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Classic Low-Grade Fibromyxoid Sarcoma: A Case Report and Literature Review

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Abstract

Background: The low-grade fibromyxoid sarcoma (LGFMS) is an exceptionally uncommon sarcoma that primarily manifests in the extremities or trunk of young adults, presenting as painless lesions. The histological features of this tumor are benign, but it exhibits an exceptionally high rate of late recurrence and a significant potential for metastasis. Imaging examinations serve as a crucial method for detecting LGFMS, while the definitive diagnosis relies on histopathological assessment. Currently, the primary treatment modality for this neoplasm is surgical resection. Early aggressive surgery with negative margins is a critical factor in mitigating the risk of tumor recurrence and metastasis. The present study presented a case of LGFMS located in the right thigh. The patient underwent a mass resection procedure following an MRI examination. During the telephone follow-up one year post-surgery, despite the absence of an imaging review, the surgical site demonstrated satisfactory recovery with no reported abnormal symptoms. **Case Presentation:** The patient, a 31-year-old male, presented to our hospital for evaluation of an asymptomatic mass in his right thigh that was incidentally discovered 13 years ago. The MRI showed a well-defined mass measuring 8.2 cm × 6.8 cm × 9.6 cm in the right thigh. The tumor signals exhibit a mixed pattern, characterized by predominantly isointense and hypointense signals on T1-weighted imaging (T1WI), a central area of hyperintensity on T2-weighted imaging (T2WI), and peripheral circular enhancement observed on contrast-enhanced scans. The patient underwent surgical resection. Microscopically, the mass was composed of intricately interwoven fibrous matrix and a distinct mucoid region. The tumor cells exhibited a distinctive arrangement in a swirling or wheel-like pattern, with minimal variation in their karyotypic characteristics. The immunohistochemical examination revealed diffuse and intense MUC4 positivity in the tumor cells.

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The diagnosis of LGFMS was confirmed by post-operative histopathological examination. **Conclusions:** The LGFMS is an exceptionally uncommon mesenchymal tumor renowned for its benign histological manifestations and malignant behavior. It is crucial to provide a comprehensive summary of the research findings and thoroughly review the existing literature pertaining to this rare disease.

Keywords

Low-Grade Fibromyxoid Sarcoma, Rare Sarcoma, Case Report

1. Introduction

Since its initial description by Dr. Harry Evans in 1987, low-grade fibromyxoid sarcoma (LGFMS) has been recognized for its benign histological characteristics coupled with malignant behavior [1]. The majority of reported cases occur in the limbs or trunk of young adults, with rare instances found in the head and neck region. LGFMS is classified as an extremely rare subtype of sarcomas, occurring at a frequency of less than 1 per million individuals [2] and representing less than 5% of all soft tissue sarcomas [3] [4]. This report presents a case of LGFMS located in the lower limb of a 31-year-old male patient and includes a review of the existing literature.

2. Case Presentation

A 32-year-old man discovered a painless mass in his right thigh during a routine examination following a car accident he experienced 13 years prior. The physician performed external fixation traction on the patient's right lower limb but did not address the mass in his thigh. Over the past 13 years, there was no significant growth of the tumor noted by the patient. Upon admission to our hospital, physical examination revealed a firm, mildly tender, and immobile mass measuring 15 cm × 15 cm that protruded from the inner aspect of the patient's right thigh. Hematological and biochemical parameters were within normal limits. The patient had no other detrimental habits apart from smoking.

MRI demonstrated a well-defined mass located in the right femoral intermediate muscle, measuring 8.2 cm × 6.8 cm × 9.6 cm (**Figure 1**). The T1-weighted image demonstrated predominantly hypointense signal within the lesion, with the local surface area exhibiting a signal intensity equivalent to or slightly higher than that of the adjacent muscle. STIR/T2-weighted fat-saturated images revealed irregular hyperintense areas along with a flocculent shadow that was isointense to muscle within the central region of the mass. The peripheral portions of the mass displayed low signal intensity on T2-weighted imaging and showed distinct ring-like enhancement following contrast material injection. No significant enhancement was observed in the tumor's center.

