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COMPLEX RESECTION AND CAVAL RECONSTRUCTION IN PRIMARY INFERIOR VENA CAVA LEIOMYOSARCOMA

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Abstract:: This article describes a primary leiomyosarcoma of the inferior vena cava in the context of retroperitoneal sarcomas, emphasizing its epidemiological, diagnostic, and therapeutic aspects. A case report is presented of a patient with an invasive retroperitoneal mass, highlighting the diagnostic investigation, surgical planning, and the need for oncological resection associated with vascular reconstruction. The discussion addresses the rarity of the entity, the technical challenges of the surgical approach, prognostic factors, and the limited role of adjuvant therapies, in light of the current literature. It is concluded that surgical treatment with free margins remains the main determinant of survival, and multidisciplinary management in specialized centers is essential for optimizing oncological outcomes.

Introduction

Retroperitoneal sarcomas are rare malignant mesenchymal neoplasms characterized by slow and insidious growth in the retroperitoneal space, which often results in late diagnosis and complex therapeutic management (1). Among the histological subtypes, liposarcoma and leiomyosarcoma stand out as the most prevalent forms, together accounting for the majority of cases described in the literature (1,4). In this context, primary sarcomas of the inferior vena cava, predominantly leiomyosarcomas originating from the smooth muscle of the venous wall, represent an exceptional entity, but of high clinical and surgical relevance, due to the diagnostic complexity and frequent need for extensive vascular resections with reconstruction (2,5). From an epidemiological point of view, they mainly affect middle-aged adults, predominantly females, and their etiopathogenesis is still unclear, with hypotheses involving hormonal factors and local changes in the vascular wall (3,6). Given its rarity, potentially aggressive behavior, and therapeutic challenges, case reports remain essential to expand knowledge about the clinical presentation, management, and outcomes of these tumors (7).

Case Report

The case in question involves a 67-year-old white female patient with no previous history of cancer and a surgical history limited to tubal ligation and cesarean section. Her clinical comorbidities included systemic arterial hypertension, hypothyroidism, and anxiety disorder, with no other relevant conditions.

The diagnostic investigation began after the onset of diffuse abdominal pain, initially mild, progressive, and exacerbated by eating, beginning in December 2023, culminating in an episode of acute exacerbation in early 2024, which prompted a visit to the emergency room. At that time, abdominal computed tomography showed a large retroperitoneal mass with irregular contours, measuring 4.8 × 5.3 cm, closely related to and inseparable from the inferior vena cava. Given the radiological findings, histopathological clarification was performed by radiointerventional-guided biopsy, which revealed spindle cell neoplasia, with immunohistochemistry positive for smooth muscle actin (SMA) and desmin, in addition to a high proliferative index (Ki-67 of 50%).

The patient was then referred to a tertiary oncological surgery service, where complementary tomographic examinations, aimed at surgical planning and restaging,

demonstrated tumor progression, reaching dimensions of 7.4 × 9.9 × 4.9 cm in May 2025, associated with signs of invasion of the inferior vena cava, left gonadal vein, and left renal vein. After discussion in a multidisciplinary meeting, a curative surgical approach was chosen, in conjunction with the oncological and vascular surgery teams, aiming at complete resection of the lesion.

For total excision of the tumor, a median laparotomy associated with a right transverse incision was necessary, allowing wide exposure of the entire inferior vena cava. It was decided to section the affected venous segment with vascular reconstruction using a Dacron prosthesis and reimplantation of the left renal vein. The postoperative course was satisfactory, with maintenance of full anticoagulation, allowing hospital discharge without significant complications. Histopathological examination of the surgical specimen confirmed the diagnosis of leiomyosarcoma, with neoplasm-free surgical margins and microscopic involvement of 1 of the 2 lymph nodes evaluated, configuring pathological staging pT1pN1, according to the 8th edition of the AJCC, in addition to positive immunohistochemistry for AML and desmin, with a Ki-67 index of 30%.

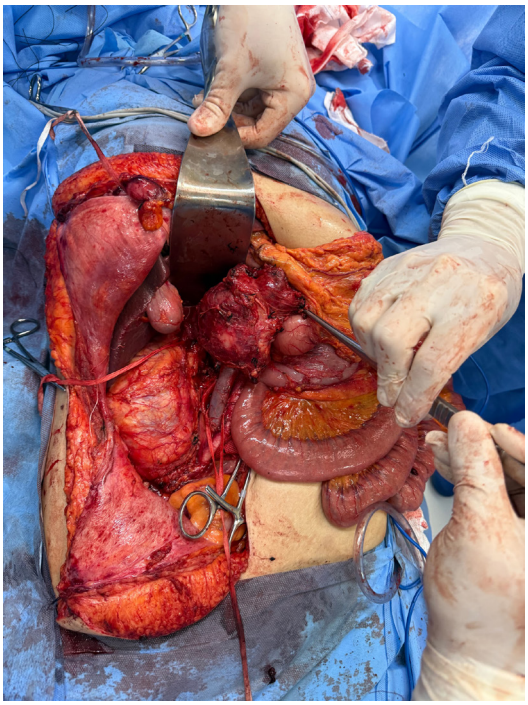

Fig. 1: Exposure of the surgical field with central visualization of the tumor prior to complete resection.

