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(CASE REPORT)

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Primary Carcinosarcoma of the breast: Report a very rare case

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Abstract

Background: Sarcoma of the breast is a rare malignancy with heterogeneous histology. angiosarcoma is the most common type of sarcoma of the breast secondary from previous radiation.There is limited clinical and survival information from other types of breast sarcomas.

Case Presentation: Here we present A 56-year-old woman which presented with a mass in her right breast. On examination the tumor was not fixed to chest wall. But skin over mass was erymatous and with nipple retraction.A cor-needle biopsy was performed which showed leiomyosarcoma. After IHC the difintive diagnos was sarcoma. A radical resection of mass with radical mastectomy and axillary dissection were performed, in permanent pathology all margins was free of tumor. After surgery patient refer to radio oncologist for irradiation therapy

Conclusions: Surgical resection seems to be the most important treatment modality in sarcoma of the breast. Radiation therapy added a minor survival benefit to the patients who received surgical resection. Systemic chemotherapy did not any role in sarcoma of the breast.

Keyword: Sarcoma; histology; Carcinosarcoma; Breast cancer; Primary Carcinosarcoma

1. Introduction

Breast sarcoma is extreme rare tumor; it is a group of uncommon tumor arising from mesenchymal tissues of the breast. Breast sarcomas accounted for 0.0006% of all breast malignancies[1]. Cases of breast sarcoma usually present after breast irradiation breast carcinoma and has increased[2-4]. Radical Surgery with complete resection of margin is the only curative therapy for patients with sarcomas of breast[3]. However, a recent randomized trial provided show to support the use of neoadjuvant chemotherapy for patients with soft tissue sarcomas of the extremities and trunk by European sarcoma groups[5,6]. We describe here a case of breast sarcoma treated with radical resection and free margin with radical mastectomy successful. In this case we want to discussed the presentation, diagnosis and treatment with a review of appropriate pathological evaluation.

2. Case presentation

A 56-year-old Iranian post-menopausal woman was referred with a huge right breast mass, which had rapidly increased in size in recent weeks erythema of skin and pod orange appearance. The chest wall and breast mass was started about 6 months prior to presentation. The tumor size was 14 cm in its greatest dimension. It was mobile, skin erythema nipple

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retraction and was not fixed to her chest wall (Fig. 1a, b). Ipsilateral axillary lymph nodes was not palpable. A cor-needle biopsy was performed from mass which showed sarcoma. After Immunohistochemical (IHC) staining established the definitive diagnosis of sarcoma (Fig. 2, a, b). A computed tomography (CT) scan of chest showed a mass of breast with invasion to pectoralis major muscle with no evidence of metastases to other sites of chest and without enlarged axillary lymph nodes (Fig. 3 a, b). After consultation with oncologist we decided to do radical surgery, because of two report abou chest wall sarcoma which chemotherapy is not effective (7,8). We perform a right radical mastectomy with axillary lymph node dissection with radical resection of tumor with a portion of involved pectoralis muscle with free margin during surgery with pathology. On gross examination, the tumor measured 12 × 6 × 8 cm. On histological examination, the main tumor consisted of bundles of spindle cells with well-defined bright eosinophilic cytoplasm, and pleomorphic nuclei (Fig. 2 a, b). Pathological assessment revealed that the axillary lymph nodes and the surgical margin were negative. On immunohistochemical examination, the neoplastic cells were positive for α-smooth muscle actin and desmin, and negative for AE1/AE3, CAM5.2, and S100. The Ki-67 labeling index was approximately 20%. The final diagnosis was carcinosarcoma. A follow-up contrast-enhanced CT scan of her chest and abdomen showed no residue or recurrence at 12 months. She is thriving and was disease free at 1.5-year follow-up.

