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RESEARCH ARTICLE

BREAST SARCOMA: ABOUT TWO CASES AND REVIEW OF THE LITERATURE

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Abstract

Breast sarcoma is a rare and aggressive form of cancer that is often difficult to diagnose and treat due to its rarity and lack of consensus on management. This case series presents two cases of breast sarcoma in women with no notable medical history, highlighting the importance of early detection and proper management in improving patient outcomes. The first case involves a 45-year-old woman who presented with a rapidly growing mass in her right breast, which was later diagnosed as a malignant phyllodes tumor. The second case involves a 26-year-old woman who underwent multiple surgeries for a recurrence of a high-grade phyllodes tumor in her right breast. The rarity of breast sarcoma makes it challenging to establish standardized treatment protocols, and further research is needed to determine the most effective treatment options. In this context, multidisciplinary collaboration and specialized centers for sarcoma management play a critical role in providing optimal care to patients with breast sarcoma.

Introduction:-

Breast sarcoma is a rare and aggressive form of cancer that is often difficult to diagnose and treat due to its rarity and lack of consensus on management.[1]. The rarity of breast sarcoma makes it challenging to establish standardized treatment protocols, and further research is needed to determine the most effective treatment options. In this context, multidisciplinary collaboration and specialized centers for sarcoma management play a critical role in providing optimal care to patients with breast sarcoma.[2].

Case Series

Case 1:

Mrs. M.F, a 45-year-old widow, G0P0, with no notable medical history, presented with an abscess in her right breast that was drained four years ago (without documentation). Over the past year, she has experienced a rapid increase in the size of her right breast. Clinical examination revealed a swelling that occupied the entire right breast measuring 35 cm in its greatest axis (fig. 1). A breast ultrasound showed an ACR 5 breast (fig. 2), and a TAP CT scan showed a huge mass in the right breast. Surgical biopsy revealed a borderline phyllodes tumor (intermediate grade) with no signs of malignancy. The patient underwent a right total mastectomy, and the pathological results indicated a malignant phyllodes tumor of the right breast (phyllodes sarcoma) measuring 27/22/20cm

Case 2:

Mrs. C.Y, a 26-year-old unmarried woman with no notable medical history, underwent her first operation in September 2014, which was a lumpectomy of the right breast. The pathological result revealed a benign stromal

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tumor with spindle-shaped cells. She underwent her second operation in November 2015 for a recurrence (no pathology report available), and her third operation in January 2016 for another recurrence, which was found to be a high-grade (grade 3) phyllodes tumor. Clinical examination revealed a hard, fixed, 8 cm mass in the right breast, adherent to the skin, classified as T4bNxM0. Breast ultrasound showed masses and nodules in the QSI of the right breast classified as ACR 4 (fig. 3). TAP CT scan showed no secondary locations. The patient underwent a total right mastectomy, and the pathological result showed complete excision of a mesenchymal sarcomatous process consistent with a high-grade phyllodes tumor. The surgical margins were clear, and the nipple and skin showed no signs of tumor infiltration. The patient was referred for additional radiotherapy.

Discussion:-

Breast sarcoma is rare but aggressive. Due to its rarity, a consensus on disease management has not been reached. In most cases, the etiology is unknown, although women who have received radiotherapy have an increased risk of developing breast sarcoma later on, the absolute incidence of which remains low.[3].

Clinical presentation includes a unilateral, well-defined, and large mass. It often grows more rapidly than epithelial breast carcinoma.[4] It can be suspected by physical examination or through imaging tools such as mammography or MRI. Rare spread occurs in this entity, and dissemination usually occurs in the lungs, bones, and liver.[5].

The use of radiotherapy seems to be recommended after positive margin resection due to the high risk of recurrence.[6].

Treatment includes initial surgery involving mastectomy and achieving a resection margin in a low-grade tumor,[7]. leading to the rejection of other adjuvant treatments due to the aforementioned controversy on these therapies. Completing surgical treatment with tumor margin enlargement and undergoing postoperative radiotherapy, although there is little evidence confirming the effectiveness of these two states in improving overall survival rates.

