International Surgery Journal
Rachna et al. Int Surg J. 2026 Sep;13(9):1790-1794
https://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: https://dx.doi.org/10.18203/2349-2902.isj20263014

Case Report

A rare case of heterologous breast leiomyosarcoma in a pregnant female: challenges in diagnosis and management

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Received: 09 July 2026
Accepted: 13 August 2026

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ABSTRACT

Primary breast sarcomas are exceptionally rare malignancies, accounting for less than 1% of all breast neoplasms and fewer than 5% of all soft tissue sarcomas. Primary leiomyosarcoma (LMS) of the breast is a rarer subtype, comprising less than 0.0006% of all breast malignancies. Fewer than 90 cases have been documented in the global literature since the disease was first described. The occurrence of a malignant phyllodes tumour harbouring heterologous leiomyosarcoma during pregnancy represents a rare case presentation. We present the case of a 37-year-old G2P1L1A0 women with gestation period of 24 weeks presented with a left breast lump of one year duration of size 10x8 cm that was painless and without any concerning features. Patient did not have any comorbidities or a significant family history. Ultrasonography was suggestive of a benign lactational adenoma. Core biopsy revealed borderline phyllodes tumour. Simple Mastectomy was done in second trimester with the final histopathological diagnosis of malignant phyllodes with heterologous component of leiomyosarcoma. Although breast leiomyosarcomas are rare and carry a more favourable prognosis than other primary non-phyllodes breast sarcomas, the presence of a malignant phyllodes tumour with heterologous LMS component the patient at a substantial risk for local recurrence or delayed hematogenous metastasis. Given that recurrence and metastasis can arise even after prolonged latent periods of 15 to 20 years, rigorous, lifelong clinical and radiological follow-up remains mandatory.

Keywords: Breast leiomyosarcoma, Pregnancy, Malignant phyllodes, Mesenchymal tumour

INTRODUCTION

Primary breast sarcomas are exceptionally rare malignancies, accounting for less than 1% of all breast neoplasms and fewer than 5% of all soft tissue sarcomas.[1] Primary leiomyosarcoma (LMS) of the breast is an even more rare subtype, comprising less than 0.0006% of all breast malignancies. Fewer than 90 cases have been documented in the global literature since the disease was first described.[2-4] Classically, LMS is thought to originate from smooth muscle cells lining the blood vessels or the areolar musculature within the breast parenchyma. it can present independently as a pure mesenchymal tumour.[5] The occurrence of a malignant phyllodes tumour harbouring heterologous leiomyosarcoma during pregnancy represents a rare case presentation.

CASE REPORT

A 37-year-old G2P1L1A0 women with gestation period of 24 weeks presented with a left breast lump of one year duration, insidious onset, and rapidly progressing for 1 month. It is painless, and not associated with redness, local rise of temperature, nipple discharge, trauma, nipple

retractions, ulcerations or skin changes. There is a history of cumulative breastfeeding of 1.5 years during first child birth.

Left breast showed a lump of size 10x8cm was present in all four quadrants. It was globular, the overlying skin stretched and shiny. Dilated veins were present, no peau d' orange or retraction of nipple. On palpation, irregular shaped hard swelling with well-defined margins, fully mobile, with skin pinchable and no lymph-node palpable. Axilla clear. Right breast had no palpable lump or swelling, nipple areola complex normal and axilla clear.

Ultrasonography of bilateral breast was done suggestive of well-defined hypoechoic mass lesion of 10x8.8cm in left breast involving all the quadrants. Fine needle aspiration cytology showed cluster of ductal epithelial cells with inflammatory granulations suggestive of lactational adenoma with nuclear atypia and inflammation. Core needle biopsy was done suggestive of borderline phyllodes tumour with ER, p63, PanCK negative and Ki67- 60-70%, vimentin, Bcl2, CD34 positive. CECT and Mammography was not done due to present pregnancy. Gynecological and Anesthesia clearance and opinion was taken for fetal cardiac monitoring and proper care during the time of anesthesia and then patient was planned for simple mastectomy.

