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Case Report

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# First Report of a Primary Breast Leiomyosarcoma with Lymph Node Metastasis in Lebanon: A Case Report

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## Abstract

Background: Primary leiomyosarcoma (LMS) of the breast is an extremely rare histologic subtype of malignant breast tumors. While breast sarcomas account for 0.0006% of all breast malignancies, primary leiomyosarcoma constitutes only 5-10% of breast sarcomas themselves. To date, the total number of reported cases in the literature is less than 70 cases with the first case reported in 1968. This is, to the best of our knowledge, the first case reported from Lebanon, with only 2 cases previously reported in the Middle East, specifically in Morocco.

Case Report: We report the case of a 48-year-old, white, previously healthy female patient, presenting for a breast mass. She first noticed it three months prior to presentation in her left breast and reports that since then, the lump has been growing and her breast has become more painful and erythematous over time. Physical examination revealed an immobile and tender mass of around 12 cm in diameter. A core biopsy was done followed by a left modified radical mastectomy with lymph node dissection. Pathology of the specimen showed a tumor composed of spindle cells arranged in fascicles invading the dermis, with irregular nuclei and prominent nucleoli. Immunohistochemical staining confirmed the diagnosis of high-grade leiomyosarcoma.

Conclusion: We presented this case of leiomyosarcoma with lymph node metastasis to contribute to the scarce literature regarding this disease. Due to the rarity of this diagnosis, not enough data exists regarding treatment and prognosis. Physicians need to review the literature for relevant cases to achieve the best outcome for their patients.

*Keywords:* breast mass, leiomyosarcoma, rare, case report, breast sarcomas, lymph node

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## Background

Breast sarcomas are a rare type of malignant breast tumors that arise from the mesenchymal breast tissue, with an incidence of 0.5-1% [1,2]. Out of 27,881 patients diagnosed with breast malignancies at the Mayo Clinic, only 0.0006% were found to have breast sarcomas [2].

Primary leiomyosarcoma of the breast constitutes a histologic subtype that accounts for 5-10% of all breast sarcomas, making it an extremely rare diagnosis [3]. To date, the total number of reported cases in the literature is less than 70. The first case goes back to 1968, and only 2 cases are reported in the Middle East, specifically in Morocco [3].

To the best of our knowledge, this is the first reported case of primary leiomyosarcoma of the breast in an adult female patient in Lebanon.

## Case Presentation

This is the case of a 48-year-old white female patient presenting for a breast mass. The patient is a nonsmoker, nonalcoholic, with no known food or drug allergies, and an unremarkable past medical, surgical, and family history. She first noted the appearance of an immobile mass in the left breast three months prior to presentation and reports that it was initially small and mildly painful. At that time, the patient was perimenopausal. She sought medical advice and was prescribed antibiotics, but the symptoms did not improve. The mass kept increasing in size and her left breast was becoming more painful and erythematous over time. She noticed the borders becoming irregular but denied any discharge. The physical examination showed an irregular tender mass that was around 12 cm in diameter, located in the left upper outer quadrant. The mass was tender, hard and associated with skin erythema. There was neither discharge nor retraction of the nipple and no lymphadenopathy. The contralateral breast was normal.

Mammography was opted for and showed a 15 x 10 cm spiculated mass occupying the left upper outer quadrant, suspicious for malignancy (BI-RADS 4). Core biopsies were taken and histopathology revealed spindle cells with atypia. Immunohistochemical staining revealed a positive Ki67 of 60% and was negative for all of the HER2, estrogen, and progesterone receptors (HER2-/ER-/PR-).

