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"ETIOLOGÍA DE LA PANCREATITIS AGUDA GRAVE Y MORTALIDAD EN PACIENTES DEL HGZ No.46."

**"ETIOLOGY OF SEVERE ACUTE PANCREATITIS AND MORTALITY
IN PATIENTS AT HGZ NO. 46."**

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Retroperitoneal synovial sarcoma a diagnostic challenge: an uncommon case

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INTRODUCTION

Synovial sarcoma (SS) is a rare and aggressive mesenchymal neoplasm that can arise at various anatomical sites. However, it typically occurs in the soft tissues of the extremities, particularly the lower limbs [1]. This tumor makes up 0.8%-10% of soft tissue sarcomas and about 1% of reported retroperitoneal tumors. [2,3,4]. Its highest incidence is observed in young adults, with no clear differences by sex. Because of its low frequency, epidemiological data are primarily derived from isolated reports and small series. [2,3,4,5]. Although its name suggests a synovial origin, the tumor does not develop from the synovial membrane; the term is kept because of morphological similarities to embryonic synovium [6,7]. Its diagnosis is difficult, not only because it can resemble benign lesions, but also because of morphological overlap with other sarcomas. When it occurs in the retroperitoneum, its detection is often delayed because this anatomical space allows for progressive tumor growth without specific symptoms. Retroperitoneal lesions can resemble other aggressive neoplasms; this complicates differential diagnosis and leads to significant treatment delays, which negatively affect prognosis. In this context, most retroperitoneal cases are detected at advanced stages and are linked to lower survival rates, estimated at 20%-29% at five years [4, 6]. Retroperitoneal SS is extremely rare, with fewer than 35 cases documented in the literature [4,5]. Spinal involvement is exceptional, and most reports lack or have limited follow-up [2]. Due to the absence of standardized guidelines for retroperitoneal SS—the National Comprehensive Cancer Network (NCCN), the European Society for Medical Oncology (ESMO), and the American Society of Clinical Oncology (ASCO) include this tumor within the general management of soft tissue sarcomas—its approach, diagnosis, and treatment pose a challenge for surgeons [1,2].

Keywords: synovial sarcoma, retroperitoneum, mesenchymal neoplasm, differential diagnosis, and prognosis.



Sarcoma sinovial retroperitoneal: un reto diagnóstico: un caso poco común

ABSTRACT

El sarcoma sinovial (SS) es una neoplasia mesenquimal rara y agresiva que puede surgir en diversos sitios anatómicos. Sin embargo, típicamente ocurre en los tejidos blandos de las extremidades, particularmente en las extremidades inferiores [1]. Este tumor representa el 0.8%-10% de los sarcomas de tejidos blandos y aproximadamente el 1% de los tumores retroperitoneales reportados [2,3,4]. Su mayor incidencia se observa en adultos jóvenes, sin diferencias claras por sexo. Debido a su baja frecuencia, los datos epidemiológicos provienen principalmente de informes aislados y series cortas [2,3,4,5]. Aunque su nombre sugiere un origen sinovial, el tumor no se desarrolla a partir de la membrana sinovial; el término se mantiene debido a las similitudes morfológicas con la membrana sinovial embrionaria [6,7]. Su diagnóstico es difícil, no solo porque puede parecerse a lesiones benignas, sino también por la superposición morfológica con otros sarcomas. Cuando ocurre en el retroperitoneo, su detección a menudo se retrasa porque este espacio anatómico permite el crecimiento progresivo del tumor sin síntomas específicos. Las lesiones retroperitoneales pueden parecerse a otras neoplasias agresivas; esto complica el diagnóstico diferencial y conduce a retrasos significativos en el tratamiento, lo que afecta negativamente el pronóstico. En este contexto, la mayoría de los casos retroperitoneales se detectan en etapas avanzadas y están vinculados a tasas de supervivencia más bajas, estimadas en 20%-29% a los cinco años [4, 6]. El SS retroperitoneal es extremadamente raro, con menos de 35 casos documentados en la literatura [4,5]. La afectación espinal es excepcional, y la mayoría de los informes carecen de seguimiento o tienen un seguimiento limitado [2]. Debido a la ausencia de pautas estandarizadas para el SS retroperitoneal (la National Comprehensive Cancer Network [NCCN], la European Society for Medical Oncology [ESMO] y la American Society of Clinical Oncology [ASCO] incluyen este tumor dentro del manejo general de los sarcomas de tejidos blandos), su abordaje, diagnóstico y tratamiento representan un desafío para los cirujanos [1,2].