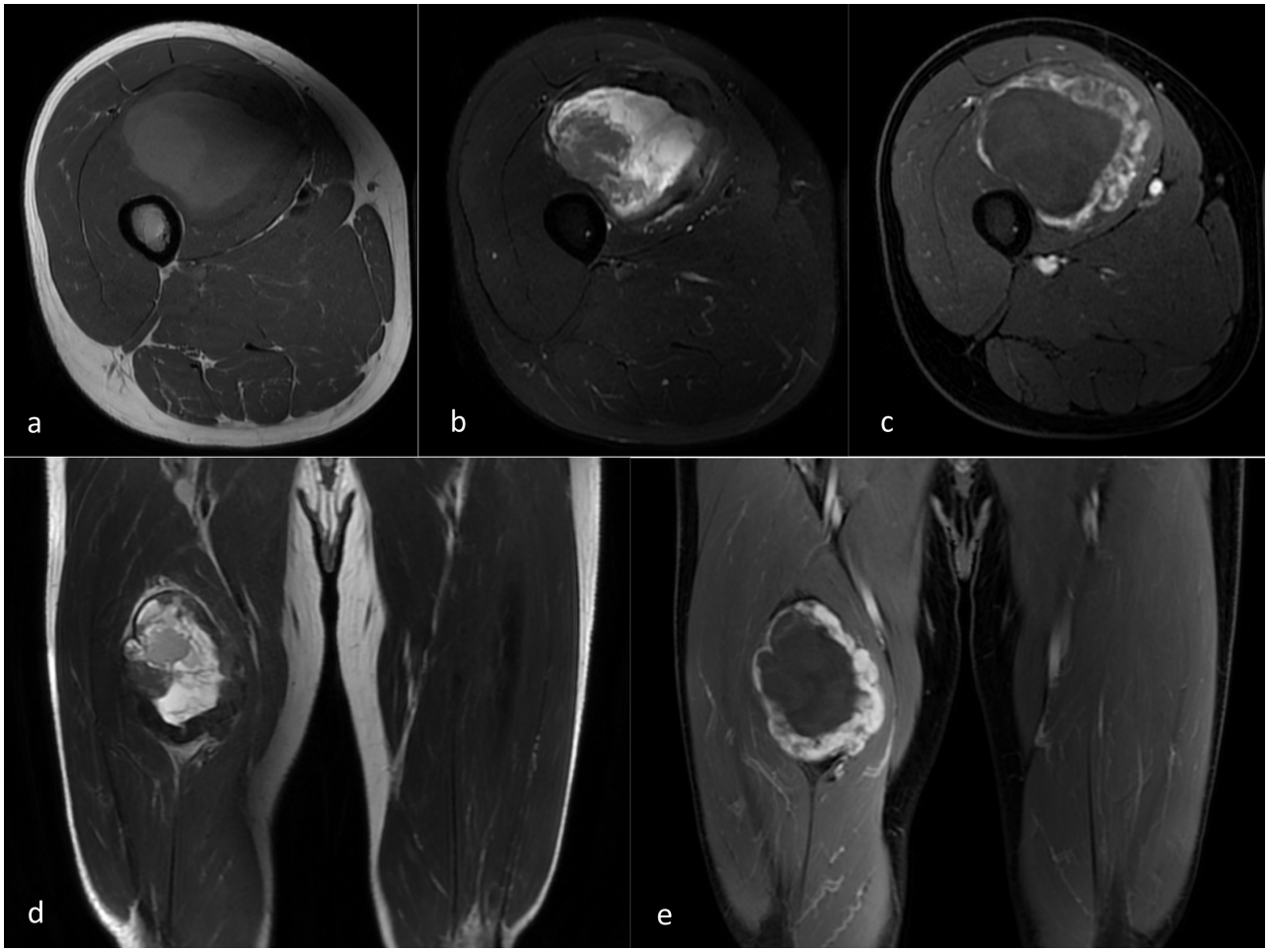

Figure 1. Right thigh MRI plain scan and enhanced scan. (a) On T1-weighted images, the center of the mass exhibited hypointense and isointense signals. The localized signals exhibit a slight elevation compared to the muscle signals. (b) The tumor exhibited heterogeneous signals on T2-FS, characterized by regions of hyperintensity. (c) After administration of the contrast agent, the tumor exhibits a ring enhancement. (d) The image depicts the mass on the coronal T2-weighted imaging. The mass is distinctly delineated from the surrounding muscle tissue. (e) The coronal T1WI enhanced imaging clearly demonstrated a well-defined circular enhancement of the mass.

The tumor was excised by the surgeon after a duration of 4 days. During the procedure, a mass measuring approximately 13 cm × 10 cm × 8 cm was noted, characterized by an irregular surface and firm texture, which facilitated its removal.