The patient underwent surgical resection of the left breast mass 3 weeks after biopsies were taken, during which the mass seemed to have doubled in size. Under general anesthesia, a left

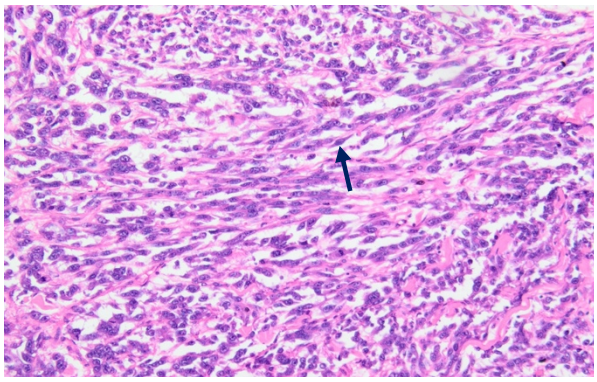

*Figure 1: Histopathology showing tumor composed of spindle cells (arrow) at 10X magnification*

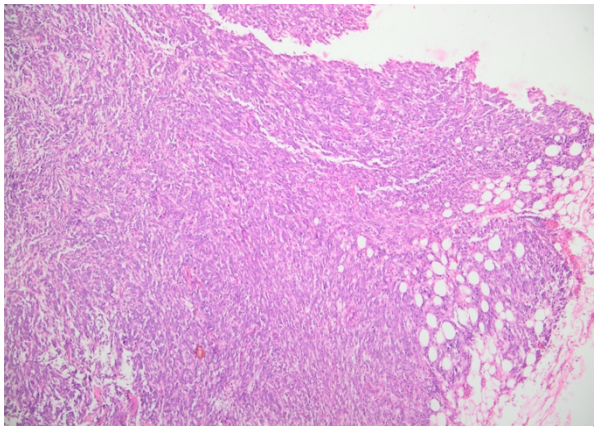

*Figure 2: Histopathology showing the spindle shaped cells arranged in fascicles (4X magnification)*

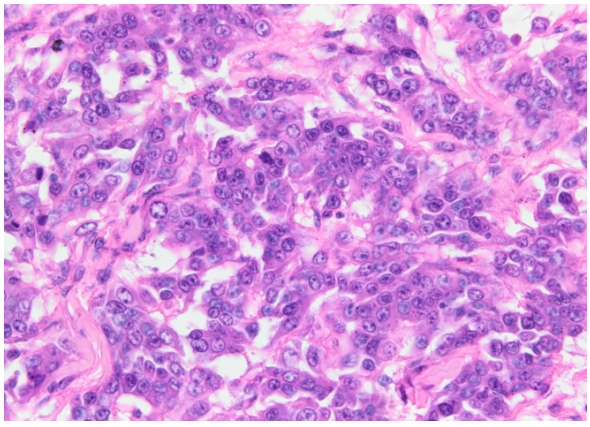

*Figure 3: The cells in this field are epithelioid and have nucleo-cytoplasmic atypia (40X magnification)*

modified radical mastectomy and a level I and II axillary lymph node dissection were carried out successfully. The specimen was removed en bloc and submitted for frozen-section tissue evaluation, which showed negative surgical margins, and the wound was drained and closed. The patient's postoperative course was smooth, and she was discharged home on postoperative day 2.

Resected left breast tissue was submitted with the nipple and overlying skin. It weighed 1195 g and had 25 x 20 x 11 cm dimensions.

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Macroscopically, the tumor was fibro-myxoid, vegetating, gray in color, hard, measuring 12 x 9 x 9 cm with around 50 % necrosis, and infiltrating the superficial skin.

Microscopically, the tumor was composed of spindle cells (Fig. 1) arranged in fascicles (Fig. 2) invading the dermis, with irregular nuclei and prominent nucleoli (Fig. 3). Abnormal mitotic figures and areas of focal necrosis were noted. An abundant myxoid matrix (Fig. 4) was found in one area.