Palabras clave: sarcoma sinovial, retroperitoneo, neoplasia mesenquimal, diagnóstico diferencial y pronóstico.

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CASE PRESENTATION

A 49-year-old male patient with a history of classic seminoma with an atypical component, treated at age 34 in 2009 and currently in complete remission, with no other significant oncological history or comorbidities. He consulted the Instituto Nacional de Cancerología (INC) (Bogotá, Colombia) in 2025 due to a six-month history of progressive left dorso-lumbar pain radiating to the inguinal region and the ipsilateral lower limb, along with intermittent episodes of paresthesia.

On physical examination, the obese patient (120 kg, 1.75 m), with increased subcutaneous fat, had a ventral hernia M2M3W2 from a previous laparotomy, and no evidence of a palpable abdominal or lumbar mass. There was no edema in the lower extremities, no signs of peripheral vascular compression, nor loss of strength or neurological alterations.

Outside the INC, it was considered a relapse of retroperitoneal lymph node after initial oncological pathology; however, because of the discrepancy between the clinical presentation and the timing, he was referred to the Breast and Soft Tissue Surgery team.

Imaging studies were ordered to characterize the lesion and assess its extent. A contrast-enhanced computed tomography (CT) scan of the abdomen and pelvis showed a large left retroperitoneal mass in the para-aortic infrarenal space, measuring 71 x 39 x 43 mm. The lesion displaced the duodenum and aorta to the right, medializing the retroperitoneal structures. In addition, a magnetic resonance imaging (MRI) scan of the lumbosacral spine revealed a solid mass involving the psoas muscle from L2 to L4 and in close contact with the L3 vertebral foramen, resulting in a pathological fracture with 50% wedging of the vertebral body (Figures 1 and 2).

Given the complex location and bone involvement, two inconclusive image-guided biopsies were needed; ultimately, the histopathological report was consistent with SS.