Macroscopically, the cut surfaces of the mass appeared grayish-white, with cystic areas containing grayish-yellow flocculent material visible within the mass (**Figure 2**). Microscopically, a distinct alternation between densely packed and sparsely distributed cell regions was observed, with relatively clear boundaries (**Figure 3**). Hemangiopericytoma-like structures were dispersed throughout. The tumor cells exhibited swirling patterns and were arranged in linear or chaotic distributions, displaying fusiform or short fusiform shapes without evident signs of nuclear division. The central region of the tumor demonstrated necrosis.

The solid mass exhibits a grayish white composition, with the presence of flocculent material within the sac.

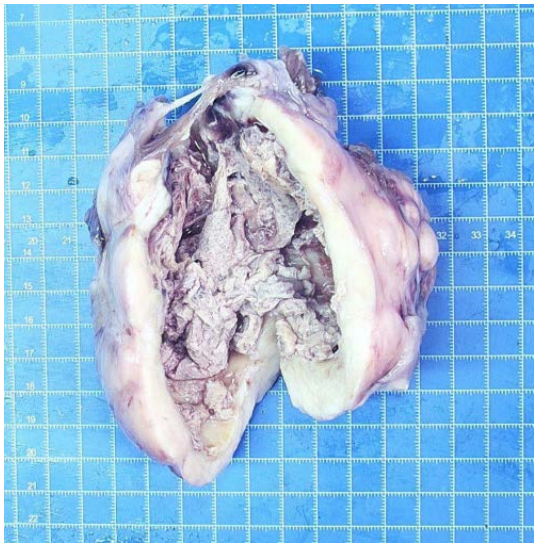

Figure 2. Gross view of resected tumor.

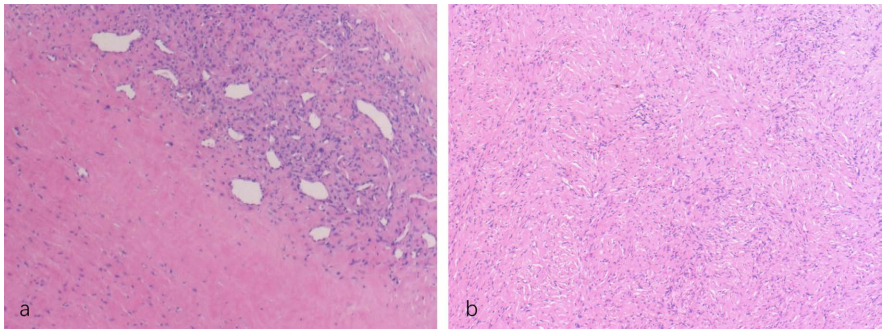

Figure 3. Pathological tissue section: hematoxylin and eosin staining. (a) Histological examination showed that the lesion was composed of fibrous matrix and mucoid region [hematoxylin and eosin staining (H&E); magnification: ×40]. (b) The bland tumor cells are distributed in swirling or wheel-like patterns [hematoxylin and eosin staining (H&E); magnification: ×40].

