Primary Leiomyosarcoma of Breast: A case report from Nepal

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Abstract
Leiomyosarcoma is an uncommon form of breast cancer, accounting for 5-10% of all breast sarcomas with only 70⁴ instances recorded to date. Waterwarth described the first instance of primary leiomyosarcoma breast (PLB) in 1968. This is the first case of PLB documented in Nepal, that we’re aware of. Leiomyosarcoma is a malignant non-epithelial smooth muscle tumor that can occur anywhere in the body. We discuss the case of a 41-year-old female who presented with a breast lump that was subsequently confirmed as Primary leiomyosarcoma and was treated with wide local excision with margins. We've been watching her progress for a year.

Keywords: Leiomyosarcoma, lipoma, breast, excision

Introduction
Leiomyosarcomas are malignant tumors originating from the smooth muscle stromal tissue. The occurrence of leiomyosarcoma in the breast is very rare. We discuss a case of this mesenchymal tumor in a 41-years-old female who presented with a breast lump.

Case Presentation
A 41-years-old female presented to the Outpatient clinic of the National Hospital and Cancer Research center after she noticed a lump in her left breast that was painless, without any nipple discharge or any overlying skin changes. She was not under any hormonal or radiation therapy. Her vitals were stable. No axillary lymph nodes were palpable. On examination, a 4 * 4 cm mass could be palpated in the left breast at the 1 o’clock position. The mammogram revealed a single 5 x 4cms, well-defined radiolucent mass in the left breast suggesting benign findings, Breast Imaging Reporting and Data System (BIRADS Category:2). The ultrasonography of the breast showed normal right breast findings and ill marginated echogenic area of 3 x 2.8 cms in upper outer quadrant likely Lipoma was seen.

A fine needle aspiration biopsy was performed but was inconclusive. Tru-Cut biopsy was also done which showed? fibroadenoma. So, an excisional biopsy of the

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mass was planned. The report showed a typical Lipomatous tumor / well-differentiated liposarcoma. The cut section showed a grey yellow capsulated mass measuring 4.5 x 4 cm with a buttery appearance.

Fig 1: Gross pathology of Specimen

Sections: A-K. Microscopy demonstrated capsulated mass composed of lobules of variable-sized adipocytes. There were embedded mammary glands lined by double layer of epithelium. Spindle-shaped stromal cells were also seen with areas of sclerosis. Numerous variable shape lipoblast was scattered. These lipoblasts showed vacuolated cytoplasm and scalloped hyperchromatic nuclei. Mitosis was around 4 per 10 high power fields (hpf) with no atypia or areas of necrosis. The cut margins were free of tumor microscopically.

Post-surgery immunology reports were within normal limits as shown in table 1:

<table>
<thead>
<tr>
<th>Test Name</th>
<th>Result</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>CA15.3</td>
<td>19.58</td>
<td>&lt;35 U/ml</td>
</tr>
<tr>
<td>CA 125</td>
<td>22.8 U/ml</td>
<td>Up to 35</td>
</tr>
<tr>
<td>CA 19.9</td>
<td>24.1 U/ml</td>
<td>Up to 37</td>
</tr>
</tbody>
</table>

CT scan of the chest and abdomen showed a non-enhancing well-defined hypodense area in the upper inner quadrant of the left breast and no evidence of metastasis in the liver, bones, and lungs. Re-excision of the cavity margins was done that showed no residual tumor and the skin and cut margin were free of tumor.

Figure 2: Microscopy of the lesion
Discussions

Due to its rare occurrence, a lot is yet to be known about this type of tumor. Leiomyosarcoma frequently manifests as a big tumoral mass with well-defined borders in postmenopausal women in their fifth to sixth decade. The breast is a rare primary site, with the uterus, retroperitoneum, subcutaneous tissues, and gastrointestinal system being the usual locations. Radiation also seems to be associated with leiomyosarcoma of the breast. Radiation-associated leiomyosarcoma accounts for 8% of all radiation-associated sarcoma patients. A diagnosis of this uncommon tumor is challenging to make by fine needle aspiration cytology and a core biopsy is necessary. Microscopically, the tumors are composed of pleomorphic and hyperchromatic spindle-shaped cells arranged in an interdigitating fascicle. Hyperchromasia in the nuclei, pleomorphism, and mitoses is seen in the cytological analysis. A definitive diagnosis is made by immunohistochemistry, which stains positively for desmin, vimentin, and muscle-specific actin, and negatively for cytokeratin, myoglobin, and S-100.

Treatment strategies for breast sarcomas have been developed based on research on soft tissue sarcoma of the extremities. The primary method of treatment is still surgical excision with negative margins. Fujita et al in their case study suggested at least 3cm of margin excision to be adequate. High-grade sarcomas are more likely to reoccur. Radiotherapy is therefore utilized as an adjuvant to achieve local control and stop recurrence. McGowan TS et al in a study suggested that if negative surgical margins can be achieved, breast sarcoma should be managed by a conservative surgery with postoperative irradiation to a microscopic tumoricidal dose (50 Gy) to the whole breast, and at least 60 Gy to the tumor bed due to high risk of recurrence of the tumor. Due to their tendency for distant metastasis, high-grade, bulky tumors (> 5 cm) have been treated with anthracycline-based chemotherapy. Various studies on breast sarcoma suggest that Ifosfamide and doxorubicin were the most effective chemotherapeutic agents, which can be inferred to breast leiomyosarcoma.
Despite some evidence from other sarcomas that suggests it would be beneficial adjuvant therapy with chemo and/or radiotherapy has not proven successful in treating these tumors.\textsuperscript{[1]} Lymphatic spread and nodal metastasis are not features of sarcoma so axillary dissection is not recommended.\textsuperscript{[10]} A case report from Korea suggests the combination of hyperthermia and pazopanib (VEGF inhibitor) may have had a strong synergistic effect on the chest wall mass of the metastasized primary breast leiomyosarcoma.\textsuperscript{[11]} PLB type of tumor tends to show local recurrence, and distant metastasis has been observed in 25% of patients. Hence, recurrent monitoring of the patient is mandatory.

\textbf{Conclusion}

Primary leiomyosarcoma of the breast is rare cancer. Although few reports have been published on the treatment strategy the definitive management is yet to be unfolded.

\textbf{Consent}

Informed written consent was taken from the patient and the patient's party.

\textbf{References}