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Case Report

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Beneath the Surface: Exploring the Complexities of Fibromyxoid Sarcoma

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HIGHLIGHTS

- Rare tumor mimics uterine fibroid
- Retroperitoneal origin causes confusion
- Fibrous myxoid pattern on histology
- MUC4 positivity confirms diagnosis
- Complete excision ensures treatment

Key Words:
Fibromyxoid sarcoma
Soft tissue tumour
Retroperitoneal

ABSTRACT

Introduction: Low-grade fibromyxoid sarcoma (LGFMS), or Evans tumor, is a rare soft tissue sarcoma with fibrous and myxoid areas, whorled pattern, and low cellularity. It comprises <5% of cases, commonly affecting trunk and extremities, and rarely the retroperitoneum or mediastinum. **Aim & Objectives:** To present a rare case of low-grade fibromyxoid sarcoma presenting as a pelvic mass mimicking a broad ligament fibroid and to highlight its diagnostic challenges and management. **Case Presentation:** A 50-year-old female (P2L2) presented with 2 months of amenorrhea and a history of T2DM and hypertension. Examination revealed a normal-sized anteverted uterus with a hard, non-tender right forniceal mass. Ultrasonography suggested a 78 × 62 mm hypoechoic pelvic lesion, likely a broad ligament or exophytic fibroid. Intraoperatively, a retroperitoneal mass (8 × 4 cm) above the right internal iliac vessels was identified, excised, and sent for histopathological examination. **Results:** Histopathological examination confirmed the diagnosis of low-grade fibromyxoid sarcoma. The tumor showed low to moderate cellularity with bland spindle-shaped (fusiform) cells arranged in a whorled pattern within a collagenized stroma, along with abrupt transition to myxoid areas. Immuno histochemistry typically shows MUC4 positivity, and cytogenetic analysis is associated with t(7;16) (q32–34; p11) translocation resulting in FUS–CREB3L2 fusion. **Conclusion:** Low-grade fibromyxoid sarcoma is a rare soft tissue tumor that can mimic a fibroid when arising in the retroperitoneum, leading to diagnostic difficulty. Accurate diagnosis relies on histopathology and immunohistochemistry. Complete surgical excision with wide margins remains the mainstay of treatment, with or without adjuvant radiotherapy.

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INTRODUCTION

Low grade fibromyxoid sarcoma (LGFMS), also known as Evans tumor, is a rare and distinctive soft tissue neoplasm characterized by deceptively benign histological features but a clinically malignant course with potential for late recurrence and metastasis. First described by Evans in 1987, LGFMS represents a unique entity within the spectrum of fibroblastic tumors, accounting for less than 5% of all soft tissue sarcomas [1,2]. It typically affects young to middle-aged adults, with a median age of approximately 34 years and a slight male predominance [3,4].

Histologically, LGFMs demonstrate a characteristic admixture of fibrous and myxoid areas arranged in a whorled or swirling growth pattern. The tumor is composed of bland spindle-shaped fibroblastic cells with low to moderate cellularity, minimal atypia, and distinctive curvilinear or arcuate vascular channels [5,6]. Additional features such as collagen rosettes and abrupt transitions between fibrous and myxoid zones further aid in diagnosis [7]. Despite this innocuous microscopic appearance, LGFMS exhibits aggressive biological behavior over long-term follow-up [8].

Immunohistochemistry plays a crucial role in confirming the diagnosis. MUC4 has emerged as a highly sensitive and specific marker for LGFMS and is widely used in routine pathological practice [9]. At the molecular level, most tumors harbor a characteristic chromosomal translocation t(7;16) (q32–34;p11), resulting in a FUS–CREB3L2 gene fusion, while rare variants involve EWSR1 rearrangements [10,11]. These molecular features are considered diagnostic hallmarks and help distinguish LGFMS from histological mimics.