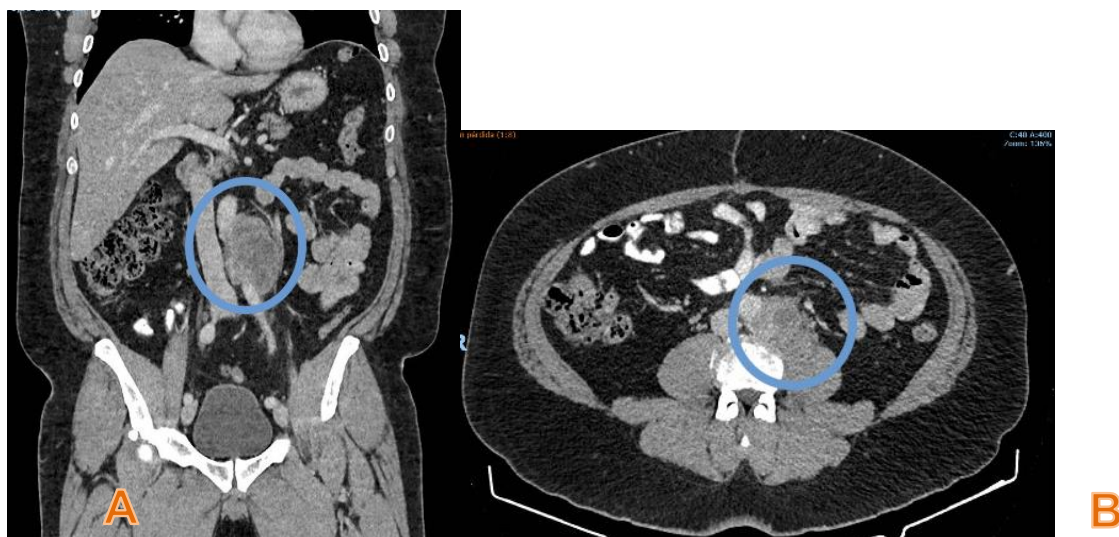


Figure 1. Initial CT scan of the abdomen and pelvis. A. Coronal view: A lobulated mass is visible, with necrotic changes inside it, near the infrarenal aorta. B. Axial view: Multilobulated mass exhibiting heterogeneous changes within it, with infiltration of the psoas muscle and the L3 vertebral body. Images courtesy of INC.



Figure 2. Initial contrast-enhanced lumbar MRI. A. Sagittal view: Tumor lesion with myxoid or cystic changes of lesser intensity in relation to the heterogeneous component. Displacement of intestinal loops anteriorly and close contact with paravertebral muscles. B. Axial view: Rounded, lobulated mass with heterogeneous signal; occupation of the neural foramen is observed, extending into the spinal canal and displacing the nerve root. In close contact with the edge of the vertebral pedicle.

With the diagnosis established, the case was referred to a multidisciplinary team (Clinical Oncology, Radiation Oncology, Surgical Oncology, Breast and Soft Tissue Surgery, Neurosurgery, Vascular Surgery, Radiological Oncology, and Pathological Oncology). The SS was considered greater than 5

cm, with borderline resectability criteria due to vertebral involvement (L3) and close contact with the duodenum, infrarenal aorta, and left iliac vessels. Therefore, the team decided on neoadjuvant management with MAI chemotherapy (mesna, adriamycin, ifosfamide) for three cycles, followed by image-guided radiation therapy (IGRT), delivering 55 Gy in five fractions on alternate days. In a new multidisciplinary team assessment, the follow-up CT scan showed a reduction in tumor volume ($65 \times 37 \times 45$ mm), with no significant changes in lumbar involvement or vascular contact (Figure 3). Therefore, it was determined that the optimal treatment was a wide surgical resection with simultaneous control of vertebral involvement, and a two-stage procedure was planned along with the Neurosurgery service.



Figure 3. Post-neoadjuvant abdominal CT scan. Coronal view: Solid, heterogeneous retroperitoneal mass. Hypodense areas indicative of necrosis are observed within it, with the contrast medium enhancing the solid component. It displaces the duodenum and intestinal loops anteriorly and superiorly. It is in close contact with the abdominal aorta without compromising the inferior vena cava.

In the first surgical stage, a left para-aortic mass measuring 12×14 cm was identified, with desmoplastic changes secondary to neoadjuvant therapy. It was in proximity to the infrarenal aorta, the left iliac vessels, and the left renal pedicle, without vascular infiltration. However, there was infiltration of the left ureter in its distal third, without a cleavage plane, and also with involvement of the distal psoas, the first portion of the jejunum (fixed loop), and the L3 vertebral body.

To achieve the oncological goal, the lesion was meticulously mobilized, dissecting the retroperitoneal planes while preserving major vascular structures. This involved a partial duodenectomy, resection of

the iliopsoas muscle, and, during the same surgical procedure, an L3 corpectomy performed by Neurosurgery. The mass was resected en bloc, ensuring wide macroscopic margins and taking out compromised bone fragments associated with the pathological fracture. No significant hemorrhagic events or iatrogenic injuries occurred during the procedure (Figure 4).