Immunohistochemically, tumor cells exhibited diffuse and strong cytoplasmic immunoreactivity to MUC4, while demonstrating very low proliferative activity as indicated by Ki-67 staining. The estimated proportion of Ki-67 positive cells within the tumor cell population is 2%. Negative results were obtained for EMA, SMA, Desmin, S-100, and β-Catenin (**Figure 4**). Furthermore, no immunoreactivity was observed for antibodies against STAT-6, effectively ruling out solitary fibrous tumors.

The diagnosis of low-grade fibromyxoid sarcoma (LGFMS) was established based on the pathological and immunohistochemical findings.

The patient underwent a follow-up imaging examination at another hospital for a month post-surgery, which revealed satisfactory recovery of the surgical site with no residual tumor signs detected. The patient did not receive any additional imaging studies during the following year. During the telephone follow-up, the patient reported no discomfort or presence of a mass on the right thigh.

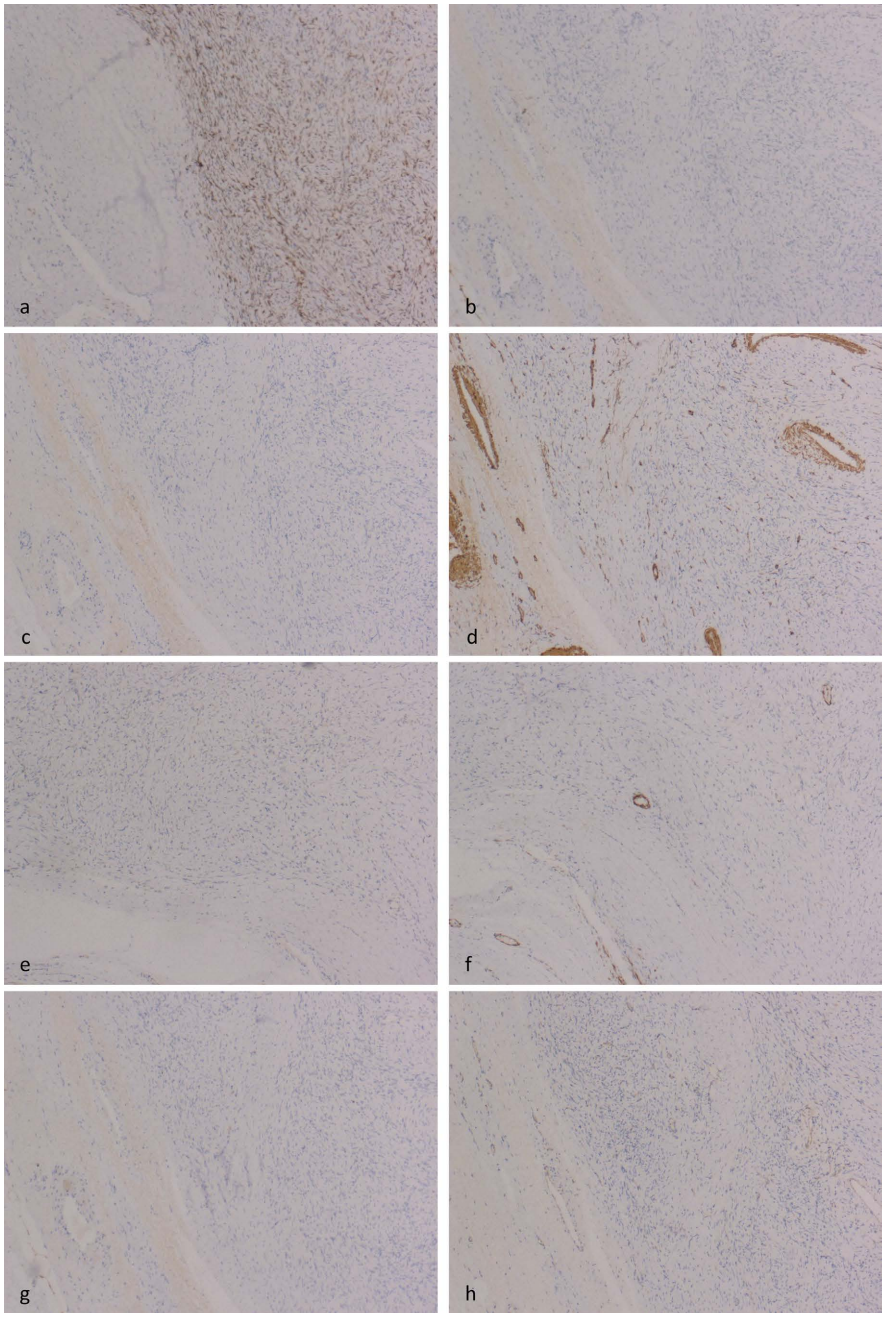

Figure 4. Pathological tissue section: immunohistochemistry. (a) The tumor cells exhibited a widespread and robust cytoplasmic immune reaction towards MUC4 [magnification: ×40]. (b) The tumor exhibited a low level of proliferative activity in Ki-67 staining [magnification: ×40]. (c) - (h) Tumor cells exhibited negative staining for EMA, SMA, STAT6, Desmin, S-100, and β-Catenin [magnification: ×40].

3. Discussion

The 2020 edition of the World Health Organization characterizes LGFMS as a "fibroblast/myofibroblastic tumor" [2], which typically manifests as a slow-growing, asymptomatic soft tissue mass. Research indicates that LGFMS predominantly affects younger adults, with a median age of approximately 34 years and no

significant differences in incidence between genders [5]. In recent years, numerous cases of LGFMS have been documented in elderly individuals and children [6]-[9]; notably, the youngest patient reported was merely 2 months old [9], suggesting that LGFMS can manifest across all age groups. Previous studies indicate that the lower extremities are the most frequently affected sites. In this instance, LGFMS presented in the