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Management of a Rare Giant Thoracic Leiomyosarcoma: Case Report and Review of Literature

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Abstract **Case Report**

Thoracic wall sarcomas account for less than 20% of all soft tissue sarcomas, with leiomyosarcomas comprising only 1% to 4%. We report a case of a giant leiomyosarcoma of the thoracic wall in a 67-year-old patient presenting with a thoracic wall mass. Imaging revealed a large tumor involving the thoracic wall, and histopathological analysis confirmed leiomyosarcoma. Staging examinations excluded distant metastases. The patient underwent surgical resection followed by adjuvant therapy with close follow-up. Early diagnosis and multimodal management significantly improve the prognosis of thoracic leiomyosarcomas.

Keywords: Thoracic Wall, Pleomorphic Leiomyosarcoma, Surgical Resection, Adjuvant Therapy.

1. INTRODUCTION

The thoracic wall is a complex anatomical structure. Tumors of the thoracic wall are rare, constituting approximately 5% of all malignant thoracic tumors and only 1% to 2% of all tumors [1, 2]. These tumors can originate from any tissue in the thoracic wall [1-3], including bone, cartilage, muscle, adipose tissue, fibrous connective tissue, nerves, and blood vessels (arterial or lymphatic). Prognosis largely depends on the disease stage, with early detection being a critical prognostic factor influencing therapeutic management.

2. CASE REPORT

A 67-year-old patient with no significant medical history presented with a left thoracic wall mass near the 7th rib, progressively increasing in size but without systemic symptoms **(Figure 1)**. The mass measured 106 × 60 × 70 mm.

Thoracic CT revealed a large soft tissue mass involving the thoracic wall, measuring 96 × 40 × 60 mm, with no evidence of associated bone lysis. Two mediastinal lymph nodes were identified, measuring 7.4 and 8.5 mm in their longest axes **(Figure 2)**. Further abdominal-pelvic and cerebral CT scans showed no secondary lesions.

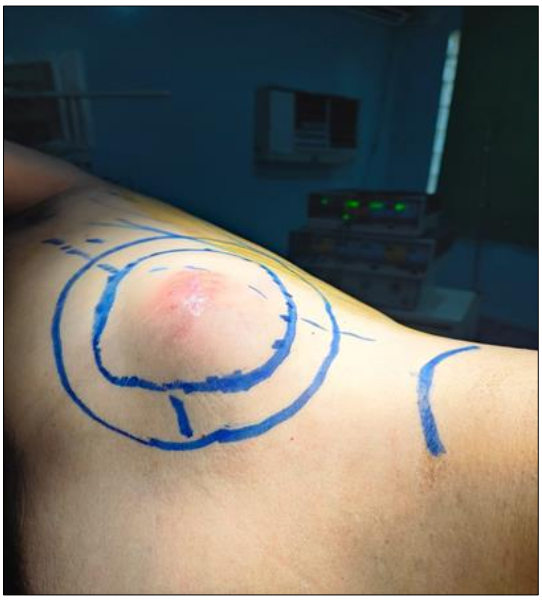

Figure 1: a left thoracic wall mass near the 7th rib

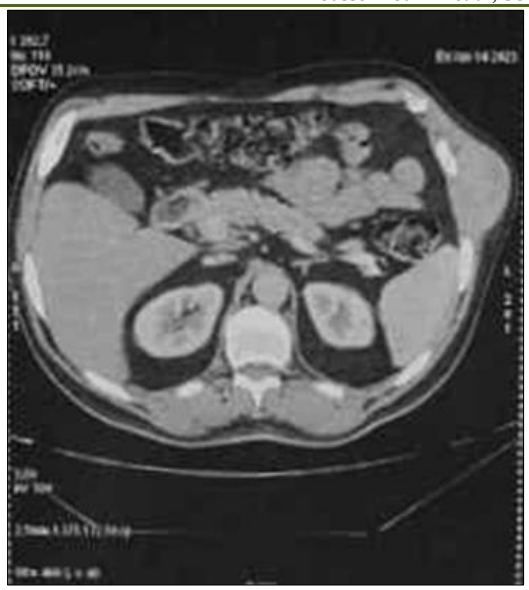

Figure 2: a large soft tissue mass involving the thoracic wall

A biopsy revealed a malignant sarcomatous tumor characterized by pleomorphic cells, marked cytonuclear atypia, numerous atypical mitoses, and minimal intratumoral necrosis. Immunohistochemistry was positive for Desmin, AML, and showed focal positivity for H-caldesmon, PS100, and cytokeratin AE1/AE3. Myogenin was negative, consistent with pleomorphic leiomyosarcoma.

The patient underwent surgical resection, which involved en bloc removal of three intercostal spaces (6th, 7th, and 8th ribs) **(Figure 3: a, b, c)**. The diaphragm was dissected on its abdominal surface and reattached without peritoneal breach **(Figure 3d)**, while thoracic wall stability was achieved using a prolene plate reinforced with a latissimus dorsi muscle flap **(Figure 3: e,f)**. Postoperative recovery was uneventful, with drain removal on postoperative day 5.

