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Primary Leiomyosarcoma of the Breast: A Rare Case Report

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ABSTRACT

Background: Primary leiomyosarcoma (LMS) of the breast is a very rare neoplasm of the breast arising from the mesenchymal tissue. The clinical presentation of this entity simulates other benign and malignant lesions of mesenchymal tissue of the breast.

Case presentation: Case 1: A 40-year-old female presented with a right-sided breast lump, which was suspected to be a malignant spindle cell tumor on needle core biopsy (NCB). A multi-disciplinary team performed modified radical mastectomy (MRM) with axillary node dissection on the patient with no post-operative chemo-radiation. Case 2: A 70-year-old female presented with a left sided breast lump and a palpable axillary node. Needle core biopsy diagnosed it as malignant spindle cell tumor. The patient underwent MRM with axillary node dissection. It was confirmed to be a case of breast LMS with axillary nodal metastasis. Both patients were followed up for one year with no evidence of recurrence.

Conclusion: Both cases underwent MRM with axillary node dissection in our study. However, the role of axillary dissection in the prognosis and disease-free survival of the patients with primary LMS of the breast with axillary metastasis has not been studied yet. The optimal management of this entity remains to be tumor excision with clear margins.

Introduction

Leiomyosarcoma (LMS) of the breast is a rare entity arising from the mesenchymal tissue of the breast. Overall, the sarcoma of the breast constitutes less than one percent of primary breast neoplasm and less than forty nine cases have been reported in the literature worldwide to date.[1] The current prevalence of primary LMS of the breast is 0.0006%.[2] The most commonly affected age-group is 45-50 years.[3] The presentation of LMS is similar to any other stromal tumor of the breast like malignant phylloides tumor. Therefore, it is difficult to diagnose this entity pre-operatively. Histopathological and immune-histochemical staining become necessary steps to make a final diagnosis. Histologically, LMS of the breast must be distinguished from tumors of mesenchymal origin which are summarized in Table 1. The optimal treatment for this entity has been tumor excision with clear margins. Almost all the articles published advocate the omission of axillary node dissection. Being a rare entity, the role of axillary dissection and adjuvant chemo-radiation in the prognosis and survival of the patients has not been studied yet and remains unearthed. We report this entity with positive axillary node metastasis followed by its implications in assessing the prognosis of the disease.

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Table 1. Diagnostic features of various differentials of spindle cell tumors

Differential Diagnosis	Histopathological Features and Immuno-histochemical Features
Leiomyoma	Bland spindle cells arranged in whorling pattern. Nuclei are blunt-ended, cigar-shaped. No necrosis. SMA(+), Vimentin(+)
Leiomyosarcoma	Infiltrative tumor composed of interlacing bundles of spindle cells. Variable degree of cytologic and nuclear pleomorphism. Areas of necrosis. Brisk mitoses. SMA(+), Desmin(+), H-caldesmon (+), S100(-), CD34(-)
Malignant Phylloides Tumor	Infiltrative border composed of hypercellular stroma with ductal slits. Prominent nuclear pleomorphism, mitoses>10/10 HPF. Vimentin(+), SMA(-), CD117(+)
Malignant Fibrous Histiocytoma	Pleomorphic cells in storiform pattern. Bizarre giant cells. Mononuclear inflammatory cell infiltrate with foamy macrophages present. SMA(-)
Sarcomatoid Carcinoma	Markedly spindle and pleomorphic epithelial cell. Necrosis and nuclear atypia. CK(+), SMA(-)
Fibrosarcoma	Spindle cells in interlacing fascicles giving herring-bone pattern, rare mitoses. Vimentin(+), CK(-), SMA(+)

Case presentation

Case 1

A 40-year-old multiparous female presented with a right-sided breast lump to the out-patient department of Mahavir Cancer Institute and Research Centre, Patna. The patient presented with a lump in the upper-outer quadrant of the right breast for the past 9 years. On clinical examination, there was a lobulated hard mass with well-defined borders measuring 9×9×8 cm. It was fixed to the overlying skin but the underlying muscle was free. Clinically, the axillary nodes were not palpable. There was no family history of breast cancer. Core needle biopsy of the mass demonstrated fascicular arrangement of spindle cells with hyperchromasia and moderate atypia. Occasional atypical ductal cells were also noted. A possibility of malignant spindle cell tumor was considered. The mammography of right breast revealed an oval high density mass with well circumscribed margins. Other investigations included chest scan and abdominal ultrasound which were normal. Uterus and cervix were unremarkable. No other primary lesion was detected even on thorough investigation. A multi-disciplinary team decided to perform modified radical mastectomy (MRM) with axillary node dissection instead of wide local excision with clear margins, because the presence of spindle cells raised the suspicion of it being a metaplastic carcinoma arising from invasive ductal carcinoma and hence the need for axillary node dissection was purported.