thigh—The most common location for this tumor. Furthermore, deep soft tissues of the trunk also represent prevalent sites, followed by regions such as the groin, upper limbs, and buttocks; occurrences in areas like the chest [10] and head and neck are less frequent. Recently published reports have also identified cases arising in superficial locations such as lip mucosa and scrotum [11]-[13].

LGFMS is characterized by its slow growth; however, it is notorious for its high risk of recurrence and significant potential for metastasis. In cohort studies, Evans *et al.* observed that the recurrence and metastasis rates of LGFMS were approximately 10% and 5% within the first five years, while long-term local recurrence and eventual metastasis rates could escalate to 64% and 46%, respectively [14]. Folpe *et al.* proposed that early intervention may mitigate the incidence of both recurrence and metastasis [15]. The lung is identified as the most common site of metastasis for LGFMS, followed by the pleura [14], with vertebrae being the predominant sites for bone metastases. The patient in the cases we reported did not report any significant discomfort during a one-year telephone follow-up. Nevertheless, due to the elevated rate of recurrence associated with LGFMS, long-term follow-up and regular re-evaluation of patients are critically important.

The definitive diagnosis of LGFMS primarily relies on histopathological examination. The lesion typically presents as a well-defined mass, with infrequent occurrences of bleeding and necrosis. Microscopic analysis of this case revealed characteristic histopathological features of LGFMS, including alternating myxoid and fibrous regions, prominent arched vasculature, and bland tumor cells arranged in swirling or wheel-like patterns. Tumor cells in LGFMS exhibit only mild nuclear pleomorphism accompanied by minimal mitotic activity [13]. Other histologic variations may include large rosettes [13] [15] [16], epithelioid areas, or focal calcifications [13]. Zhang *et al.* suggested that focal dense cellularity with reduced mass and increased atypia might correlate with the rapid proliferation and invasive behavior associated with LGFMS [14].