Clinically, LGFMS commonly arises in the deep soft tissues of the trunk and extremities, particularly the thigh, but rare occurrences in the retroperitoneum, mediastinum, and head-and-neck region have been reported [12,13]. The tumor often presents as a slowly enlarging, painless mass and may remain asymptomatic for prolonged periods, contributing to delayed diagnosis [14]. Imaging modalities such as MRI typically reveal well circumscribed lesions with heterogeneous signal characteristics corresponding to fibrous and myxoid components [15].

Long-term studies have demonstrated that LGFMS is associated with a significant risk of recurrence (up to 64%), metastasis (approximately 45%), and disease-related mortality (around 42%) [8]. Notably, recurrence and metastasis may occur decades after initial treatment, emphasizing the need for prolonged surveillance. Metastases most commonly involve the lungs, pleura, and chest wall [4]. The primary treatment remains complete surgical excision with negative margins, while radiotherapy may be considered in selected cases with close or positive margins [3,6].

Given its rarity, atypical presentations, and potential to mimic benign conditions-especially in unusual locations such as the retroperitoneum-LGFMS poses a diagnostic challenge. Awareness of its clinicopathological and molecular features is essential for accurate diagnosis and optimal management.

CASE PRESENTATION

A 50-year-old woman, para 2, living 2 (P2L2), presented with a history of amenorrhea for two months. She reported previously regular menstrual cycles occurring every 28–30 days, with bleeding lasting 3–4 days, not associated with clots or dysmenorrhea. She was a known case of type 2 diabetes mellitus and hypertension for the past two years and was on regular medical treatment. On general examination, the patient was stable. Per speculum examination revealed a pulled-up cervix. On per vaginal examination, the uterus was anteverted and normal in size. The left fornix was free and non-tender, while a hard, non-tender mass was palpable in the right fornix.

Radiological evaluation with ultrasonography demonstrated a solid hypoechoic space-occupying lesion measuring 78 × 62 mm in the right side of the pelvis, near the midline. The findings were suggestive of a broad ligament fibroid or an exophytic uterine fibroid. Based on the clinical and radiological findings, the patient was planned for total abdominal hysterectomy with bilateral salpingo-oophorectomy. Preoperatively, bilateral ureteric stenting was performed to prevent ureteric injury. Intraoperatively, the uterus was found to be normal in size, and both fallopian tubes and ovaries appeared normal. However, a large retroperitoneal mass measuring approximately 8 × 4 cm was identified above the right internal iliac vessels. The mass was noted to receive vascular supply from branches of the iliac vessels and was adherent to the lateral pelvic wall, indicating its retroperitoneal origin rather than uterine pathology. Careful dissection was performed, and the mass was completely excised. The specimen was sent for histopathological examination.

Gross examination (**Figure 1**) showed a well-circumscribed mass with a firm consistency. The cut section (**Figure 2**) revealed a characteristic whorled appearance with alternating fibrous and myxoid areas. Histopathological examination (**Figure 3**) demonstrated features consistent with low-grade fibromyxoid sarcoma, including low to moderate cellularity, bland spindle-shaped fibroblastic cells, and a mixture of fibrous and myxoid stroma arranged in a whorled pattern. The pathological staging was reported as pT, pN, and pM, not assigned/not applicable. Postoperatively, the patient was managed in the intensive care unit and had an uneventful recovery. Once hemodynamically stable, she was referred to adjuvant radiotherapy considering the tumor's malignant potential and risk of recurrence. This case highlights the diagnostic challenge posed by retroperitoneal tumors mimicking gynecological conditions such as fibroids, emphasizing the importance of intraoperative vigilance and histopathological confirmation.