Breast sarcomas are classified according to the American Joint Committee on Cancer (AJCC) classification. [10].The staging criteria include tumor size with a cut-off of 5 cm, lymph node involvement, distant metastases, histologic grade, and surgical margins. The AJCC classification for breast carcinoma is not relevant for sarcomas due to the rarity of lymph node involvement. [9]PET scan has no established role in staging breast sarcomas.[8] The risk of local and metastatic recurrence, particularly pulmonary, is highest within the first 5 years. In a French series of 83 patients with primary breast sarcomas and a median follow-up of 8 years, the disease-free survival (DFS) rate was correlated with grade and tumor size. Histologic grade was the only factor significantly correlated with overall survival (OS) in this series. Breast sarcomas in irradiated fields have a worse prognosis, with tumor size and histologic grade being major prognostic factors for local control and survival.[10] Angiosarcomas have a poorer prognosis than other histologic types of breast sarcomas, especially in cases of visceral involvement and in older patients. The survival rate of patients with angiosarcomas at initial diagnosis is estimated at 12% at 5 years.[11]

To summarize, breast sarcomas are rare tumors with various histologies, either primary or secondary to radiotherapy. Angiosarcomas occur more often in irradiated tissue and have a particularly grim prognosis due to a high risk of metastasis and the absence of effective treatment. Prospective studies are rare, and the optimal treatment for these tumors is subject to debate. The therapeutic strategy should be discussed in a multidisciplinary meeting at a specialized center for sarcoma management in the context of the French network Netsarc (netsarc.org) [12].The reference treatment is wide surgery with clear margins, usually radical, without systematic lymph node dissection because axillary involvement is rare. High histological grade, surgical margin involvement, and a tumor size of more than 5 cm are factors of poor prognosis. The role of adjuvant chemotherapy and radiotherapy remains to be defined by specific collaborative studies in high-risk forms of relapse.

Conclusion:-

Breast sarcomas are rare tumors that present unique diagnostic and therapeutic challenges. Due to their rarity, there is no consensus on the optimal treatment approach, and treatment decisions should be made by a multidisciplinary team at a specialized sarcoma center. Surgery with clear margins is the reference treatment, and the use of adjuvant radiotherapy may be considered, particularly in high-risk cases. The prognosis of breast sarcomas is dependent on several factors, including histologic grade, surgical margin involvement, and tumor size, and the risk of local and

metastatic recurrence is highest within the first 5 years. More research is needed to determine the most effective treatment strategies for these rare tumors.

