannoma ▪ Nerve Sheaths ▪ Schwann Cells ▪ Retroperitoneal Tumor ▪ Case Report.

Introduction

Schwannoma, or neurilemmoma, is a rare ectodermal tumor that primarily originates from the sheaths of peripheral or cranial nerves. Retroperitoneal schwannomas are a very uncommon variety, making up about 3% of all schwannomas and 4% of retroperitoneal tumors (1). The sporadic form of retroperitoneal schwannomas is the most common, and predominantly affects females between the second and fifth decades of life (2). The familial form of retroperitoneal schwannomas accounts for 5-18% of all retroperitoneal schwannomas, presents at a younger age, and is often associated with Von Recklinghausen's disease (3). Clinical manifestations are nonspecific, and diagnosis relies mainly on histopathological examination. Prognosis is favorable following surgical resection, however, there is a 5-10% risk of late recurrence in cases of incomplete excision, necessitating long-term follow-up (1).

Here, we present a case report of an uncommon location of schwannoma in a 36-year-old male. The patient was presented to our department due to a two-month history of back pain. The schwannoma was successfully treated using conventional surgery.

Case Presentation

A 36-year-old male was referred to our surgical clinic due to the existence of a retroperitoneal

mass. The pathology department of the hospital discovered this mass during a thorough imaging evaluation in response to a reported two-month history of back pain. It is important to mention that there were no documented issues with urinating or symptoms related to the lower extremities. The patient's medical history, clinical examination and laboratory evaluations were otherwise unremarkable. A lumbar spine Magnetic Resonance Imaging (MRI) scan revealed a well-circumscribed lesion in the left retroperitoneal space, adjacent to the left iliopsoas muscle. The lesion exhibited low signal intensity in T1 weighted images (Figure 1, panel A) and high signal intensity in T2 weighted images (Figure 1, panel B).

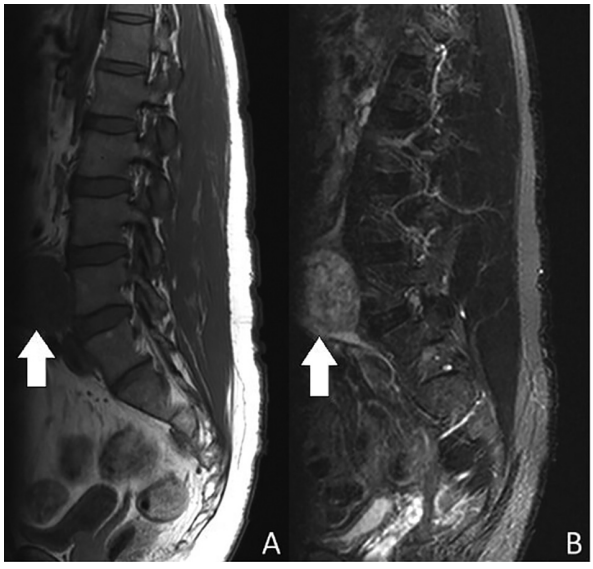

Figure 1. Sagittal lumbar MRI without contrast. A well-circumscribed lesion with signal isointense to the muscle in T1 weighed images (panel A) and hyperintense signal in T2 weighed images (panel B).

An abdominal Computed Tomography (CT) scan demonstrated a 52×47 mm mass with heterogeneous contrast enhancement, located anterior to the L4 vertebra, abutting the left ureter and left common iliac artery. Brain and chest CT scans were unremarkable (Figure 2).

The lesion exhibited fluorodeoxyglucose (FDG) avidity (SUVmax 5.2) on Positron Emission Tomography (PET-CT). A CT-guided core needle biopsy was performed, and histopathological analysis of the specimen revealed a nodular mass surrounded by a fibrous capsule, with areas of mixed cellularity and the presence of thrombi in the blood vessels. Immunohistochemistry of the biopsy tissue suggested a benign nerve sheath tumor.

The decision to operate was based on the patient being symptomatic. Beyond the surgical issue, no concurrent diseases were discovered during the routine preoperative examination. During the surgical procedure, the patient had both general and epidural anesthesia. A midline incision was conducted, and a thorough examination of the liver and peritoneal cavity revealed