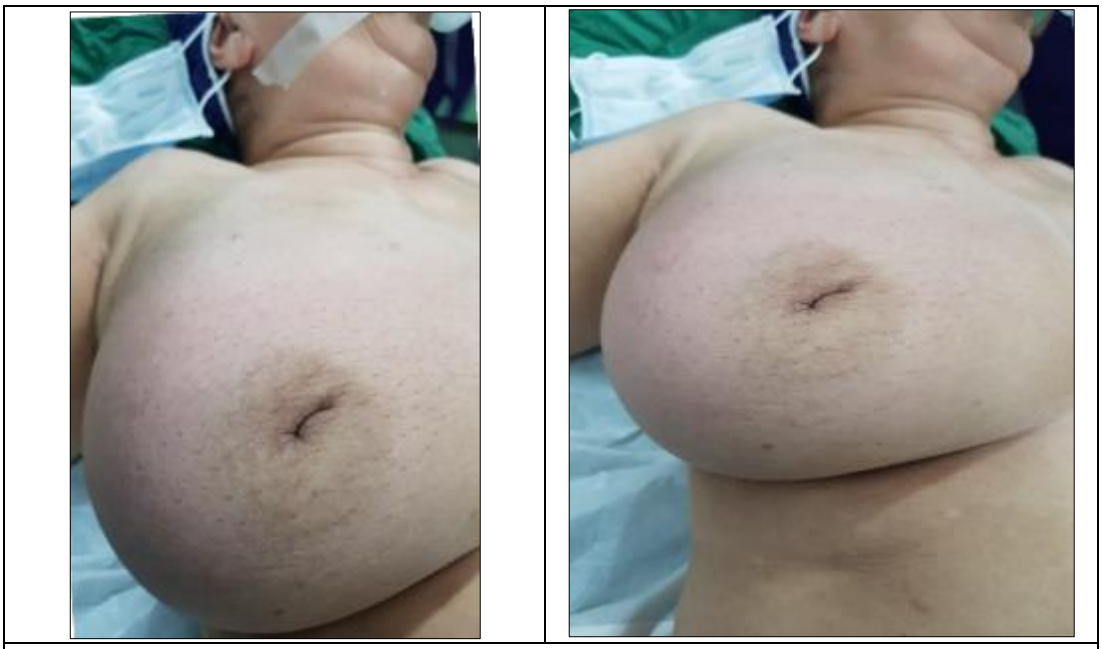


Figure 1a

Figure 1b

Figure 1 a, b The size of the tumor was 14 cm in the largest dimension. Mobile, cutaneous erythema was nipple constriction and was not fixed to her chest wall

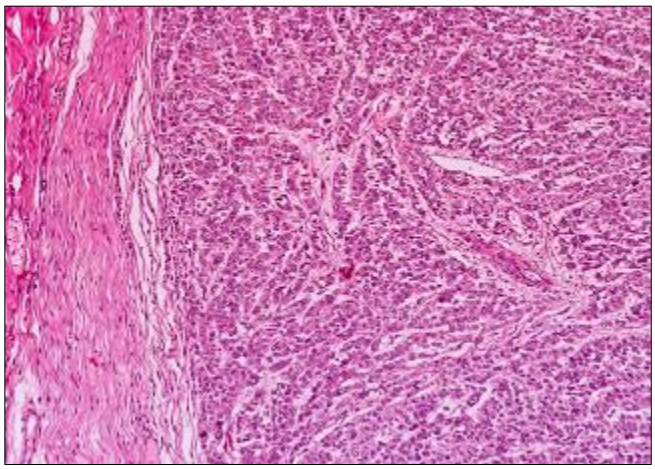
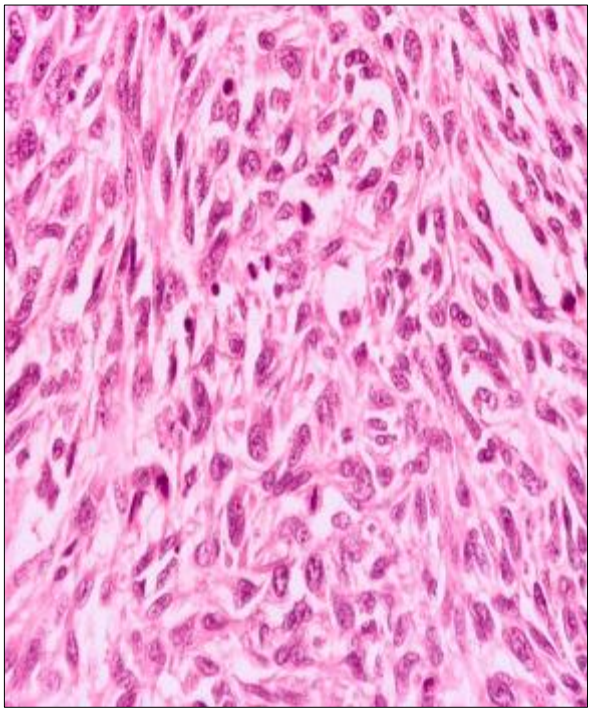