Subsequently, in a second surgical procedure eight days after surgery, the duodenal anastomosis was revised, and the neurosurgery team completed the L3 corpectomy through the same initial approach. Following resection of the affected vertebral body, spinal reconstruction was performed with arthrodesis and bone graft placement—a 45 mm titanium lumbar cylinder (Johnson & Johnson) filled with 6G bone graft—with proper fluoroscopic verification, restoring mechanical stability (Figure 5). The patient was transferred to the Intensive Care Unit for recovery under close neurological monitoring. No new deficits or immediate complications were observed.

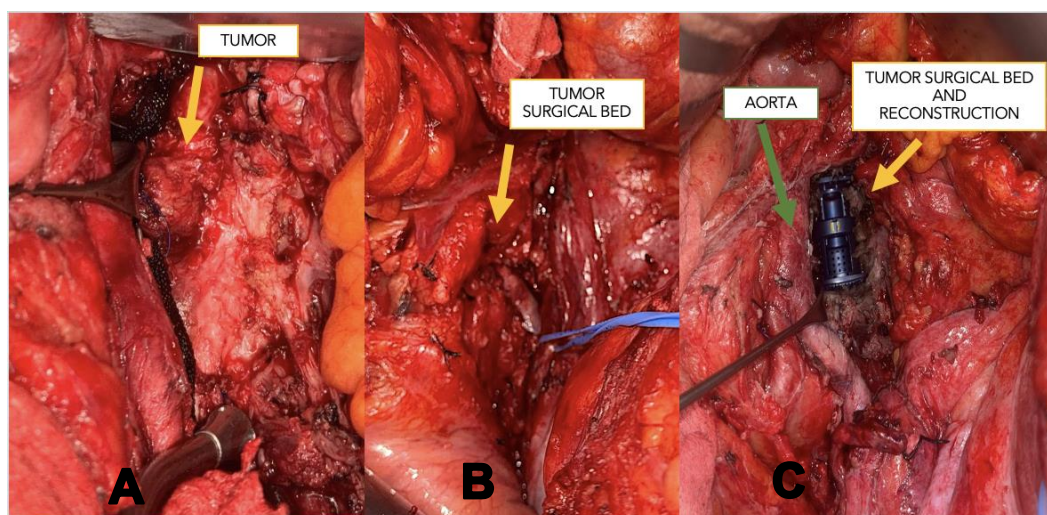


Figure 4. Intraoperative findings. A. Intraoperative image showing the retroperitoneal tumor (arrow), solid and lobulated in appearance, adhered to deep planes and close to vascular structures and the psoas muscle in the retroperitoneum. B. Tumor surgical bed after complete resection of the retroperitoneal tumor (arrow). The deep planes are clear, with adequate exposure of retroperitoneal structures and no macroscopic residual mass. C. Tumor surgical bed after resection of the retroperitoneal sarcoma, showing the exposed abdominal aorta (green arrow) and the site of vascular and tissue reconstruction in the area of the resected tumor (yellow arrow). Titanium fixation hardware is visible in the vertebral body.

Postoperative CT imaging showed adequate vertebral reconstruction, correct positioning of the arthrodesis material, and no fluid collections, residual bleeding, or involvement of nearby structures. Subsequent clinical evaluation confirmed progressive improvement in low back pain and resolution of paresthesias that had prompted the initial consultation.

The patient was discharged with clear instructions for physical rehabilitation and close follow-up by the Oncology, Soft Tissue Surgery, and Neurosurgery services.

Surgical pathology was consistent with a monophasic synovial sarcoma measuring $11 \times 4 \times 3.3$ cm, with 40% residual viable cells and 60% hyalinization/sclerosis (Figure 5). The superior, inferior, and anterior surgical margins were positive (R1); the lateral and medial margins were 0.2 cm and 0.4 cm from the tumor, respectively, with no lymphatic, vascular, or neural invasion. Immunohistochemistry showed that the tumor cells were positive for CD99, TLE1, and CK AE1/AE3, negative for SALL4, OCT4, EMA, and CD30, and had a Ki-67 index of 20%.