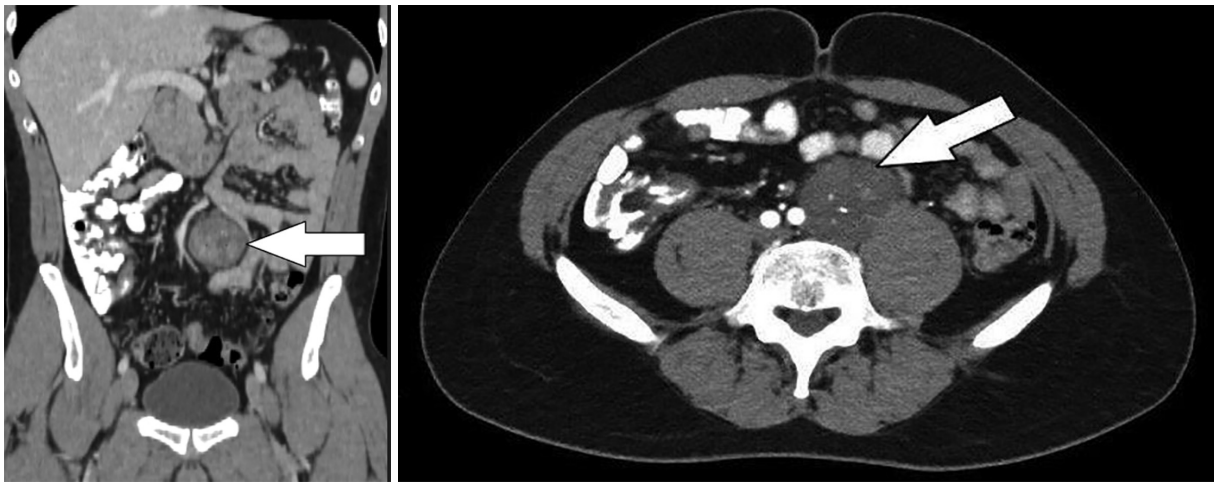

Figure 2. Computed tomography (CT) of the abdomen with intravenous contrast in coronal (A) and axial (B) sections. (A-B) A well-circumscribed mass with heterogeneous contrast enhancement, located anterior to the L4 vertebra, abutting the left ureter and left common iliac artery.

no pathological abnormalities. The posterior peritoneum was incised, we accessed the retroperitoneal area and detected the mass. The left ureter was located superior and lateral to the structure, with outward displacement. The left common iliac artery was situated within it and to its right. The left psoas muscle was positioned posterior to it. Subsequently, we surgically accessed the tumor and securely ligated the lumbar vessels located superior to it. The process entailed surgically removing the tumor and establishing thorough control of bleeding (Figure 3).

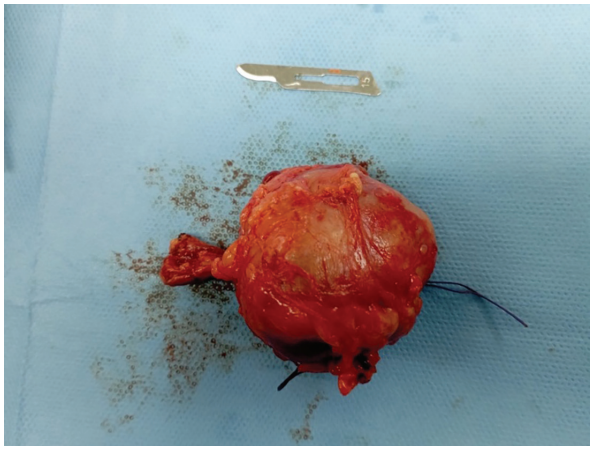

Figure 3. The surgical specimen. Macroscopic examination of the surgical specimen revealed a well circumscribed tumor, encased in a fibrous capsule and surrounded by fatty tissue.

The abdomen was closed in layers. Histopathology of the resected specimen confirmed the diagnosis of a retroperitoneal schwannoma, with negative margins and no evidence of nerve or blood vessel infiltration. Histologically, the lesion consisted of hypercellular (Antoni A areas, Figure 4-panel A and B) and hypocellular areas (Antoni B areas, Figure 4-panel A). The immunohistochemistry was suggestive of a benign peripheral nerve sheath tumor. The cells stained diffusely positive for S100 and vimentin. The patient was discharged on postoperative day 8. He did not receive any adjuvant therapy, and was scheduled for follow up with serial MRI scans for a minimum of five years. At the 12-month follow-up, the patient had not developed any recurrence, his condition remained satisfactory, and he reported an excellent quality of life.