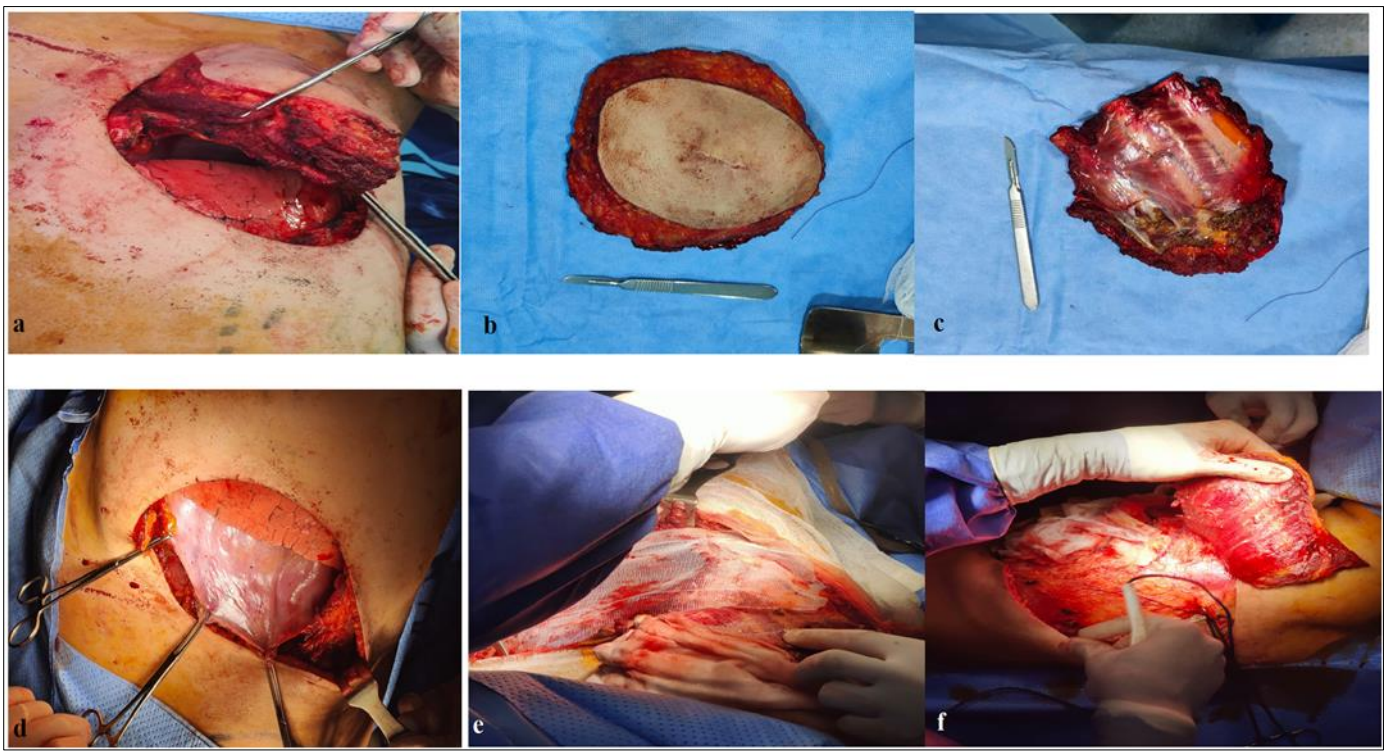

Figure 3: a, b, c: En bloc removal of the 6th, 7th, and 8th ribs
d: The dissection of the diaphragm without peritoneal breach
e, f : The implementation of a prolene plate, reinforced with a latissimus dorsi muscle flap

Histopathological analysis confirmed an R0 resection with clear margins exceeding 3 cm. Multidisciplinary evaluation recommended adjuvant radiotherapy followed by chemotherapy due to the tumor's size and aggressiveness.

3. DISCUSSION

Leiomyosarcoma is a malignant tumor arising from mesenchymal cells with smooth muscle differentiation [4], representing 5 to 10% of all soft tissue sarcomas. While it can occur in any anatomical location, leiomyosarcoma of the thoracic wall remains uncommon [5, 6].

Thoracic leiomyosarcomas may manifest with nonspecific symptoms such as cough, chest pain, and dyspnea or can be asymptomatic as observed in our patient [7]. CT scans typically characterize thoracic leiomyosarcomas as large, soft tissue masses with heterogeneous enhancement due to hemorrhage, necrosis, and other heterogeneous structures. Larger tumors may displace surrounding organs and occasionally exhibit invasiveness [8]. MRI, superior for soft tissue sarcoma assessment, typically reveals the tumor with iso- to hypo-intensity, enhancing on T1W1, and presenting iso- to hyper-intensity on T2WI [9]. Leiomyosarcoma demonstrates high FDG uptake on PET-CT [10].