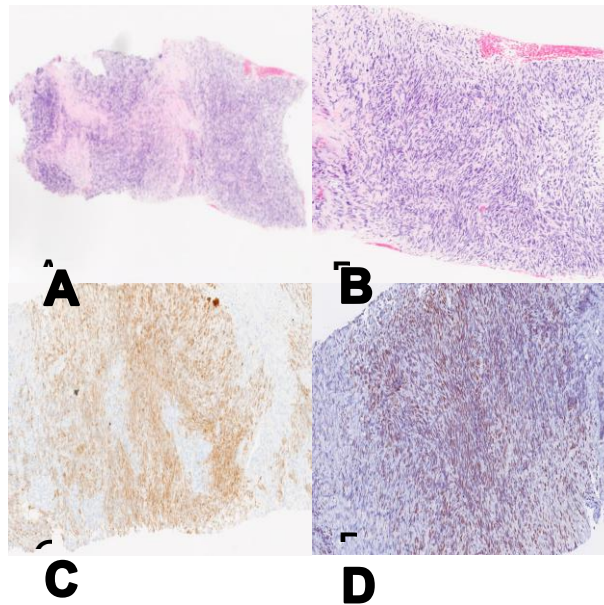


Figure 5. Pathology. A-B. Hematoxylin-eosin stain. A mesenchymal neoplasm consisting of spindle cells with slight atypia arranged in fascicles is observed. Few mitoses are seen. No necrosis is present. C. Focal staining for AE1/AE3. D. Nuclear staining for TLE1.

The patient was evaluated by the Clinical Oncology service, and follow-up imaging was recommended due to the risk of local and distant recurrence given planned R1 margins, with no indication for adjuvant chemotherapy. Systemic management was advised only in the event of progression. At the time of writing, the patient has shown no signs of local, regional, or distant recurrence and remains under follow-up with clinical improvement.

Patient perspective and informed consent

From the patient's perspective, he reported significant improvement in symptoms and functional recovery after surgery, expressing satisfaction with the treatment received and, in particular, grateful for the absence of neurological deficits after the intervention.

The patient provided full informed consent for the surgical procedure that included an explanation of the major neurological and vascular risks. He also signed a specific consent form authorizing the publication of his clinical case and diagnostic images, in accordance with international ethical standards and the CARE guidelines.

DISCUSSION

Synovial sarcoma is a rare malignant tumor that arises in mesenchymal tissues and commonly affects the extremities of young adults. It makes up 5–10% of soft tissue tumors, with an estimated incidence of 1 to 1.5 cases per million people annually [3,8]. Its exact incidence remains uncertain due to its low frequency, and most available data are from case reports or small series [3,6].

Primary retroperitoneal presentation is rare, accounting for less than 1% of sarcomas in this location, which was first described in 1954 by Pack and Tabah [3,5,9,10]. This rarity poses significant diagnostic challenges, as the anatomical depth of the retroperitoneum allows the tumor to grow silently until advanced stages, when vascular or nerve structures are already affected, or neurological pain symptoms appear, as in this case [3].

Differential diagnoses include liposarcoma, leiomyosarcoma, and undifferentiated pleomorphic sarcoma [3, 6]. As in this case, it is important to consider the patient's oncological history to adopt a comprehensive approach and exclude other retroperitoneal pathologies with lymph node involvement, such as lymphoma, seminoma, or tumors of gynecological or urothelial origin.

Clinical presentation

The interval between symptom onset and diagnosis can range from 6 months to 10 years [2]. Retroperitoneal SS is often diagnosed late, which worsens the prognosis and poses a challenge for the surgeon.

The most common symptoms include chronic abdominal or lower back pain, radiation to the extremities, sensory or motor disturbances, a palpable mass, weight loss, or symptoms caused by compression of structures such as the bowel, urinary system, or kidneys [2,9]. The study by Yang et al. [2] found that 81.25% of participants had neurological deficits.