Discussion

Peripheral nerve schwannomas are neuroectodermal neoplasms with an annual prevalence of 6 per 1,000,000 people (4). They are the most common peripheral nerve sheath tumor in adults and arise from Schwann cells (1). The retroperitoneal location of a schwannoma is exceedingly rare, accounting for only 3% of all schwannomas (2, 3). The sporadic variant of retroperitoneal

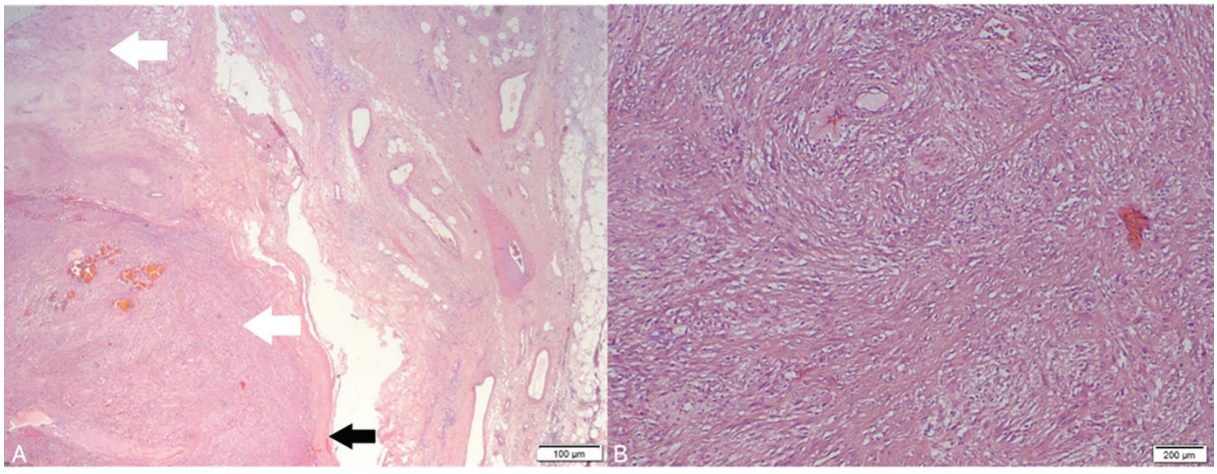

Figure 4. Histopathology of the surgical specimen. A well-circumscribed nodular mass, encased in a fibrous capsule (panel A - thin black arrow). The lesion consists of hypercellular areas (Antoni A areas- bottom white arrow), and hypocellular areas (panel A - Antoni B areas - top white arrow) (magnification ×100). Microscopic image of a hypercellular – Antoni A area (panel B) (magnification × 200).

schwannoma demonstrates the highest occurrence rate during the second to fifth decades of life, displaying a slight predominance in females (4, 5). Conversely, the familial form manifests at an earlier age, exhibiting a higher likelihood of malignancy (with reported rates of up to 60% of cases), and a strong association with von Recklinghausen's disease (3). Schwannomas, which are usually benign growths that develop from Schwann sheath cells, mostly impact nerves in the head and upper limbs. Schwannomas primarily originate from neural crest cells. Except for cranial nerves I and II, which do not have Schwann cells, schwannomas can develop in any other organ or nerve trunk (6). Schwannomas can arise in any anatomical region of the body, with the limbs being the most common site in 53.1% of cases, followed by the trunk in 13% of instances, and the head and neck in 13.9% of cases (7). Additional sites include the posterior mediastinum, retroperitoneum, spinal roots, bone, gastrointestinal tract, pancreas, liver, thyroid, adrenal glands, and lymph nodes (8).

Table 1. The Characteristics of Retroperitoneal Schwannomas in Our Case Report and Cases in Pubmed

Characteristics	Our case report	Cases report in Pubmed (N=28)
		N; (%)
Symptom		
None	-	5 (18)
Abdominal pain	-	16 (57)
Back pain	Yes	12 (42)
Urinary symptoms	-	18 (64)
Neurological symptoms	-	8 (29)
Preoperative imaging		
US*	-	22 (79)
CT†	Yes	15 (54)
MRI‡	Yes	12 (42)
PET /CT§	Yes	4 (14)
Preoperative biopsy	Yes	13 (46)
Surgery method		
Abdominal resection	Yes	13 (46)
Laparoscopy resection	-	15 (54)

* Ultrasound; †Computed Tomography; ‡Magnetic Resonance Imaging; §Positron Emission Tomography/ Computed Tomography.

A total of 28 cases of retroperitoneal schwannoma were retrieved from the PubMed database (1-3, 5-7, 9-16). These items are documented in the supporting information (Table 1).

Out of the total of 28 patients, the larger portion, specifically 18 individuals, did not exhibit any symptoms. Neurological problems were reported by only eight patients. In general, the predominant method of assessment for patients was ultrasound, with 22 out of 28 patients undergoing this procedure. CT was used for 15 patients, while MRI was used for 12 individuals. A PET scan was employed as a preoperative evaluation method for the tumor in just four cases. Fewer than half of the patients (13/28) had preoperative biopsy, as in our case. Out of the total of 28 patients, 13 patients underwent abdominal resection, while the remaining 15 patients had laparoscopic tumor excision.