LGFMS must be morphologically distinguished from other benign tumors that exhibit mucoid morphology or low-grade spindle-cell hyperplasia, including low-grade myxofibrosarcoma, malignant peripheral nerve sheath tumor, liposarcoma, neurofibroma, solitary fibrous tumor (SFT), and protuberant cutaneous fibrosarcoma, etc. [17] [18]. Immunohistochemistry facilitates the rapid and accurate differentiation of diagnoses. Previous studies involving large cohorts have identified MUC4 as a highly sensitive marker for diagnosing LGFMS [19]. Usman Hassan *et al.* utilized mouse anti-MUC4 monoclonal antibodies to investigate a range of tissue samples and discovered that malignant peripheral nerve sheath tumors,

fibrosarcoma, leiomyosarcoma, liposarcoma, and myxofibrosarcoma exhibited no MUC4 expression [20]. It is crucial to acknowledge that the overexpression of MUC4 is not a specific marker for LGFMS. MUC4 may also exhibit positive expression in sclerosing epithelioid fibrosarcoma (78%), synovial sarcoma (30% - 90%), ossifying fibromyxoid tumors, and EWSR1-CREB family rearrangement tumors [19] [21] [22]. In clinical practice, ossifying fibromyxoid tumor is characterized by peripheral metaplastic bone and fibromyxoid matrix [23], and the morphological characteristics of synovial sarcomas typically do not coincide with those of LGFMS [24]. SEF has the potential for cystosis and calcification, and the typical SEF can be seen microscopically with epithelioid tumor cells arranged in the nest or cord, uniformly embedded in the dense collagen matrix [25]. Several SEF and LGFMS exhibit comparable morphological, immunohistochemical, and molecular characteristics, necessitating genetic testing for accurate differentiation [26].

Additionally, several prior cases demonstrated positivity for EMA, CD99, and Bcl-2, with EMA expression varying from 43% to 91% [27] [28]. A few instances were focally positive for SMA, desmin, CD34, and cytokeratin; however, staining for S100 protein, GFAP, H-calciformin beta-catenin MDM2, and CD117 is generally negative [3] [27] [28]. Negative STAT6 immunohistochemical staining assists in ruling out SFT [29]. The histopathological findings of this case, along with the strong and diffuse positive immunohistochemical staining of MUC4, together with negative immunohistochemical staining for EMA, SMA, STAT6, S-100, etc., provide compelling evidence supporting the diagnosis of LGFMS. Ki-67 serves as an indicator of the proliferative activity of endogenous cell populations within malignant tumors, and it is also utilized to assess the invasive potential of these tumors. In this instance, the proportion of Ki-67 positive cells within the LGFMS tumor cells was determined to be 2%, suggesting a relatively low level of tumor malignancy.

Radiological examination plays an indispensable role in the diagnosis, grading, and follow-up of LGFMS. On CT and MRI scans, initial presentations of LGFMS typically manifest as well-defined, solitary solid-cystic masses, whereas recurrent lesions may appear as multiple invasive masses [30]. Conventional CT imaging reveals LGFMS as a heterogeneous mass with predominantly low attenuation compared to surrounding muscle tissue [31]; areas of low density suggest mucinous components while isodense regions indicate dense tumor cellularity. In certain cases, calcifications may present as punctate hyperdense opacities [32]. Contrast-enhanced CT often demonstrates peripheral enhancement along with internal heterogeneity within most masses [6] [31]. MRI provides an enhanced characterization of LGFMS through subtle multi-sequence imaging techniques previously documented in literature [6] [31] [32]. Mucinous areas generally exhibit low signal intensity on T1-weighted images and high signal intensity on T2-weighted images, while fibrous structures display hypointensity on both T1W and T2W sequences. Following gadolinium contrast administration, the fibrous region shows

mild enhancement; conversely, the mucoid area displays varying degrees of enhancement potentially correlated with blood vessel distribution density within that region [31]. The MRI findings in our case were characteristic and align with previously reported pathology results.