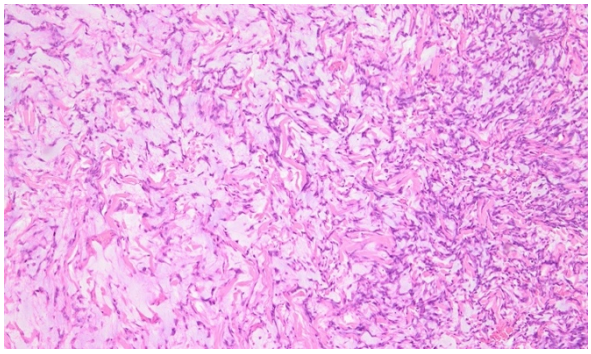

*Figure 4: Histopathology showing myxoid stroma (10X magnification)*

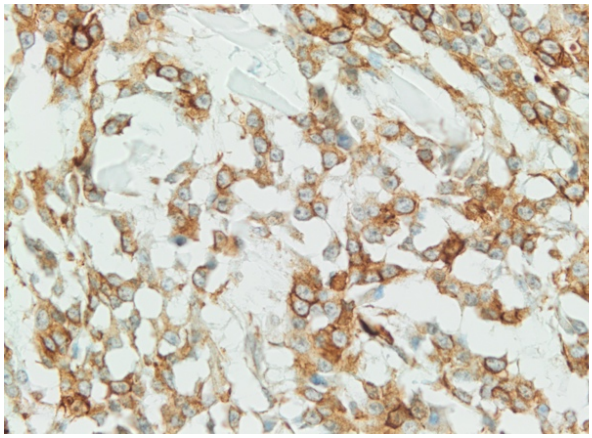

*Figure 5: Immunohistochemistry showing SMA positive neoplastic cells (40X magnification)*

Immunohistochemical staining revealed a strong expression of smooth muscle actin (SMA) in the tumor cells (Fig. 5). Cytokeratin was focally positive in the myxoid area, while epithelial membrane antigen (EMA), S100, and CD34 were negative (Fig. 6A-D). Hence the results were significant for high-grade leiomyosarcoma.

Surgical margins and deep muscles were free of tumor. As for axillary lymph nodes, there was metastasis to 1 of 27 lymph nodes with capsular invasion.

PET-CT scan was done 8 weeks post-op and was negative for local disease. However, it showed multiple small FDG-avid lymph nodes in both jugular-carotid regions measuring up to 0.6 x 1 cm, which need surveillance.

The patient did not receive any adjuvant therapy and is still free of any progression after 26 months follow-up.