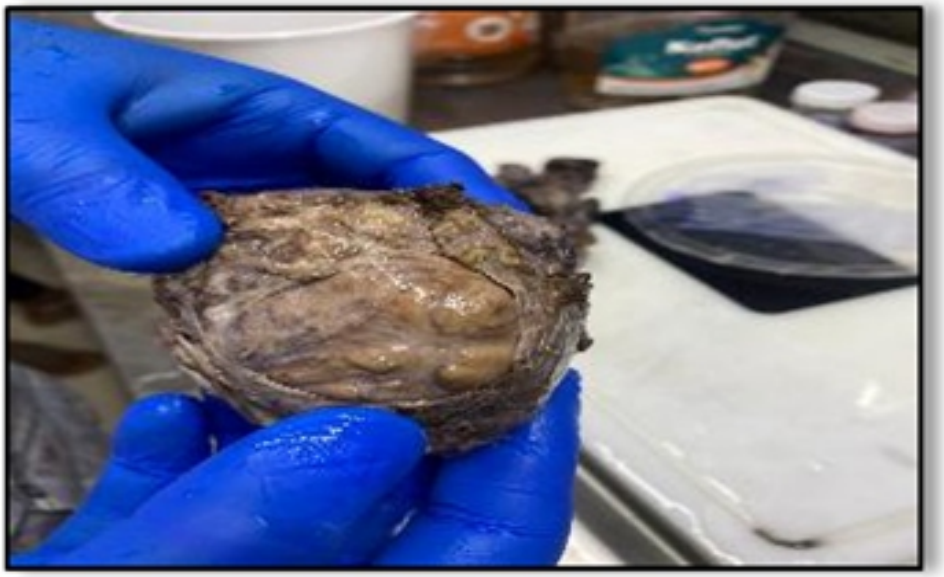

Figure 1: Gross appearance of fibromyxoid tumor

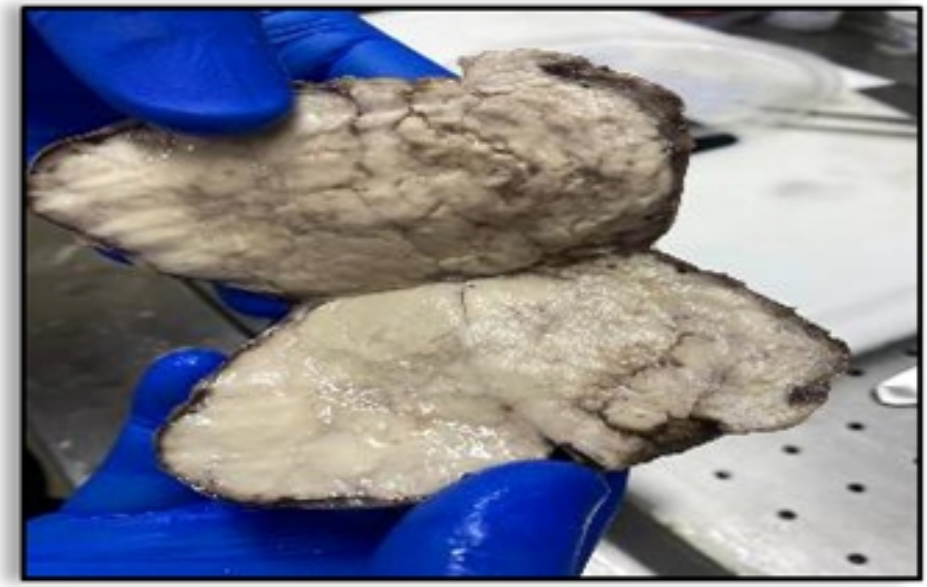

Figure 2: Cut section of fibromyxoid tumor

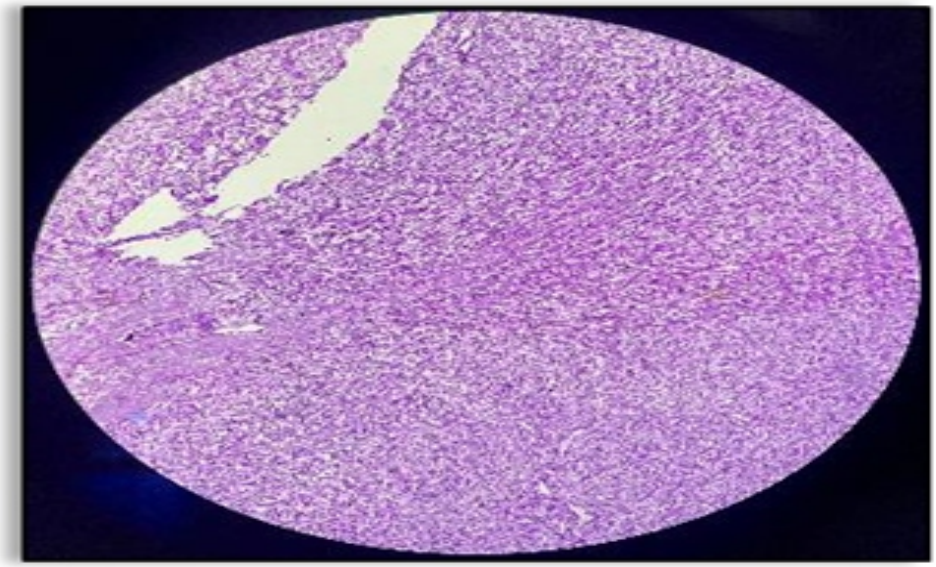

Figure 3: Histological image of fibromyxoid sarcoma

RESULTS

Histopathological examination of the excised mass confirmed the diagnosis of low-grade fibromyxoid sarcoma. Microscopy revealed a characteristic admixture of fibrous and myxoid areas with low to moderate cellularity, composed predominantly of bland spindle-shaped fibroblastic cells arranged in a whorled pattern. Curvilinear blood vessels were also noted, supporting the diagnosis. The tumor was completely exercising intraoperatively, and surgical margins were grossly free of disease.

Postoperative recovery was uneventful, with no immediate complications. The clinical and intraoperative findings correlated with the histopathological diagnosis, highlighting the unusual retroperitoneal origin and its preoperative misdiagnosis as a fibroid. Based on the known biological behavior of LGFMS, the patient was advised to monitor regular and long-term follow-up for recurrence or metastasis

DISCUSSION

Fibromyxoid sarcoma, also known as Evans tumor, is a rare soft tissue sarcoma that is histologically low grade but clinically significant due to its potential for late recurrence and metastasis [8]. Despite its deceptively benign microscopic appearance, it behaves as a malignant tumor over long-term follow-up, a paradox that has been consistently emphasized in the literature. As