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Case Report

A chest wall sarcoma, a recurrence 8 years after index excision: a case report

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ABSTRACT

Soft tissue sarcomas (STS) occur in 2-5 patients per 100,000 people, as a rare heterogeneous group of tumors arising in connective tissues, embryologically derived from mesenchyme. These tumors comprise <1% of all neoplasms with a 4:1 male preponderance. Prognosis depends on size, grade, depth and metastasis. Even in STS classified as low-grade, the risk of local recurrence requiring further intervention remains high, therefore emphasizing the importance of long-term follow-up. While nodal metastasis is rare, the tumor immediately becomes stage III when this occurs, regardless of size or depth. More than 50 subtypes exist and originate from cartilage, muscle, blood vessels, nerves, fat, tendons and deep skin tissues. Low-grade fibromyxoid sarcoma (LGFMS), one rare subtype with a relatively benign histologic appearance, has a high recurrence rate, late metastatic spread, and chemotherapy and radiotherapy insensitivity. We present the case of a 57-year-old male with a right chest wall mass diagnosed as a recurrent low-grade fibromyxoid sarcoma (LGFMS) 8 years after index excision. We emphasize the importance of early detection and management given the value of early surgical intervention due to the limited response of adjuvant therapy in this low-grade yet aggressive tumor.

Keywords: Sarcoma, Recurrent soft tissue sarcoma, Chest wall tumor, Fibromyxoid sarcoma

INTRODUCTION

Soft tissue sarcomas (STS) are a rare and diverse group of malignancies that develop in connective tissues of mesenchymal origin, with an incidence ranging from 2 to 5 cases per 100,000 individuals annually despite their rarity in accounting for less than 1% of all neoplasms, STS demonstrate a marked male predominance, with a reported ratio of 4:1.[1] Over 50 histological subtypes have been identified, arising from structures such as cartilage, muscle, vasculature, nerves, adipose tissue, tendons, and dermis. Among these, undifferentiated pleomorphic sarcoma is the most frequently diagnosed subtype, as confirmed in a two-decade epidemiological review conducted from 1984-2004.[2-4] In contrast, low-grade fibromyxoid sarcoma (LGFMS), first characterized in 1987, is exceedingly uncommon, compromising approximately 0.6% of all STS cases, incidence of 0.18 per million" to "incidence of 0.18 cases per 1 million people.[5] Although histologically indolent, LGFMS poses significant clinical challenges due to its potential for delayed metastasis, local recurrence, and resistance to conventional chemotherapy and radiotherapy. While the extremities and trunk are the most typical sites of origin, rare cases involving the chest wall have been reported.[2,6,7] The median age at diagnosis is 59 years.[2] We present the case of a 57-year-old male with a recurrent right chest wall mass diagnosed as a recurrent low-grade fibromyxoid

sarcoma with positive margins on re-excision 8 years after index operation.

CASE REPORT

A 57-year-old male with a past medical history of hypertension, hyperlipidemia, and diabetes mellitus presented in 2025 with a nontender, right upper chest wall mass. His past surgical history was significant for excision of a 13 cm right chest wall sarcoma at the same location in 2017, identified at the time as a low-grade fibromyxoid sarcoma. The patient was then lost to follow-up. His chest computed tomography (CT) showed a 16×9.7×11 cm predominantly cystic mass with mildly enhancing septations and invasion into the pectoralis muscles, which appeared to be in close proximity to the subclavian vein, although no invasion on imaging could be identified (Figure 1). Evaluation for metastatic disease with CT abdominal/pelvis was negative. Preoperative labs (CBC, BMP) were unremarkable. Due to the patient's history of a prior chest wall sarcoma and CT findings indicating a likely recurrent mass, no biopsy was done preoperatively. The patient was scheduled for wide resection of this recurrent chest wall mass.

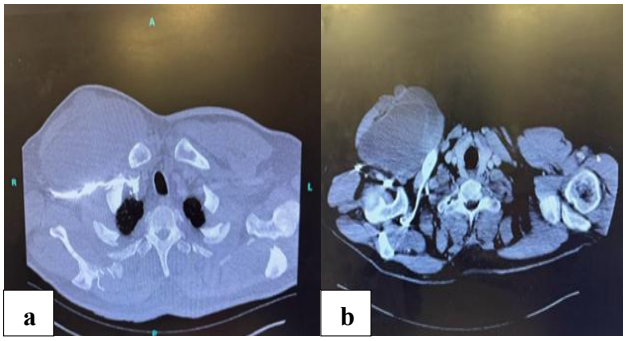

Figure 1: (a) Contrast-enhanced computed tomography reveals right chest wall mass invading the pectoralis muscle and in close proximity to, but not invading, the subclavian vein, and (b) contrast CT revealing large right chest wall sarcoma.