Figure 2a



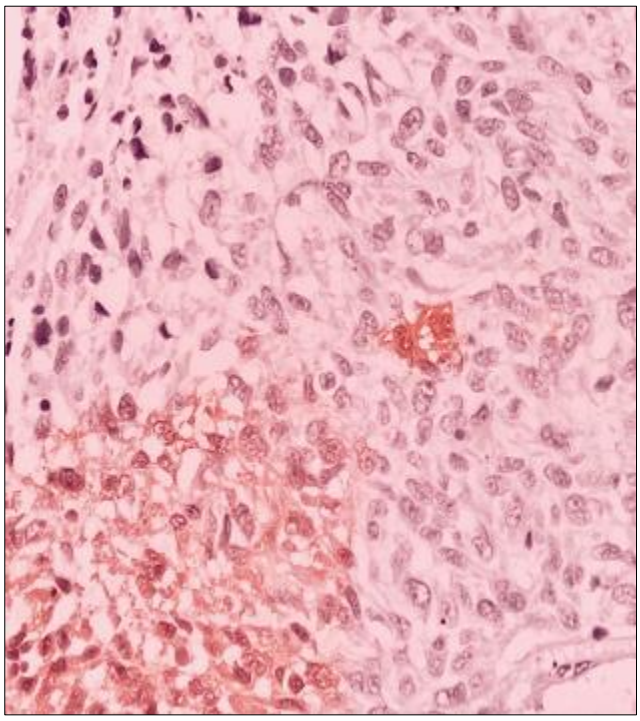


Figure 2 b

Figure 2 a, b Immunohistochemical (IHC) staining provides a definitive diagnosis of sarcoma. On histological examination, the main tumor consisted of bundles of spindle cells with well-defined bright eosinophilic cytoplasm, and pleomorphic nuclei