noted in the present case, its occurrence in the retroperitoneum and clinical resemblance to a fibroid further complicate the diagnosis. Similar diagnostic challenges have been described by Arnaoutoglou et al., who reported that LGFMS may mimic benign soft tissue lesions, leading to delayed recognition [3].

Histologically, LGFMS is characterized by a mixture of fibrous and myxoid areas arranged in a whorled growth pattern. The tumor typically shows low cellularity, composed of bland spindle-shaped fibroblastic cells, along with characteristic curvilinear or arcuate blood vessels. These findings are consistent with those described by Folpe et al., who further identified features such as collagen rosettes and occasional higher-grade areas, thereby broadening the histological spectrum [4]. Guillou et al. also highlighted the morphological variability and its overlap with related entities, reinforcing the importance of thorough histopathological assessment [7].

Immunohistochemistry plays a crucial role in confirming the diagnosis. MUC4 is the most sensitive marker for LGFMS and is widely used in practice. Doyle et al. demonstrated that MUC4 shows high sensitivity and specificity, making it a reliable diagnostic marker, especially in differentiating LGFMS from other spindle cell neoplasms [9]. At the molecular level, most cases demonstrate a characteristic gene fusion involving FUS and CREB3L2. Kurisaki-Arakawa et al. and Mertens et al. confirmed that this translocation is a defining molecular feature and aids in diagnostic confirmation [5,11].