First surgery of a left-sided simple mastectomy was performed, during which a 10 × 15 cm lump was excised with a 2-cm margin of surrounding tissue; the pectoralis major and minor muscles were preserved, and no axillary lymph node dissection was performed.

Histopathology findings suggestive of left malignant phyllodes with heterologous component of leiomyosarcoma of size 11x9x7.5cm tumour with involved deep resected margin. Elevated mitotic rate of 14 per 10 hpf and significant stromal atypia with pleomorphic cells. Immunohistochemistry indicates SMA positive and desmin and myoD1 negative.

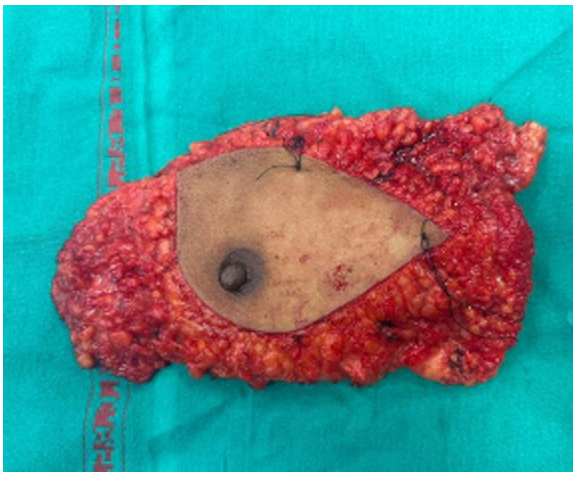

Figure 1: Resected specimen of left simple mastectomy.

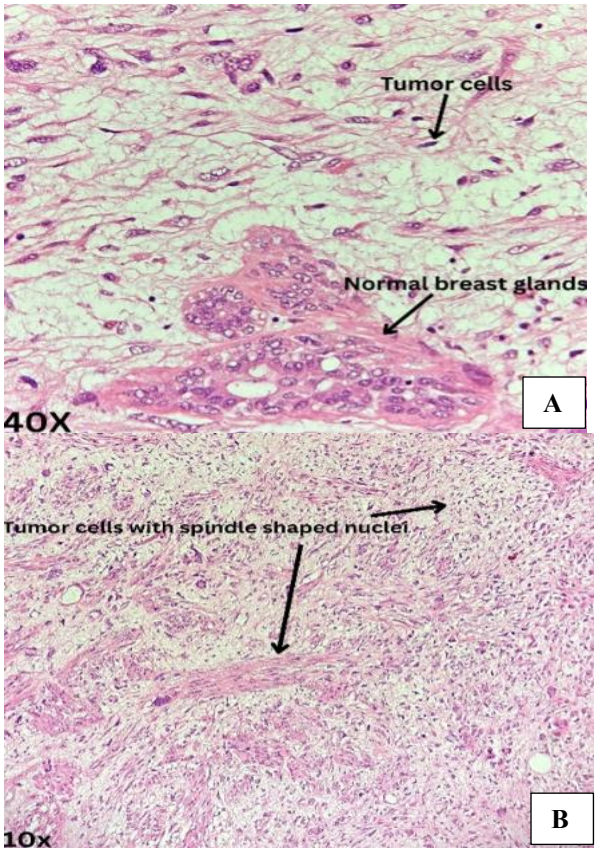

Figure 2: (A) 40X Histopathology picture of left breast mass; (B)10X Histopathology picture of left breast mass.

Because of deep resected margin positive status, patient was taken up for revision surgery. The patient underwent revision surgery involving resection of the pectoralis major muscle on the left side. Overlying skin and subcutaneous tissue were free. No involvement of intercostal muscles, ribs seen. Preserved tissue planes.

