Case report:

Primary Breast Sarcoma

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Abstract:
Breast sarcomas are an extremely rare and heterogeneous group of malignancies derived from non-epithelial elements of the gland constituting less than 1% of total breast malignancies and less than 5% of all soft tissue sarcomas. It can be divided into two categories: de novo development (primary) or secondary. These tumors are important as they have a high rate of recurrence and poor prognosis. In addition it is important to differentiate these tumors from breast carcinoma as the treatment differs in these two entities. We present a case of primary breast sarcoma in a 46 year old nulliparous Chinese woman.

Keywords: breast sarcoma; heterogenous; malignancy; non-epithelial; poor prognosis

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Introduction

Sarcoma of the breast was first described by Chelius in 1828. It is an unusual condition accounting for less than 1% of all breast malignancies and less than 5% of all soft tissue sarcomas[1]. It can be divided into two categories: de novo development (primary) or secondary and primary sarcoma of the breast can be further subdivided into four categories namely, cystosarcoma phylloides, lymphoma, angiosarcoma and pure sarcoma.

The main risk factor for development of breast sarcomas is previous radiation treatment for breast carcinoma while chronic lymphoedema is an additional risk factor. Other debatable risk factors include professional exposure to vinyl chloride, artificial implants and patients carrying a mutation in p53[2]. However it must be remembered that the majority of breast sarcomas is present without an identifiable etiologic factor.

Primary breast sarcomas occur over a wide age range, but most (except for angiosarcomas) occur in women in their fifth or sixth decade of life[3,4]. They usually present as painless breast masses that are mobile, often large and mimic fibroadenomas. However, a history of rapid growth in a previous long standing indolent mass should raise the suspicion of a sarcoma. Diagnosis is made with the help of clinical findings, imaging and biopsy which allows a preoperative diagnosis to be made reliably while prognostic factors are size and grade with a trend towards better survival in those with superficial tumors[5,6]. We present a case of left breast sarcoma in University Malaya Medical Center (UMMC).

Case Report

A 46 year old nulliparous Chinese lady presented to a private hospital with history of a left breast lump noticed 3 weeks earlier which appeared to be growing rapidly. There was no history of pain, fever or skin changes. The patient was apparently well before this with no previous medical illness. There was also no family history of breast carcinoma. Examination of the left breast revealed a lump measuring about 3-4 cm in size and there were no palpable axillary lymph nodes. She then had a bilateral mammogram done which revealed a large dense opacity in the upper outer quadrant of the left breast (**Figure 1**).

This was followed by an ultrasound of the left breast which revealed a 2.8 cm x 2.1 cm lesion in the upper outer quadrant of the left breast and was reported to be probably benign (**Figure 2**).

The right breast was normal. She was later referred to UMMC for further evaluation. The informed consent of the patient has been obtained verbally.

At UMMC, the patient had fine needle aspiration cytology (FNAC) of the lesion done twice as the first sample was inadequate for interpretation. The second FNAC revealed presence of stromal tissue

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and pleomorphic spindle shaped cells suggestive of a stromal tumour and biopsy was recommended. She then had a hook wire localization and lumpectomy of the left breast lesion which according to histopathology examination revealed stromal tissue and pleomorphic cells with prominent nucleoli in keeping with sarcoma of the breast (**Figure 3** and **4**). Contrasted computed tomography (CT) scan of the thorax and abdomen did not reveal any evidence of metastasis to the lung, liver or other abdominal organs. She then had a left mastectomy and axillary clearance done 3 weeks later which showed no residual tumour and the lymph nodes showed no evidence of metastasis. Follow up visits of the patient to the hospital over a period of 2 years remained uneventful.

Discussion

Sarcoma of the breast is an unusual condition accounting for less than 1% of all breast malignancies and less than 5% of all soft tissue sarcomas with an overall incidence in United States being about 17 new cases per 1 million women[7]. Thus an in depth understanding of this lesion is lacking due to its relative rarity.

Sarcoma of the breast can be divided into two categories: de novo development (primary) or secondary with histologic subtypes which include malignant fibrous histiocytoma, liposarcoma, fibrosarcoma, clear cell sarcoma, neurogenic sarcoma, leiomyosarcoma and alveolar soft tissue sarcoma[6]. The most common subtype of primary breast sarcoma however is malignant phyllodes tumor and angiosarcoma[5].

Breast sarcoma usually presents as a painless breast mass that is large and mobile. This is the same as that of a typical fibroadenoma, although a history of rapid growth should raise the suspicion of a sarcoma. The age of presentation varies and ranges from prepubertal to post menopausal with a peak incidence in the 40s and 50s. This is one to two decades older than that for a fibroadenoma.

The diagnosis is made based on clinical findings, imaging and biopsy. However, core needle biopsy is the procedure of choice as it facilitates the grading of the tumor and determines the histological sub type[1,2]. Fine-needle aspiration cytology has low accuracy in the diagnosis of this disease and should be avoided if a sarcoma is suspected. Routine mammography is useful to demonstrate the lesion, but is not particularly useful to differentiate from other benign or malignant lesions. It is seen as a dense mass with indistinct margins and is rarely associated with tumor calcification while sonography demonstrates a solid lesion that is indistinguishable from a fibroadenoma and hence necessitates a biopsy of the lesion[3,8].

The mainstay of treatment is surgery. Thus, localized breast sarcomas should be treated by complete surgical excision, which offers a chance for long-term survival. Mastectomy is generally performed for more sizeable tumors. However lumpectomy offers equivalent oncologic results only if it is technically feasible. Obtaining negative surgical margins is more important than the type of surgery which entails a simple mastectomy. Adjuvant radiation may be indicated when the primary tumor is bulky and when local recurrence is a problem[2,9].

The M.D Anderson experience revealed that breast sarcomas smaller than 5 cm have significantly better prognosis, irrespective of any other factor. It has also been established that other tumor characteristics like cellular appearance, infiltrating borders, number of mitoses and stromal atypia have a negligible prognostic role compared to tumor size. Axillary nodal status also had a minimal impact on prognosis[10].

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Ethical clearance

Consent obtained from patient was submitted to University Malaya Ethics Committee prior to submission of this manuscript.

Disclosure of Interest

The author of this case report believes that there is no conflict of interest that could potentially be construed to affect the material contained in the manuscript that is being submitted to the Journal.

Authors's contribution

Data gathering and idea owner of this study: Mansharan Kaur Chainchel Singh

Study design: Mansharan Kaur Chainchel Singh

Data gathering: Mansharan Kaur Chainchel Singh

Writing and submitting manuscript: Mansharan Kaur Chainchel Singh

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