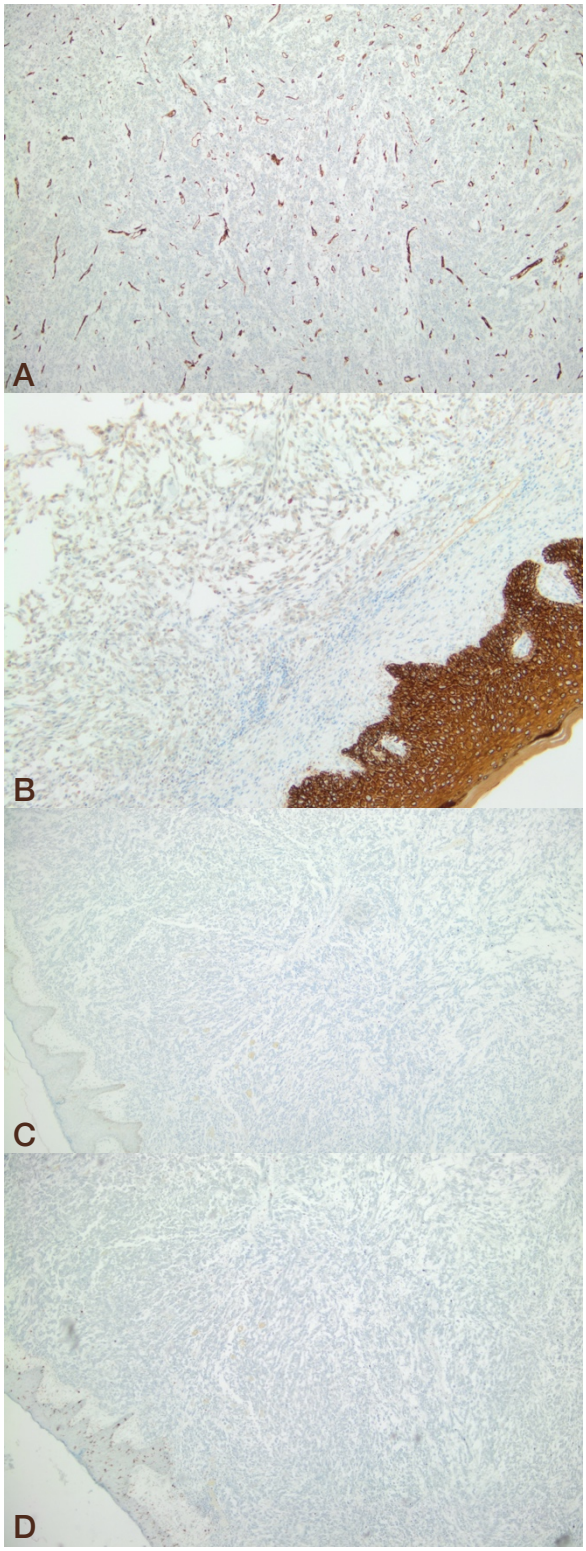


*Figure 6: Immunohistochemistry staining with (A) **CD34** negative in the neoplastic cells but positive in vessels (positive control), (B) **Cytokeratin** focally positive in the myxoid area (positive in surface epidermis - positive internal control), (C) **EMA** negative in neoplastic cells and (D) **S100** negative in neoplastic cells*

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## Discussion

Sarcomas of the breast account for less than 1% of all breast malignancies, and leiomyosarcoma is a rare histologic subtype of the former.

Throughout the cases reported, clinically, breast leiomyosarcoma (LMS) often presents as a large breast lump in post-menopausal women and it may be difficult to distinguish from other breast neoplasms. However, pain is not always a common presentation [4-9].

Mammography and ultrasound findings are also non-specific, as breast LMS appears as a dense, circumscribed, lobulated mass that is often mistaken for most benign and malignant breast neoplasms [3]. The definitive diagnosis is made with a core needle biopsy, or excisional biopsy and confirmed with immunohistochemical stains. LMS stains positive for smooth muscle actin, desmin, vimentin, and muscle-specific actin, and negative for S-100, cytokeratins, and epithelial markers [10-12].

Due to its rarity, there are no prospective randomized controlled trials to guide therapy. Treatment principles have been derived from small retrospective case reviews of breast sarcomas and extrapolated from studies of non-breast soft tissue sarcomas [5]. The cornerstone of treatment is complete surgical excision with free surgical margins [6]. A margin of 1 cm is sufficient to be curative, while also being considered to be the main prognostic factor [7]. Simple mastectomy and wide local excision have been used. Compared to wide excision, simple mastectomy showed no improvement in long-term survival for sarcomas of the breast [8].

The local recurrence rate is around 13% with resection margins less than 1 cm and around 0% with resection margins larger than 1 cm [9, 10].

Breast sarcomas spread mainly through the hematogenous route. Axillary lymph node involvement is rare, occurring in less than 10% of breast sarcomas, so there is no established role for axillary lymph node dissection [11]. Even in cases of breast LMS with clinically palpable axillary nodes, axillary node dissection showed no evidence of metastasis [12].

Until now, there is no clear correlation between axillary lymph node dissection and disease-free survival. However, more studies are needed to better asses the prognosis and disease-free survival of these rare cases [13].

There is still no definite consensus concerning the use of adjuvant chemotherapy or radiotherapy in such cases, with most patients reporting doing well in the few initial years. There is still much controversy regarding the benefits of chemotherapy and radiotherapy in preventing local recurrences which need to be balanced by the high risk of a secondary malignancy induced by ionizing radiation [14]. Also, the impact of adjuvant chemo-radiotherapy on overall survival remains uncertain [14].

## Conclusion

This is, to the best of our knowledge, the first case of leiomyosarcoma of the breast to be reported in Lebanon. We presented this case of LMS with lymph node metastasis to contribute to the literature regarding the clinical presentation, diagnosis, treatment, and prognosis of this disease. When facing this diagnosis, physicians need to review the literature for relevant cases to achieve the best outcome.

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