Imaging studies

Imaging studies are crucial for characterizing these lesions in order to guide diagnosis, evaluate resectability, and determine the presence of metastatic involvement [4,6]. The combination of CT and MRI helps define the tumor's relationship to solid organs, vascular structures, and the spine, which is essential for surgical planning [3,4].

Features that help identify SS on imaging include intratumoral hemorrhage and the presence of calcifications. On CT scans, calcifications are seen in 27-47% of cases and can also reveal areas of active bleeding and bone involvement [6–9].

Regarding MRI, it allows identification of a heterogeneous mass with solid or cystic components, caused by hemorrhage or fibrosis [3–5]. It is important to note that the absence of an intratumoral lipomatous component on CT or MRI is a key sign for the differential diagnosis between SS and liposarcoma [1,6]. In this patient, the lesion's characteristics, infiltration of the L3 vertebral body, and associated pathological fracture indicated a locally aggressive behavior rarely described in the literature, which is consistent with high-grade tumors and rules out differential diagnoses such as lymphoma or regional recurrence of seminoma in this case. Similarly, bone involvement has been reported only sporadically in the literature, which could be considered a borderline resectability factor and a poor prognostic indicator for treatment planning.

Histopathology

The definitive diagnosis requires histopathological confirmation and an immunohistochemical panel. The morphology of spindle-shaped cells arranged in fascicles strongly indicates monophasic SS. This type of tumor is often confused with other mesenchymal tumors such as malignant fibroepithelial tumors, solitary fibrous tumors, and sarcomatoid carcinomas, among others. Therefore, immunohistochemical and cytogenetic studies are necessary to distinguish SS [4].

Histologically, there are three main subtypes: poorly differentiated, biphasic, and monophasic spindle cells, the latter being the most common subtype in SS [1, 6,11]. The monophasic spindle cell type consists of a matrix made up of relatively small spindle cells with a uniform, oval, short nucleus and long vacuolar structure. The chromatin is evenly dispersed, the nucleus is not prominent, and the cytoplasm is rarely eosinophilic [1, 6]. Immunohistochemistry typically shows epithelial membrane antigen (EMA) expression and diffuse Bcl-2 and CD99 expression [1].

Confirmatory diagnosis is achieved through translocation (x;18)(p11;q11) tests, since the SS18-SSX fusion type is critical due to its high sensitivity and specificity for SS [6,8]. For these tests, the use of next-generation sequencing (NGS) panels is recommended, as they can help distinguish between SS18-SSX1 and SS18-SSX2 [10,11]. While there is no strong consensus on the link between fusion subtype

and outcomes, histological grade has been identified as a potentially more significant prognostic factor [3,8,12].

Multiple studies have identified prognostic factors associated with survival, including age, tumor size, surgical margins, mitotic index, bone or neurovascular invasion, histological subtype, p53 overexpression, and Ki-67 proliferation index [2,8].

In this specific case, TLE1 positivity and the absence of germline markers helped rule out recurrence of the previous seminoma. Furthermore, the intermediate proliferation rate (Ki-67 ~20%) aligns with the aggressive behavior observed on imaging and at surgery, consistent with reports from other series of this neoplasm.

Treatment justification

Optimal management of these neoplasms requires a multidisciplinary approach at sarcoma referral centers. Wide surgical resection with negative margins is the cornerstone of treatment and the most important prognostic factor for survival [12]. Surgical resection may involve multivisceral resection or resection with planned R1 margins when vascular or nerve structures are affected, provided that neoadjuvant or adjuvant therapy is administered [2,13]. In the series by Yang et al. [2], en bloc resections achieved prolonged local control without relapse, with follow-up ranging from 11 to 74 months.