LGFMS accounts for less than 5% of all soft tissue sarcomas and commonly affects young to middle-aged adults, with a median age of around 34 years and a slight male predominance [3,6]. The tumor most frequently arises in the trunk and extremities, particularly the thigh, although rare cases have been reported in the retroperitoneum and mediastinum [12,13]. Clinically, the tumor often presents as a slow-growing, painless soft tissue mass and may remain asymptomatic for a prolonged period, contributing to delayed diagnosis, as also emphasized by Kim et al. in their systematic review [6].

Although initially considered indolent, more recent long-term studies have demonstrated that LGFMS has a significant risk of recurrence (approximately 64%), metastasis (about 45%), and disease-related mortality (around 42%) [8]. Recurrences are more likely in cases with positive or uncertain surgical margins and may occur even up to 15 years after initial treatment, with a median interval of approximately 3.5 years. Metastases most commonly involve the lungs, pleura, and chest wall, and can present decades later, even up to 45 years after diagnosis [4,8].

The mainstay of treatment is complete cytoreductive surgical excision with negative margins. Radiotherapy may be used as an adjunct in selected cases, particularly when margins are close or positive, as supported by Arnaoutoglou et al. and Kim et al. [3,6]. Given its potential for late recurrence and metastasis, long-term follow-up is essential for optimal patient outcomes [14,15].

CONCLUSION

Low-grade fibromyxoid sarcoma is a rare soft tissue tumor that presents a diagnostic challenge due to its deceptively benign histological appearance and its ability to mimic more common benign conditions, particularly when occurring in atypical locations such as the retroperitoneum. This case emphasizes the importance of maintaining a high index of suspicion in patients presenting with atypical pelvic masses. Definitive diagnosis relies on careful histopathological evaluation, supported by immunohistochemical markers such as MUC4 and, where available, molecular studies demonstrating FUS–CREB3L2 gene fusion. Complete surgical excision with adequate margins remains the mainstay of treatment and is crucial in reducing the risk of recurrence. Despite its low-grade nature, LGFMS has a significant potential for late recurrence and distant metastasis, sometimes occurring many years after initial treatment. Therefore, prolonged and vigilant follow-up is essential to ensure early detection and management of disease recurrence, ultimately improving patient outcomes.

LIMITATIONS & FUTURE PERSPECTIVES

The study was limited by its single-centre design, relatively small sample size, and short duration, which may restrict gener-

alizability. Future research could focus on multi-center studies with larger cohorts to validate findings, evaluate long-term outcomes, and explore innovative diagnostic and management strategies for appendicular perforation, improving patient prognosis and reducing complications.

ABBREVIATIONS

LGFMS: Low-grade fibromyxoid sarcoma
SOL: Space-occupying lesion
USG: Ultrasonography
HPE: Histopathological examination
T2DM: Type 2 diabetes mellitus
HTN: Hypertension
IHC: Immunohistochemistry

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AUTHOR CONTRIBUTIONS

All authors significantly contributed to the study conception and design, data acquisition, or data analysis and interpretation. They participated in drafting the manuscript or critically revising it for important intellectual content, consented to its submission to the current journal, provided final approval for the version to be published, and accepted responsibility for all aspects of the work. Additionally, all authors meet the authorship criteria outlined by the International Committee of Medical Journal Editors (ICMJE) guidelines.

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CONFLICT OF INTEREST

Authors declared that there is no conflict of interest.

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CONSENT FOR PUBLICATION

All necessary consent for publication was obtained by authors.

DATA AVAILABILITY

All data generated and analyzed are included within this research article. The datasets utilized and/or analyzed in this study can be obtained from the corresponding author upon a reasonable request.

USE OF ARTIFICIAL INTELLIGENCE (AI) & LARGE LANGUAGE MODEL (LLM)

The authors confirm that no AI & LLM tools were used in the writing or editing of the manuscript, and no images were altered or manipulated using AI & LLM.

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