Gross examination of the MRM specimen revealed a lobulated solid mass measuring 9×9×8 cm in size with ulceration of the overlying skin. The tumor was 1.8 cm from the underlying muscle. On microscopic examination, spindle cells were arranged in interlacing fascicles with moderate atypia and brisk mitoses. Focal areas of hemorrhage and necrosis were seen. Axillary nodal metastasis was present (Fig 1). IHC analysis showed diffuse

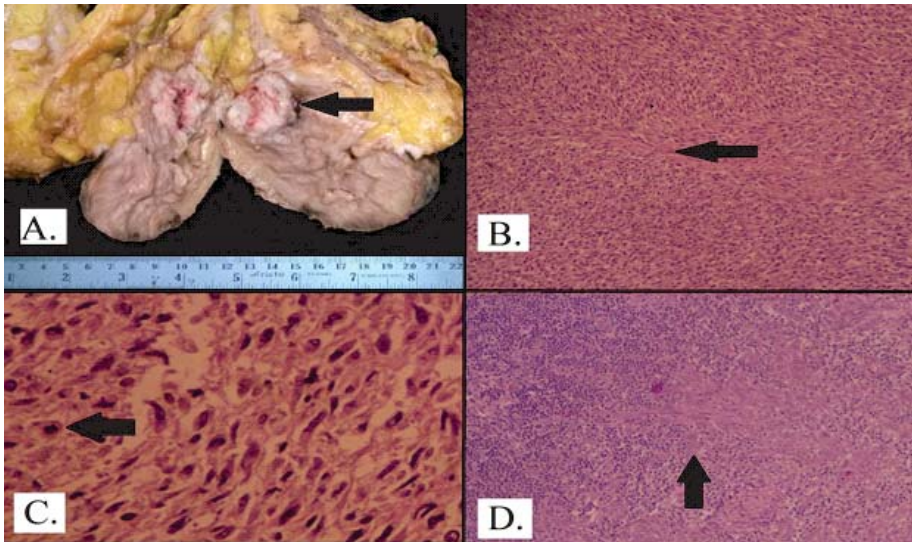

Figure 1. A. Gross photograph showing a well-circumscribed mass with areas of necrosis (arrow) and hemorrhage. B. Low-power photomicrograph showing spondle cell tumor with intersecting fascicles (arrow) H&E, 100X. C. High-power photomicrograph showing frequent mitoses (arrow) H&E, 400X. D. Axillary lymph nodes showing metastasis (arrow) H&E, 100X.

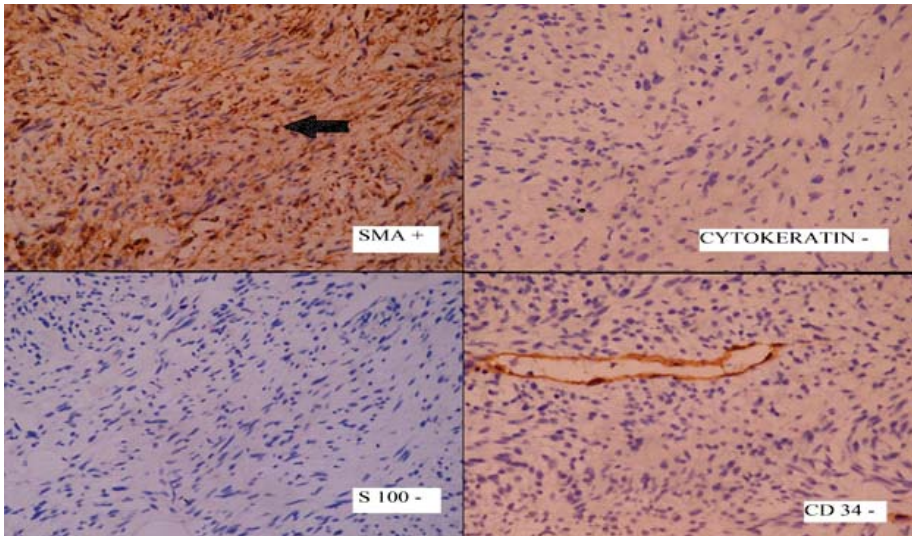

Figure 2. Low-power peroxidase based immunohistochemistry showing diffuse positivity with SMA (arrow) and negativity with CK, S-100, CD34.