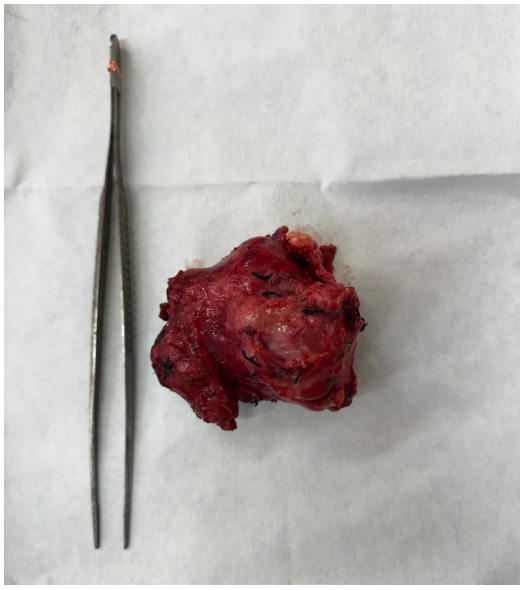

Fig. 2: Anterior view of the tumor after resection;

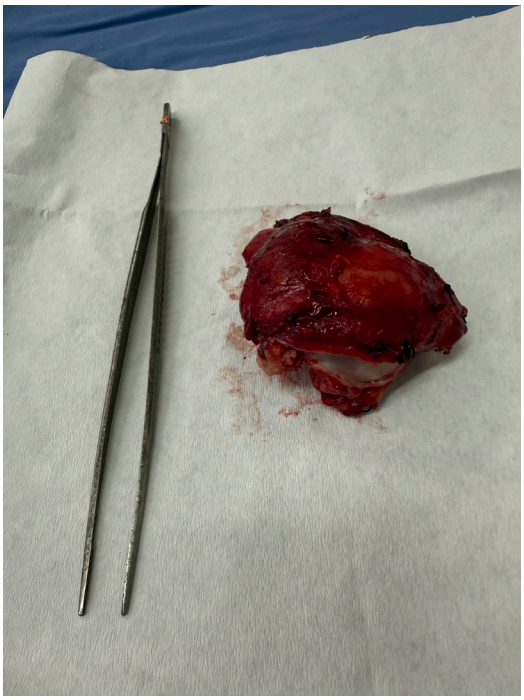

Fig. 3: Inferior tumor view after resection;

Discussion

Epidemiologically, retroperitoneal sarcomas account for approximately 10–20% of all soft tissue sarcomas and about 0.1–0.2% of solid cancers in the adult population, with an estimated annual incidence of around 0.3–0.4 cases per 100,000 inhabitants (1). Among the histological subtypes, liposarcoma is the most frequent, followed by leiomyosarcoma and, to a lesser extent, undifferentiated pleomorphic sarcoma. These neoplasms are characterized by slow and silent growth in the deep retroperitoneal y compartment, which often results in late diagnosis and presentation with large tumors. Evidence from large institutional series shows that tumor size at diagnosis and locoregional extension of the disease are directly associated with local recurrence rates and overall survival, reinforcing the prognostic impact of late presentation (4,7).

Within this spectrum, primary sarcomas of the inferior vena cava, predominantly leiomyosarcomas originating from the smooth muscle layer of the venous wall, are an exceptional entity and represent the most common primary malignant tumor of the central venous system (2). The clinical relevance of these tumors stems not only from their rarity but also from their close relationship with critical vascular and visceral structures, which often necessitates en bloc resections associated with complex vascular procedures, as in the case reported. Studies show that the location of the tumor along the different segments of the inferior vena cava directly influences the clinical presentation, surgical planning, and oncological outcomes, since the involvement of adjacent structures varies according to the affected segment (5,8).

It is predominantly observed in middle-aged adults, with a higher incidence in females, describing a female:male ratio ranging from 3:1 to 9:1, with most cases occurring between the fifth and sixth decades of life (3,6). Despite multiple proposed etiopathogenic hypotheses, including possible hormonal influences, local changes in the vascular wall, and previous thrombotic events, the etiology remains largely unknown, and most cases occur sporadically, without clearly established risk factors (6).

With regard to therapeutic management, complete surgical resection with free margins (R0) remains the cornerstone of oncological treatment for primary vena cava sarcomas and is the main independent prognostic factor associated with overall survival and locoregional control of the disease (2,3). Achieving negative margins often requires

extensive vascular resections, with ligation or reconstruction of the vena cava using synthetic prostheses or autologous grafts, as well as selective venous reimplantation, particularly of the renal veins. Data from the literature indicate that caval reconstruction is associated with better preservation of venous drainage and a reduction in late complications, especially in patients with a prolonged life expectancy, provided that it is performed in specialized tertiary centers (5,8). The role of radiotherapy, whether in a neoadjuvant or adjuvant context, remains controversial and may be considered on an individual basis, especially in high-grade, large tumors or in the presence of narrow surgical margins. Similarly, systemic chemotherapy has limited benefit, with modest responses to anthracycline-based regimens, and is generally reserved for unresectable, recurrent, or metastatic disease. Retrospective studies and case series suggest that these modalities should be used judiciously, within a carefully selected multimodal strategy(1,3,7).

Conclusion

Although rare, primary vena cava sarcomas should be considered in the differential diagnosis of retroperitoneal masses closely related to large vessels, especially when identified incidentally or in the context of nonspecific symptoms. Therapeutic management should be individualized, with careful surgical planning and detailed assessment of the possibility of complete oncological resection, often associated with the need for vascular reconstruction and lymph node analysis, in order to optimize oncological outcomes.

In addition, periodic clinical and radiological follow-up is essential, given the aggressive biological behavior of these tumors and the risk of late recurrence, even after resection with clear margins. This report reinforces the importance of adequate preoperative staging, a multidisciplinary approach, and management in centers specializing in the treatment of unusual and complex presentations, such as vena cava sarcomas.

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