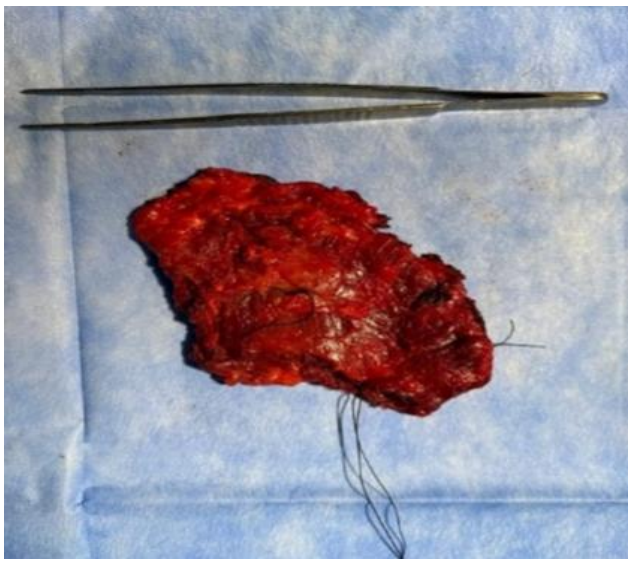

Figure 3: Resected specimen of left pectoralis major muscle.

Histopathology of Resected Left Pectoralis Major muscle: Sections show deposition of leiomyosarcoma. All margins are free from tumor.

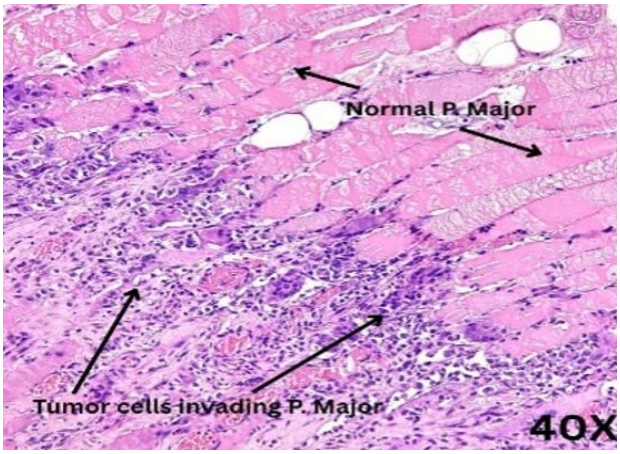

Figure 4: 40X histopathology picture of left pectoralis major muscle showing invasion.

Patient is doing well post-surgery with continuous gynecological assessment and fetal monitoring and further planned for radiotherapy postdelivery.

DISCUSSION

Primary breast sarcomas are rare malignancies, accounting for less than 1% of all breast neoplasms and fewer than 5% of all soft tissue sarcomas.[6] Etymologically, these mesenchymal tumours are thought to originate from smooth muscle cells lining the subsegmental blood vessels or from the areolar/nipple smooth muscle musculature within the breast parenchyma.[5]

While primary breast LMS typically presents independently as a pure mesenchymal tumour, the present case represents an extraordinary, high-stakes oncological intersection: a malignant phyllodes harbouring a heterologous leiomyosarcoma component arising during the 24th week of pregnancy. To the best of our knowledge, this specific presentation in a pregnant patient is virtually unprecedented in global literature.[7] As observed in our patient and supported by the reviewed literature, breast leiomyosarcomas typically manifest as large, painless, well-circumscribed, and mobile masses that can easily mimic benign lesions like fibroadenomas or phyllodes tumour.

Preoperative diagnosis is challenging because physical examinations and imaging modalities are non-specific. On ultrasound, these masses frequently present as well-defined hypoechoic lesions, matching our patient's findings (10x8cm hypoechoic mass involving all quadrants).[8] Furthermore, this case underscores the high probability of diagnostic pitfalls during initial cytological and histopathological assessment.