immune-reactivity for SMA and vimentin and focal positivity for h-caldesmon. The tumor section were immune-negative for CK, EMA, CD34, desmin, ER, PR, HER-2 (Fig 2). Hence, on histo-pathological and immune-histochemical analysis, a final diagnosis of primary leiomyosarcoma was made. In this case, no post-operative chemo-radiation was administered.

Case 2

A 70-year-old multiparous female presented to the outpatient department with a massive left-sided mass since 5 years ago. On examination, the lump was hard and lobulated with well-defined margins measuring 8×7×6 cm. It was fixed to the overlying skin but the underlying muscle was free. An axillary node was palpable which measured 1.5×1×1 cm. There was no family history of breast cancer. Ultrasonogram of the left breast showed an iso-echoic mass with indistinct margins. Core needle biopsy of the mass revealed a spindle cell tumor with few atypical ductal cells. No other primary lesion was detected on further investigation. The patient underwent MRM with axillary node dissection because of the clinically palpable node which raised the suspicion of it being involved by the primary.

Gross examination of the MRM specimen revealed a well-defined solid mass with irregular margins measuring 7.5×7×6 cm. Microscopic examination revealed intersecting fascicles of spindle cells, diffusely immunoreactive with SMA & vimentin and focally positive for h-caldesmon. It was immunonegative with CK, EMA, CD34, desmin. The axillary nodes were negative for metastasis. This case was thus diagnosed as primary LMS of the breast.

After follow-up of one year, no evidence of recurrence was detected in either of the cases.

Discussion

Primary LMS of the breast is a rare entity encountered in peri-menopausal age group of patients. The most common sites involved by the tumor are the uterus, retroperitoneum, subcutaneous tissue, and gastrointestinal tract.[4] The origin of this tumor is debatable. Some authors consider the origin to be from smooth muscle cells of the blood vessels, while others consider myofibroblasts of the nipple-areolar complex to be the main cell undergoing malignant transformation.[5,6] Core needle biopsy is considered the diagnostic procedure of choice in a suspected case of sarcoma. Generally, the diagnosis is challenging as the clinical picture and the radiological findings are nonspecific. Clinically, the patients present with a large breast lump which is a dense, circumscribed, lobulated mass on radiology showing features similar to phylloides or fibro-adenomas. Therefore, differential diagnosis between LMS and other benign lesions is difficult on image analysis alone. Histopathological examination followed by immunehistochemical analysis is very important for an accurate diagnosis.

Most authors consider LMS as a locally invasive neoplasm and hence advise either simple mastectomy or wide local excision as the best possible management.[7,8] In proven cases of primary LMS of the breast, no correlation has been found between axillary dissection and improvement in disease-free survival. Because the literature suggests that cases undergoing wide local excision/ simple mastectomy are associated with a high incidence of recurrence, they advocate the need for axillary dissection in such cases.[9] However, there is no clear consensus yet. More case reports and case series are needed to study the role of axillary node dissection in the prognosis and disease-free survival of such patients.

Primary LMS of the breast is a rare tumor that is a diagnostic challenge for the clinicians, radiologists, and pathologists, as well. Histopathological examination remains the gold standard but application of ancillary techniques helps with further confirmation of the diagnosis. Microscopically, it can be suspected by typical histological features like high cellularity, fusiform spindle cells with blunt end nuclei, moderate atypia, and frequent mitoses. IHC confirmation by SMA, vimentin and desmin is helpful. The mainstay of treatment remains surgical excision with clear margins and long-term follow-up.

We recommend that more case reports and case series need to be published to determine the exact role of the axillary node dissection in the prognosis and disease-free survival of the patients.

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Conflicts of interest

The authors declare that they have no conflict of interest related to the publication of this manuscript.

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