The imaging characteristics of LGFMS lack specificity and must be differentiated from a range of benign and malignant soft tissue lesions. LGFMS typically manifest in the deep soft tissues of younger individuals, whereas myxofibrosarcomas predominantly localize in the subcutaneous layer of the extremities in older adults. Myxofibrosarcomas demonstrate an infiltrative growth pattern around the fascia, which appears as a high-signal spiculated margin on T2-weighted imaging. Additionally, low-grade myxofibrosarcomas are more frequently associated with necrosis and hemorrhage compared to LGFMS [33] [34]. Myxoid liposarcoma is identifiable on MRI by its septate adipose tissue, which may exhibit an amorphous, lace-like, or linear appearance, and occasionally presents as small nodules within the lesion. The adipose tissue demonstrates a high signal intensity on T1-weighted images (T1WI) and a relatively lower signal intensity on T2-weighted images (T2WI) compared to the mucoid component [35] [36]. Myxoid neurofibroma is intricately associated with neural structures, and the presence of nerve entries and exits within the masses can be observed [37]. Furthermore, it is essential to differentiate LGFMS from conditions such as fibromatosis, solitary fibrous tumour, nodular fasciitis, and synovial sarcoma. 18F-FDG PET/CT serves as a highly effective modality for monitoring tumor metastasis and recurrence; tumors exhibiting elevated uptake rates of 18F-FDG demonstrate heightened metabolic activity. A case study by Yoshimura *et al.* involving primary pulmonary LGFMS, recorded a maximum standardized uptake value (SUVmax) of 5.59 during preoperative 18F-FDG PET/CT assessment [38]. Furthermore, Zhang reported a remarkable metabolic uptake rate reaching up to 11.2 in recurrent instances of LGFMS [5].

Numerous previous studies have indicated that a characteristic feature of LGFMS is the specific translocation t(7; 16) (q33; p11), resulting in the formation of the FUS-CREB3L2 fusion gene [39]. This genetic marker aids in distinguishing LGFMS from morphologically similar tumors with greater precision. A limited number of cases involve the FUS::CREB3L1 or EWSR1::CREB3L1 gene fusions [28] [40].

Currently, radical surgery is the preferred treatment modality for LGFMS. Patients who underwent early and aggressive surgical intervention with negative margins exhibited a reduced risk of recurrence and an extended interval between relapses [41]. In addition to surgical margins, tumor size and invasion into adjacent tissues are also critical factors influencing the prognosis of LGFMS [5]-[14]. Recent research has demonstrated that age and the presence of metastasis at the time of diagnosis are independent prognostic factors for overall survival in patients with LGFMS [41] [42]. Despite the high likelihood of curing localized LGFMS through surgery alone, with an overall survival rate approaching 100% at

the 10-year mark, patients who have undergone treatment still require long-term follow-up [43]. The French study specifically recommends a follow-up period of at least 10 years post-surgery [42]. The impact of chemotherapy and radiotherapy on the long-term prognosis of LGFMS warrants further investigation. C. Giani has suggested that pazopanib demonstrates superior therapeutic efficacy in treating LGFMS compared to other commonly utilized chemotherapeutic agents [26]. Radiotherapy is typically employed in scenarios where the lesion is extensive and deemed unresectable.

4. Conclusion

In summary, this report presents a representative case of LGFMS in the lower extremities and provides a comprehensive review of the relevant literature. Histopathological examination is an essential diagnostic modality for LGFMS, while imaging studies offer critical information that aids in both diagnosis and treatment planning for this condition. Currently, surgical resection remains a crucial method for treating this type of tumor. Patients who have undergone treatment should be monitored over an extended period to vigilantly detect any recurrence or distant metastasis of LGFMS.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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