FNAC limitations: Fine needle aspiration cytology in our patient initially suggested a lactational adenoma with nuclear atypia, likely confounded by gestational physiological changes. This is consistent with literature showing that FNAC is often inconclusive or misleading for stromal malignancies.[4,9] Core needle biopsy: The initial core biopsy pointed towards a borderline phyllodes tumour. It was only after final excisional histopathology of simple mastectomy specimen that the diagnosis of a malignant phyllodes with a heterologous leiomyosarcoma component was made.

The critical role of Immunohistochemistry (IHC): Because soft tissue and stromal breast malignancies exhibit significant histologic diversity, differentiating a pure or heterologous leiomyosarcoma from differential diagnosis such as spindle cell sarcomatoid carcinoma, metaplastic carcinoma, or fibrosarcoma- requires a robust IHC panel.[4]

In our patient's case, the strong positivity of smooth muscle actin (SMA) combined with negativity for epithelial markers (PanCK) and myoepithelial markers (p63) definitively confirmed smooth muscle cell differentiation of the malignant stromal component, validating the diagnosis of a heterologous leiomyosarcoma.

Table 1: IHC markers used in breast sarcomas to differentiate from other types.[10,12]

Marker	Current case (heterologous LMS)	Masadah et al (2023)[8]	Lv et al (2025)[10]	Samenova et al (2023)[11]	Diagnostic significance
SMA	Positive	Positive	Positive	Positive	Confirms smooth muscle origin
Vimentin	Positive	Positive	Positive	Positive	Confirms mesenchymal origin
Desmin	Negative	Negative	Positive	Negative	Variable expression
PanCK/CK	Negative	Negative	Positive (CK7)	Negative	Excludes epithelial/metaplastic carcinoma
P63	Negative	Not specified	Negative	Not specified	Excludes myoepithelial/carcinomatous origin

In genetics, pathology, imaging, medical oncology, radiation oncology, and surgery. Undoubtedly, the management of patients afflicted with colorectal cancer will evolve as advances continue to be made in the multiple disciplines that contribute to the diagnosis and treatment of colorectal cancer.

Surgical strategies and margin status

The primary treatment strategy for breast sarcomas and malignant phyllodes tumours is surgical resection with negative margins (>1cm), achieved via either wide local resection or simple mastectomy.[1,13] Because breast sarcomas preferentially spread hematogenously rather than via the lymphatic system, axillary lymph node dissection (ALND) is not routinely indicated unless clinically palpable or radiographically suspicious lymph nodes are present.[3,14] In our patient, the axilla was clinically and radiologically clear, justifying the preservation of the axillary lymph nodes.

A major clinical hurdle in this case was the positive deep resected margin after the first simple mastectomy, revealing an 11x9x7.5cm tumour with an elevated mitotic rate (14 per 10hpf) infiltrating the deep aspect. The literature emphasizes that the adequacy of the initial surgical margin is the most dominant predictor of local recurrence and overall survival.[13,15] To achieve the local control, the patient promptly underwent aggressive revision surgery involving the resection of the left pectoralis major muscle. The final histopathology of the resected muscle confirmed a deposit of leiomyosarcoma but, crucially, achieved R0 resection with all margins free of tumour.

Gestational considerations and multidisciplinary care

Managing a high-grade malignant breast stromal tumour during pregnancy poses significant therapeutic challenges. Standard contrast-enhanced computed tomography (CECT) and mammography were omitted in our patient to avoid fetal radiation exposure, with ultrasonography preferred for initial assessment in accordance with general gestational oncology principles. Given the high-grade and large size (>5 cm) of the tumour, adjuvant radiotherapy and potentially anthracycline-based chemotherapy would ordinarily be considered to reduce the risk of local and distant recurrence; however, these treatments were deferred until completion of the pregnancy and delivery because of the potential fetal risks.[1,3,4,8]

By utilising an intensive multidisciplinary approach-combining obstetric monitoring, anaesthetic clearance for fetal cardiac stability, and surgical oncology expertise-the patient successfully underwent two major surgical interventions at 24+ weeks of gestation without fetal compromise.