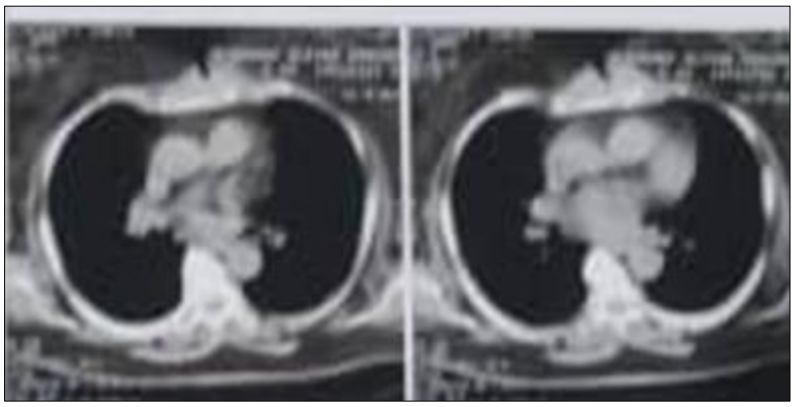


Figure 3 a

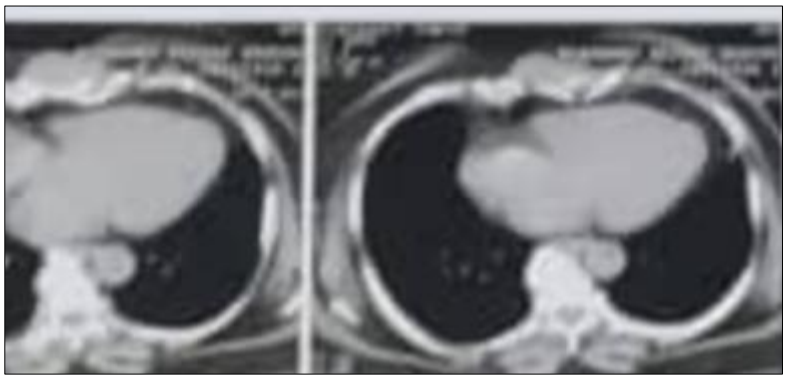


Figure 3 b

Figure 3 a, b A computed tomography (CT) scan of chest showed a mass of breast with invasion to pectoralis major muscle with no evidence of metastases to other sites of chest and without enlarged axillary lymph nodes

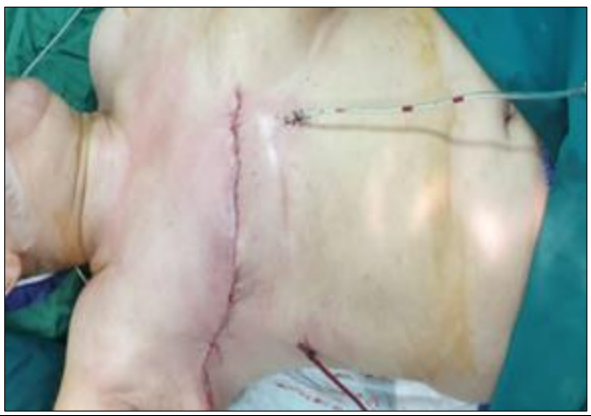


Figure 4 a

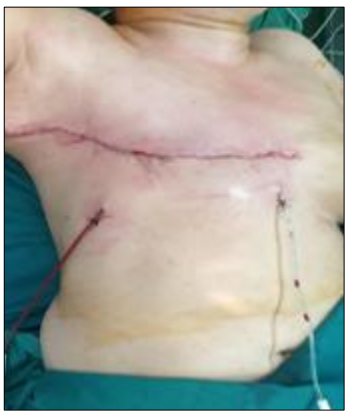


Figure 4 b

Figure 4 a, b Performing radical surgery on the patient

3. Discussion

Sarcomas arise from all part of body with connective tissue , including the breast and chest bwall[7, 8]. Breast sarcomas are defined as a group of mesenchymal malignant tumors similar to other soft tissue sarcomas and excluding malignant phyilods sarcoma[1]. Leiomyosarcoma which arise from the smooth muscle , which is one of the most common soft tissue sarcomas, between 10 and 20% of all soft tissue sarcomas[9]. Leiomyosarcomas of the breast are rare, In the world

literature nearly 50 cases reported[10]. Most report have shown no effective in survival patient with adjuvant chemotherapy for the treatment of soft tissue sarcomas[7-10]. The first reported from a randomized trial for high-risk soft tissue sarcomas of extremities and trunk including leiomyosarcoma was benefit[3]. This trial showed an improved overall survival increased in patients who received neoadjuvant therapy[3, 5, 6]. In the present patient, we used radical surgery and resect all tumor with breast mass which invading the pectoralis major muscle and suggested the use of radiotherapy[7, 8].Recently, neoadjuvant chemotherapy has been used more frequently to treat patients with soft tissue sarcomas[3, 5, 6]. The European Organisation for Research and Treatment of Cancer - Soft Tissue and Bone Sarcoma Group (EORTC-STBSG) proposed that therapeutic response should be evaluated by the change on pathological findings of stainable tumor cells and necrotic area for the entire tumor following neoadjuvant chemotherapy[10].