Retroperitoneal schwannomas are typically benign and slow-growing in nature (5). They are mostly asymptomatic; however, they can present as a palpable mass, or cause abdominal or back pain, urinary dysfunction, or bowel obstruction due to their location (5, 11, 17). On imaging, retroperitoneal schwannomas often appear as encapsulated lesions with low density and heterogeneous contrast enhancement on CT scans (11, 17). On MRI scans they appear isointense to the muscle on T1-weighted images, and exhibit high signal intensity (similar to fat tissue) on T2-weighted images (2, 18).

Differential diagnosis of retroperitoneal schwannoma includes a wide spectrum of retroperitoneal lesions, including: malignant peripheral nerve sheath tumors, sarcoma, lymphoma, neuroendocrine tumors (pheochromocytoma, paraganglioma), vascular tumors (hemangioma, cystic lymphangioma), fatty tissue tumors (angiomyolipoma, myelolipoma, lipoma), rhabdomyoma, or extragonadal tumors (teratoma or seminoma), among others (9). The need for establishing a diagnosis by means of a preoperative biopsy has been a matter of debate (12). The similarity in the clinical and imaging characteristics between schwannomas and other lesions is countered by the risk of tumor seeding, hemorrhage, or a hypertensive

crisis during a biopsy attempt (5). Nevertheless, in the appropriate clinical setting, a preoperative core needle biopsy of a retroperitoneal mass, in order to guide further treatment decisions, is in accordance with the European Society for Medical Oncology (ESMO), the National Cancer Comprehensive Network (NCCN) and the Trans- Atlantic Retro Peritoneal Sarcoma Working Group (TARPSWG) Guidelines (19).

Surgical excision is the mainstay of treatment for all symptomatic patients, and is generally associated with a favorable long- term prognosis. There have been no reports of retroperitoneal schwannoma with distant metastases, although local recurrence rates of 5-10% have been reported, especially in cases of incomplete resection (9, 10). Malignant transformation is exceedingly rare, but it has been reported in cases of von Recklinghausen's disease (5). Long-term follow-up remains necessary to minimize the slight risk of late recurrence and malignant transformation.

At this juncture, we also note that our article serves as another instance illustrating the limited efficacy of Fluorodeoxyglucose Positron Emission Tomography (FDG PET) in differentiating schwannomas from malignant peripheral nerve sheath tumors (20). Due to the FDG uptake observed in schwannomas, it is not feasible to differentiate these tumors from malignant peripheral nerve sheath tumors, which also exhibit high FDG uptake, prior to undergoing a biopsy or surgery (21, 22).

Conclusion

Retroperitoneal schwannomas constitute a small percentage of retroperitoneal tumors and present a diagnostic challenge due to their nonspecific clinical and imaging features. A multidisciplinary approach and early tissue biopsy are required. Surgical resection is the primary treatment modality for symptomatic patients, with an overall favorable prognosis. However, long-term follow-up is recommended to mitigate the risk of late recurrence.

What Is Already Known on This Topic:

Schwannomas are the predominant neoplasms of the peripheral nerve sheath in adults, originating from Schwann cells. Their vague clinical and imaging features make them difficult to diagnose. Schwannomas located in the retroperitoneum are extremely uncommon, accounting for only about 3% of all schwannomas. The necessity of confirming a diagnosis with a biopsy before surgery has been a subject of discussion. The definitive diagnosis relies on the histopathological examination of the operative specimen. Surgical excision is the primary treatment for all individuals experiencing symptoms, and is typically linked to a positive long-term prognosis.

What This Study Adds:

This study offers thorough insights into the imaging and histological diagnosis and management of these uncommon tumors. The preoperative imaging, which encompassed both CT and MRI scans, afforded us a meticulous depiction of the anatomical components encompassing the tumor. In conjunction with the CT-guided core needle biopsy, this played a pivotal role in the favorable outcome of the surgery. The paper showcases the restricted efficacy of Fluorodeoxyglucose Positron Emission Tomography (FDG PET) in differentiating schwannomas and malignant peripheral nerve sheath tumors. Histology and immunohistochemistry markers were used to identify and study this condition, offering insights for future research in this area. Our primary objective is to enhance the limited body of research on retroperitoneal schwannomas by emphasizing the significance of being cognizant and doing comprehensive assessments in similar cases.

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Conflicts of interest: The authors declare that they have no conflict of interest.

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