Histologically, leiomyosarcoma appears as a fascicular spindle cell neoplasm. Tumor cells display abundant eosinophilic cytoplasm and elongated cigar-shaped nuclei, mimicking normal smooth muscle structure [11]. Poorly differentiated tumors resemble to the normal smooth muscle structure. The differential diagnosis for leiomyosarcoma traditionally includes other fascicular spindle cell neoplasms such as neurofibrosarcoma, synovial sarcoma, spindle cell rhabdomyosarcoma, and inflammatory myofibroblastic tumors.

Immunohistochemistry is essential for confirming smooth muscle differentiation, with markers such as Desmin, α-SMA, muscle actin (HHF-35), and H-caldesmon being particularly useful [11]. Actins and h-caldesmon may be more sensitive than desmin in detecting myogenic differentiation in smooth muscle neoplasms, with desmin expression occurring in 70% to 80% of cases [12]. Some leiomyosarcomas, especially those in the retroperitoneum, may exhibit significant pleomorphism. In such cases, tumor recognition as leiomyosarcoma may only occur after identifying focal areas with identifiable smooth muscle differentiation. Although immunoreactivity to desmin and/or α-SMA may be present in pleomorphic tumor cells, desmin staining is often less diffuse than in areas with morphologically recognizable smooth muscle differentiation [13].

The optimal treatment for thoracic leiomyosarcoma is not clearly defined. The standard for localized disease treatment is surgical resection, with or without neoadjuvant or adjuvant radiotherapy and chemotherapy [14]. The recommended excision margin size is similar to other sarcomas. Kawaguchi *et al*., proposed a 2 cm margin for sarcomas involving soft tissues and bone [15], while King *et al*., recommended a 4 cm margin to improve 5-year survival rates in thoracic sarcoma patients [16]. Mesko *et al*., recently suggested a 2 cm margin for low-grade tumors and a 4 cm R0 margin for high-grade tumors [17].

Neoadjuvant radiotherapy is frequently used in LMS treatment. Studies have shown that 90% of patients receiving neoadjuvant radiotherapy for limb-localized LMS achieved margins with no residual disease (R0), compared to reduced percentages in groups receiving adjuvant radiotherapy or no radiotherapy [18].

Adjuvant doxorubicin-based chemotherapy reduced overall recurrence with an odds ratio of 0.69, while the combination of doxorubicin and ifosfamide reduced overall recurrence with an odds ratio of 0.61. However, this advantage was not observed in a pooled analysis of two European Organisation for Research and Treatment of Cancer (EORTC) trials. Adjuvant chemotherapy in 819 patients with soft tissue sarcoma (STS) (including LMS) randomized into adjuvant or non-adjuvant therapeutic arms did not show improved survival, regardless of sarcoma subtype [19].

Nevertheless, contradictory results from multiple clinical trials evaluating the benefit of adjuvant chemotherapy (with or without radiotherapy) after surgical excision to prevent recurrences exist. A meta-analysis of 18 trials involving 1953 patients with soft tissue sarcomas, including leiomyosarcoma, with resectable localized tumors, showed that adjuvant chemotherapy led to a statistically significant reduction in both local and distant recurrence, with odds ratios of 0.73 and 0.69, respectively [20]. A subsequent EORTC study randomized 351 patients with intermediate to high-grade STS (including LMS) without metastases to receive adjuvant chemotherapy after excision, but no survival benefit was observed compared to the control group in terms of overall survival (OS) or relapse-free survival (RFS) [21]. These trials are limited by the inclusion of heterogeneous patient populations, encompassing different tumor subtypes, sizes, grades, and anatomical locations, as well as small patient numbers and suboptimal chemotherapy regimens.

The potential benefit of neoadjuvant chemotherapy is still under evaluation, with current studies also providing conflicting results [22, 23]. The findings from a phase 3 randomized trial demonstrated that neoadjuvant chemotherapy led to no complete response and improved overall survival (OS) or disease-free survival (DFS) only in patients treated with standard

chemotherapy such as anthracycline and ifosfamide, not histology-directed chemotherapy, with a DFS odds ratio of 0.47 [24]. Due to these conflicting reports, adjuvant chemotherapy is not systematically recommended.

4. CONCLUSION

The reported case underscores the rarity of leiomyosarcoma in the thoracic wall and highlights the complexity of its management. Surgical resection remains the cornerstone for localized forms, with considerations on excision margins. Adjuvant approaches such as radiotherapy and chemotherapy are gaining importance, emphasizing the value of collective decisions made during multidisciplinary team meetings for an increasingly individualized therapeutic approach.

Conflicts of Interest:

The authors of this article declare that they have no conflicts of interest. The authors are solely responsible for the information and writing of this article.

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