Lymphatic spread is uncommon in sarcomas. In a prospective analysis of 1722 soft tissue sarcomas, lymph node metastases were present in 2.6% of patients[11]. In the present patient, based on physical examination and imaging studies axillary lymph node metastases were not present and axillary dissection not performed. In sarcoma of breast axillary lymph nodes dissection should be perform individually, depending on the situation for that patient[12]. Radiation therapy may reduce the risk of local recurrence after surgery but does not increase on the survival[3, 7, 8]. In a retrospective study for soft tissue sarcomas of extremity show, that postoperative radiation therapy may improve the survival rate in tumor larger than 5 cm and high-grade lesions[13, 14]. Due to the size of the tumor in this patient, and the fact that it was a high-grade subtype, recommendation is to use radical resection and adjuvant radiotherapy[13, 14]. In my 35 years practice in thoracic surgery I had one patient with sarcoma of breast in a 14 year girl who underwent wide radical resection of right breast with margin free and postoperative radiotherapy, she is alive yet and has a 8 year boy, after nearly 20 year post surgery and live with good condition. Other type of sarcoma of breast are phyllodes tumor, in a report most series considered it a distinct entity from breast sarcoma in view of its epithelial component[1-4, 15]. Other mass of breast tumor is phyllodes tumor which is the most common nonepithelial neoplasm of the breast[16]. However, about 85-90% of phyllodes tumors are benign and that approximately 10-15% are malignant[17].

Like other sarcomas, malignant phyllodes tumors metastasize hematogenously.

The lungs are the most common metastatic site, other are skeleton, heart, and liver[18].Moreover, on histologic examination some fibroadenomas can look like phyllodes tumors[18, 19].The difficulty of distinguishing among fibroadenomas, benign phyllodes tumors, and malignant phyllodes tumors depended to experienced ofpathologist[19, 20]. However, the number of secondary sarcomas is increasing due to breast irradiation for previous breast carcinoma, as breast conservation in the surgical treatment necessitates adjuvant radiotherapy[4, 12]. A greater proportion of the radiation-induced sarcomas are angiosarcomas[3]. The proportion of angiosarcomas reported in the literature varies from 41%, 42% to 92%[2-4].

In general, breast sarcomas have a poorer prognosis than breast cancer. Five-year overall survival ranged between 44 and 67%[1, 3, 15] and 5-year sarcoma-specific survival ranged between 56.6 and 78%[1, 2, 4]. Tumor size (> 5 cm), secondary sarcoma (radiation-induced sarcoma, chronic lymphedema), residual tumor after treatment, cellular pleomorphism, and angiosarcoma were found to be prognostic factors for survival rate[3]. In these studies, although the mainstay of treatment should be radical surgical excision with negative margins, with radiotherapy may or not improved survival as other research[1-4, 15]. Our patient underwent wide radical resection with margin free and post radiotherapy.

4. Conclusion

For all patients with soft tissue sarcomas, an en bloc radical resection with negative margins is choice and potentially curative therapy. Post resection adjuvant radiotherapy can prevent local recurrence abd survival but, neoadjuvant chemotherapy for aggressive high-grade lesions is widely accepted as a therapeutic option and its benefits have been reported. We believe that breast sarcomas could be treated in the same way as other soft tissue sarcomas in terms of adjuvant radiotherapy. Because of the rarity of these lesions for decision to best approach will be difficult.

Compliance with ethical standards

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Authors' contributions

CM summarized the case study and drafted the manuscript. MS made decisions about patient treatment and reviewed the literature. RK collaborated in manuscript drafting. KO, YS, SS, SN, and MS performed the chemotherapy and operation. NF performed original histologic staining and provided high resolution images as well as descriptions of pathology. TF helped in managing the patient. HM, AKL, and JK revised the manuscript critically. NS aided in final review of the paper.

Availability of data and materials

The datasets used during the current study are available from the corresponding author on reasonable request.

Disclosure of conflict of interest

All authors read and approved the final manuscript and gave their consent for publication.

Statement of ethical approval

The present research work does not contain any studies performed on animals/humans subjects by any of the authors

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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