Intraoperative findings confirmed the right chest wall mass in the infraclavicular region to be in close proximity to the subclavian vessels, with invasion of the pectoral muscles. The procedure included the mass excision and resection of 90% of the pectoralis major muscle, with surgical clips placement (Figure 2). Histopathological findings showed a neoplasm of low cellularity with predominantly myxoid and locally fibrotic components of monomorphic spindle cells arranged in patternless or short fascicles. Results also described the mass as having highly infiltrative growth into the skeletal muscle, FUS (16p11.2) gene rearrangements and MUC4 immunohisto positivity (consistent with 2017 findings), with positive inferior, lateral, and medial margins. These results all confirmed a diagnosis of a low-grade fibromyxoid sarcoma. The patient's postoperative course was uncomplicated, and he was scheduled to receive adjuvant radiation under the guidance of surgical clips placed intraoperatively.

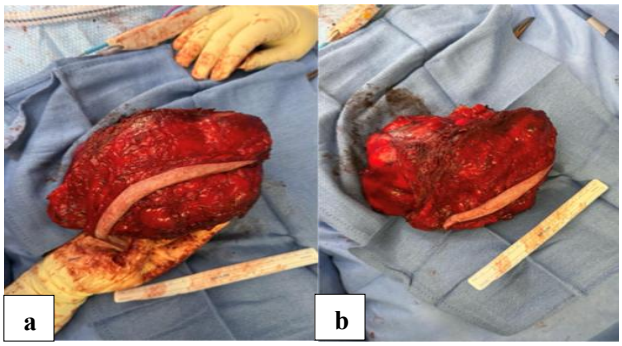

Figure 2 (a and b): Intraoperative chest wall mass measuring 16x11cm. Mass was excised alongside the invaded pectoralis muscle and was closely excised from the subclavian vein. Clips were placed in the area to facilitate targets for postoperative radiation.

DISCUSSION

STS are a rare, heterogenous group of tumors arising in connective tissues embryologically derived from mesenchyme.[1] They comprise <1% of all adult neoplasms. While most cases of STS occur sporadically, germline mutations found in patients with neurofibromatosis type I, Li-Fraumeni syndrome and familial adenomatous polyposis have been reported" to "with neurofibromatosis type I (NF- 1), Li-Fraumeni syndrome and familial adenomatous polyposis (FAP) have been reported". Radiation and carcinogen exposure, and conditions such as chronic lymphedema, may also predispose patients to the development of these neoplasms.[7,10] LGFMS are estimated to represent fewer than 5% of soft-tissue sarcomas, with an incidence of 0.18 per million.[5,8] LGFMS are distinctly described as tumors with alternating fibrous and myxoid areas and having a bland histology featuring with alternating fibrous and myxoid areas, having a bland histology, featuring fibroblastic spindle cells with scant cytoplasm arranged in a short fascicular or whorled pattern. Despite its seemingly benign and indolent pattern of growth, they frequently metastasize late to the lungs, prostate or liver.[8,9]

Initially, LGFMS typically present as slow growing, painless masses, most commonly on the trunk or extremities, but can also evolve for decades in a patient with metastatic disease. Occasionally, they may also be found in the groin/perineum[11] and in the head and neck (such as scalp, face, nasal cavity).[12] They are often located deep to the fascia in adults, while superficial masses are seen in pediatric patients.[8,9,11] These tumors can range in size from 1 cm to in excess of 20 cm and are characterized by a white appearing, fibrous cut surface on macroscopic examination intraoperatively.[5]

Because of its histology and ambiguous presentation, LGMFS can be mistaken for other benign and malignant

conditions. LGMFS as an isolated chest wall mass should be differentiated from desmoid fibromatosis, neurofibromas, undifferentiated pleomorphic sarcoma (malignant fibrous histiocytoma), myxoid and spindle cell liposarcomas, lipomas, low-grade myxofibrosarcoma, nodular fasciitis, metastatic hepatocellular, squamous cell carcinoma, and intramuscular hemangioma.[5,13-17]

Ultrasound has been used to assess depth of invasion and features of suspicious soft tissue masses, but magnetic resonance imaging (MRI) is preferred for local assessment of tumors, while CT is used for staging.[3,6] To support imaging findings, a core needle biopsy, with or without CT or ultrasound guidance, is required to obtain an adequate tumor sample for diagnosis.[3,18] When performing this procedure, it is essential to keep the biopsy needle within the intended area of excision to avoid seeding along the biopsy tract to minimize contamination of surrounding structures while ensuring the possibility of achieving negative surgical margins.[3,18,19]