CONCLUSION

Although breast leiomyosarcomas generally carry a favourable prognosis than other primary non-phyllodes breast sarcomas, the presence of a malignant phyllodes tumour with high grade heterologous LMS component, a high mitotic index, and initial deep muscle invasion places this patient at a substantial risk for local recurrence or delayed hematogenous metastasis (commonly targeting the lungs and liver). Achieving completely free margins through R0 pectoralis major resection was an essential prognostic milestone. The postdelivery phase will be critical as immediate adjuvant radiotherapy to chest wall is planned to optimize long-term local disease control. Given that recurrence and metastasis can arise even after prolonged latent periods of 15 to 20 years, rigorous, lifelong clinical and radiological follow-up remains mandatory.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Duncan MA, Lautner MA. Sarcomas of the breast. Surg Clin North Am. 2018;98(4):869-76.
2. Horton L, Wohlfeil M, Al-Kourainy N, Choi L. Rare case of primary leiomyosarcoma of the breast treated with wide local excision and planned cosmetic breast reduction surgery. BMJ Case Rep. 2020;13(11): e236013.
3. Miyazaki C, Shiozawa M, Koike R, Ogihara K, Sasaki Y, Shiba S, et al. Neoadjuvant chemotherapy for primary sarcoma of the breast: a case report. J Med Case Rep. 2019;13(1):289.
4. Amberger M, Park T, Petersen B, Baltazar GA. Primary breast leiomyosarcoma with metastases to the lung in a young adult: case report and literature review. Int J Surg Case Rep. 2018;47: 34-7.
5. Menon G, Mangla A, Yadav U. Leiomyosarcoma. [Updated 2026 Mar 23]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2026
6. Moore MP, Kinne DW. Breast sarcoma. Surg Clin North Am. 1996;76(2):383-92.
7. Alimi A, Abbassi H, Abid S, Dinari A, Khlifi A, Hidar S, et al. Leiomyosarcoma of the breast: case report and review of the literature. Eur J Breast Health. 2025;21(3):277-80.
8. Masadah R, Anwar F, Nelwan BJ, Faruk M. Primary leiomyosarcoma of the breast: a case report and literature review. Int J Surg Case Rep. 2023;106:108290.
9. Kim BR, Lee JH, Cho E, Kim DC, Park YM, Ha DH, et al. Primary breast leiomyosarcoma located in the premammary zone: a case report. Clin Imaging. 2015;39(6):1105-7.
10. Lv H, Ren H, Zhang S, Ding G, Tian P. Case report: primary leiomyosarcoma of the breast. Front Oncol. 2025;15 :1571029.

11. Samenova D, Midlenko A, Khamzina Y, Kaldybayev M, Khamzina S. Primary leiomyosarcoma of the breast: a successful surgical treatment in a 45-year-old woman. Am J Case Rep. 2023;24: e939437.
12. Félix C, Bispo M, Chagas C. Pancreatic metastasis from primary leiomyosarcoma of the breast. GE Port J Gastroenterol. 2019;26(3):223-35.
13. Adem C, Reynolds C, Ingle JN, Nascimento AG. Primary breast sarcoma: clinicopathologic series from the Mayo Clinic and review of the literature. Br J Cancer. 2004;91(2):237-41.
14. Lamyman MJ, Giele HP, Critchley P, Whitwell D, Gibbons M, Athanasou NA. Local recurrence and assessment of sentinel lymph node biopsy in deep soft tissue leiomyosarcoma of the extremities. Clin Sarcoma Res. 2011;1(1):7.
15. Fujita N, Kimura R, Yamamura J, Akazawa K, Kasugai T, Tsukamoto F. Leiomyosarcoma of the breast: a case report and review of the literature about therapeutic management. Breast. 2011;20(5):389-93.

Cite this article as: Rachna, Saha S, Borgharia S, Rattu PK, Singh K, Chandra MRS et al. A Rare case of heterologous breast leiomyosarcoma in a pregnant female: challenges in diagnosis and management. Int Surg J 2026;13:1790-4.