Vertebral involvement remains a topic of discussion, as it includes tumors with *borderline resectability*. According to NCCN and ESMO guidelines, histological grade, histological subtype, tumor size, and involvement of vascular or nerve structures should be considered when deciding on neoadjuvant management [1,2,11,13]. In our case, the multidisciplinary team determined that the patient met the criteria for neoadjuvant chemotherapy with an MAI regimen and concurrent radiotherapy, aiming to improve the likelihood of complete resection and locoregional control.

Preoperative radiotherapy may be considered in retroperitoneal sarcomas (STRASS trial), although its benefit in specific SS is limited; however, it contributes to improved local control in high-grade tumors or in cases with vertebral involvement [3, 4,13].

In our case, the patient received neoadjuvant chemotherapy and radiotherapy, and a wide multivisceral local resection was performed, with planned positive margins (R1) on pathology, to preserve adjacent structures, improve function, and maintain the patient's quality of life (4,5). This approach was based on

international and national consensus guidelines, such as the INC's guidelines for managing retroperitoneal sarcomas in patients with tumors in borderline resectability scenarios [13].

In the adjuvant setting, it is recommended for patients with microscopically positive margins or high-grade subtypes, especially when neoadjuvant treatment was not administered, to help reduce the risk of recurrence [2, 3,4,13].

Local recurrence after resection of retroperitoneal SS can reach up to 80%, and distant involvement in 25%, with pulmonary involvement being the most common, which directly affects oncological outcomes [6,11].

Follow-up

Follow-up should be conducted at centers with extensive experience to evaluate early relapses and provide interdisciplinary patient management. Surveillance is essential for detecting recurrences that could potentially be curable. However, evidence on the most effective surveillance methods remains limited [12,14].

It is recommended to perform MRI or CT scans of the tumor bed every 6 months during the first 2 to 3 years, then annually [11,14]. For patients undergoing major vertebral reconstructions, follow-up CT scans should be performed every 6 months, along with regular neurological evaluations in collaboration with the Neurosurgery service [14].

CONCLUSIONS

Primary retroperitoneal SS is a rare condition. Its diagnosis and treatment present a significant challenge due to its location, aggressive behavior, and involvement of vascular and/or nerve structures. Imaging studies such as CT and MRI are essential for characterization and surgical planning.

A multidisciplinary approach at referral centers is vital for optimizing resection and oncological management, ensuring the treatment's oncological objectives are met. Retroperitoneal SS with vertebral involvement is a rare and highly complex condition. This case demonstrates that multidisciplinary management and careful surgical planning enable complete resection while preserving neurological function, even in complex anatomical scenarios. We present the first reported case in Colombia and Latin America of primary retroperitoneal SS with vertebral involvement, providing key clinical information for the approach and management of future cases.

COMPLIANCE WITH ETHICAL STANDARDS

Informed consent: The study adhered to current international bioethical standards, such as the Nuremberg Code, the Declaration of Helsinki, and the Belmont Report. It also complied with the regulations set forth in Article 1502 of the Colombian Civil Code, Law 23 of 1981, Decree 3380 of 1981, and Resolution 008430 of 1993 of the Ministry of Health, which establish the scientific, technical, and administrative standards for health research. Confidentiality was maintained in accordance with Article 15 of the 1991 Political Constitution of Colombia. Furthermore, written informed consent was obtained from the patient for the publication of the case and its corresponding images.

Conflicts of interest: The authors reported no conflicts of interest.

Use of Artificial Intelligence: The authors stated that they used artificial intelligence-assisted tools to optimize writing style and clarity.

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Authors' contributions

- Study design and conception: Natalia Lasprilla, Erika Benito, Laura Arce Polanía, Emma Gómez, Mauricio García.
- Data analysis and interpretation: Natalia Lasprilla, Erika Benito, Mauricio García, Sandra Díaz
- Manuscript writing: Natalia Lasprilla, Erika Benito, Laura Arce Polania
- Critical review: Natalia Lasprilla, Mauricio García, Sandra Díaz.

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