Histologic features of scant cellularity with bland morphology along with ample collagen can be found in both benign and malignant tumors. For this reason, misdiagnosis of fibromatosis, malignant fibrous histiocytoma, and neurofibroma has occurred.[5] Immunohistochemistry, cytogenetic and molecular genetic testing on tissue samples will help accurately identify sarcomas and guide treatment.[20] Mucin-4 (MUC4) has emerged as a highly sensitive and specific immunohistochemical marker for LGFMS. Hassan et al have found MUC4 expression in 100% of LGFMS cases, whereas most other soft-tissue tumors do not include this expression, reinforcing the diagnostic utility of MUC4 in challenging soft-tissue sarcoma.[21] More than 90% of LGFMS also exhibit a characteristic translocation at chromosome 16p11, forming a fusion gene of FUS RNA binding protein (FUS)–cAMP responsive element binding protein 3-like 2 (CREB3L2), named FUS-CREB3L2. The FUS-CREB3L1 fusion gene can also be seen in approximately 5% of LGFMS cases. These translocations are detected as gene rearrangements at chromosome 16p11 via fluorescence in situ hybridization (FISH).[5,8,9] This patient's tumor was positive for both MUC4 protein and FUS rearrangements at chromosome 16p11, both specific findings strongly suggesting a fibromyxoid sarcoma diagnosis.[9,22]

LFGMS tend to be chemotherapy and radiotherapy insensitive due to their low mitotic activity, and are therefore largely dependent on aggressive surgical resection for treatment.[5] Surgery is considered the mainstay treatment for STS, with the goal of wide excision with negative margins, as it is the most important factor in preventing recurrence and improving survival in both primary and recurrent STS.[1, 23] An example is shown in a previous case of rare LGFMS where due to the tumor's large size, midline location, and infiltrative nature of the sternum and adjacent costal cartilages, an en bloc surgical resection, with a joint thoracic and plastic surgery approach, included complete removal and reconstruction of the sternum in order to achieve clear margins. An en bloc resection, a technique where a tumor and surrounding margin of healthy tissue are removed as a single intact piece, may be needed in order to achieve complete resection of the mass and obtain clear margins.[24] Our patient underwent radical resection of the recurrent tumor, with excision of approximately 90% of the pectoralis major muscle considering the tumor's invasion within the muscle compartment. Surgery is the only treatment that can result in disease-free periods. Staged surgical resection should be considered to optimize chances of clear margins while also optimizing patient recovery in cases of extensive resection due to locally advanced tumors.

Debulking operations (cytoreductive surgery) have a limited role and no survival advantage in STS.[25] However, it can be considered with palliative intent, as a precursor to other therapies, or as a method for reduction of tumor burden in metastatic disease.[26]

Neoadjuvant or adjuvant radiotherapy is usually considered in tumors with infiltrative growth to improve local control.[8] Neoadjuvant radiation is preferred for its benefits in tumor downstaging and early treatment of micrometastases, allowing lower total radiation doses.[27] Adjuvant radiation, however, still remains an effective strategy. In this patient, adjuvant radiation is indicated due to recurrent disease, tumor size >5 cm, and positive excision margins.[27]

Chemotherapy is typically reserved for high-risk STS, with anthracycline-based regimens in the neoadjuvant or adjuvant settings.[28] Currently available conventional systemic therapies have limited efficacy in LGFMS with no significant difference in median time from presentation to death compared to those treated without systemic treatment for recurrent or advanced disease.[8] The use of chemotherapy as stand-alone or adjuvant treatment for LGFMS remains unclear and may merely slow disease progression. Conventional chemotherapy has shown limited efficacy in the treatment of low-grade fibromyxoid sarcoma (LGFMS), with studies reporting no objective response rate and a median progression-free survival of less than two months.[8,29] While targeted drug therapy can be used in certain cases of STS, no guidelines have been set.[7] Preliminary data indicate a positive potential response rate to tyrosine kinase inhibitor (TKI) axitinib, and agents like trabectedin and pazopanib may offer prolonged disease stabilization in select cases, but efficacy in LGFMS remains unknown.[8]

Even in STS classified as low grade, the risk of local recurrence requiring further intervention remains high.[30] Despite its benign nature, LGFMS has a significant potential for recurrence and metastasis, often occurring years after initial treatment, most commonly to the lung. In approximately 6% of patients with LGFMS, the disease will recur, while metastasis will occur in 9% of patients within 2 years of primary resection. In contrast, long-term

recurrence rates as high as 64% and metastasis rates of 46% have been observed.[31] Generally, localized tumors (stage I and II) have better survival rates of 70-90% compared to metastatic disease (stage IV), which is associated with 10-20% survival.[32] These statistics demonstrate the importance of long-term surveillance in patients with STS, even when tumors are considered low risk. Current guidelines recommend imaging every 3–6 months for the first 2–3 years after surgery, then less frequently over time, to detect recurrence early.[7] Given the risk of late local recurrence and distant metastasis, long-term, preferably lifelong, surveillance is recommended for patients with LGFMS, with periodic MRI and chest CT imaging to monitor for recurrence and pulmonary spread.[31]

CONCLUSION

This case of a patient with a LGMS of the chest represents a rare location of an uncommon disease. The deceptively benign histology of this tumor is misleading and its insensitivity to chemotherapy and radiotherapy highlights the critical role of complete surgical resection, the need for multidisciplinary care, and the value of continuous follow-up. Despite low-grade status, LGFMS carry a high recurrence rate often requiring additional extensive surgical intervention with negative margins and adjuvant treatment in certain cases. Long-term follow up is crucial to early detection and management of recurrent disease.

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