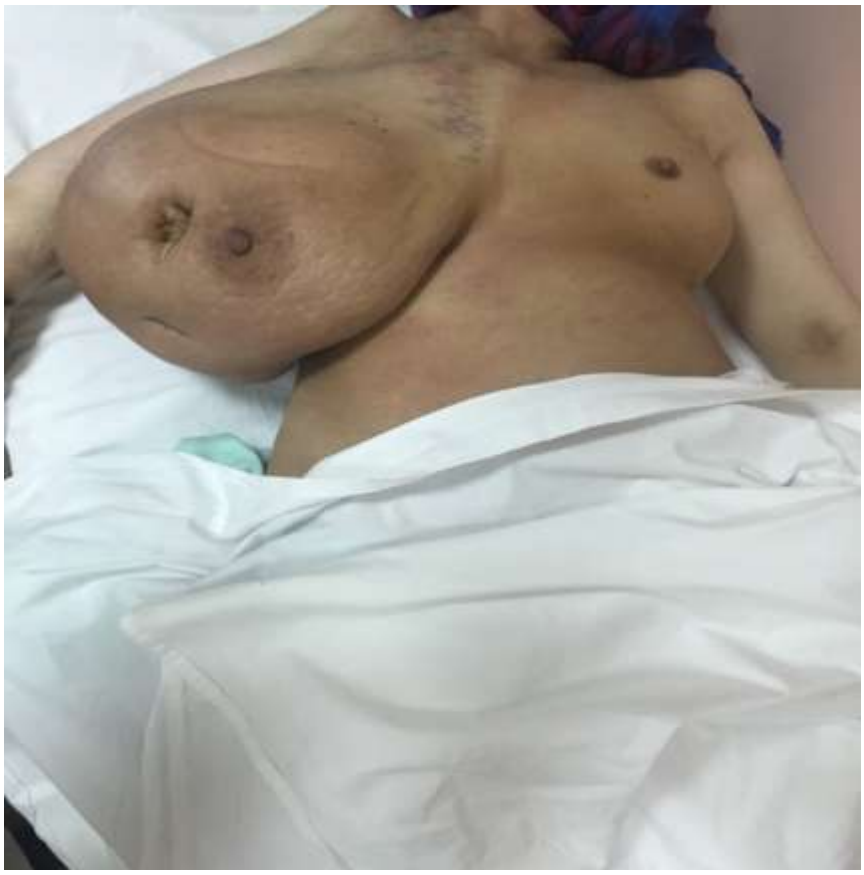

Fig. 1:- Image showing the tumor of the right breast.

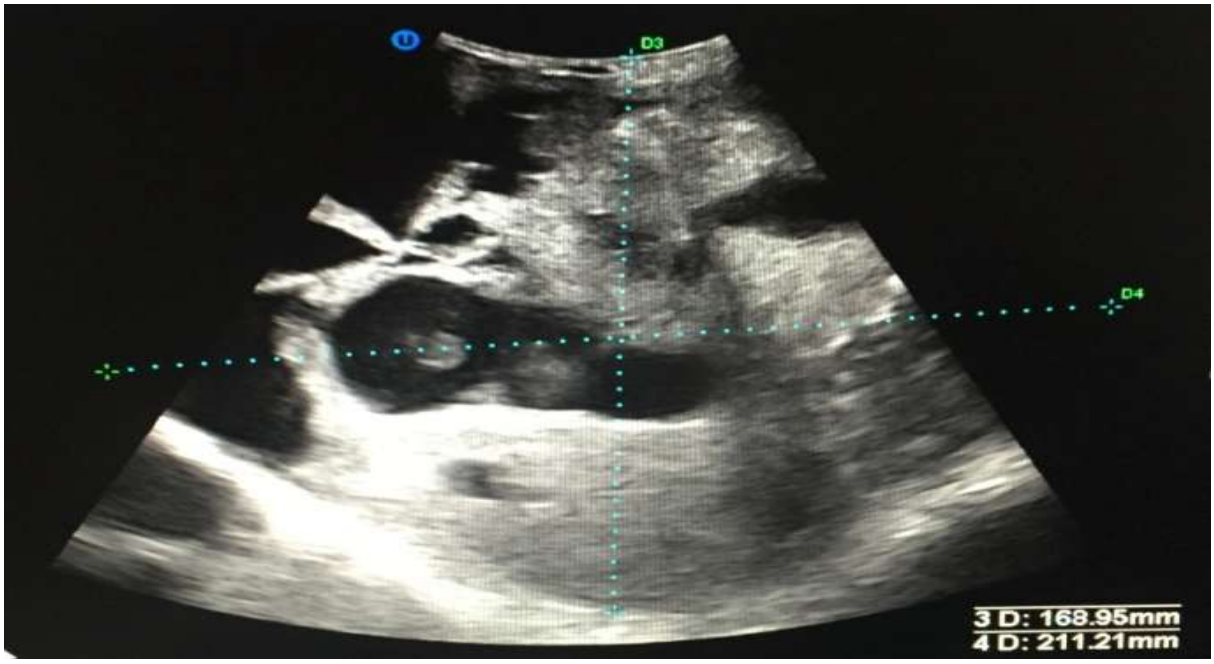

Fig. 2:- Image of breast lesion classified as ACR 5.

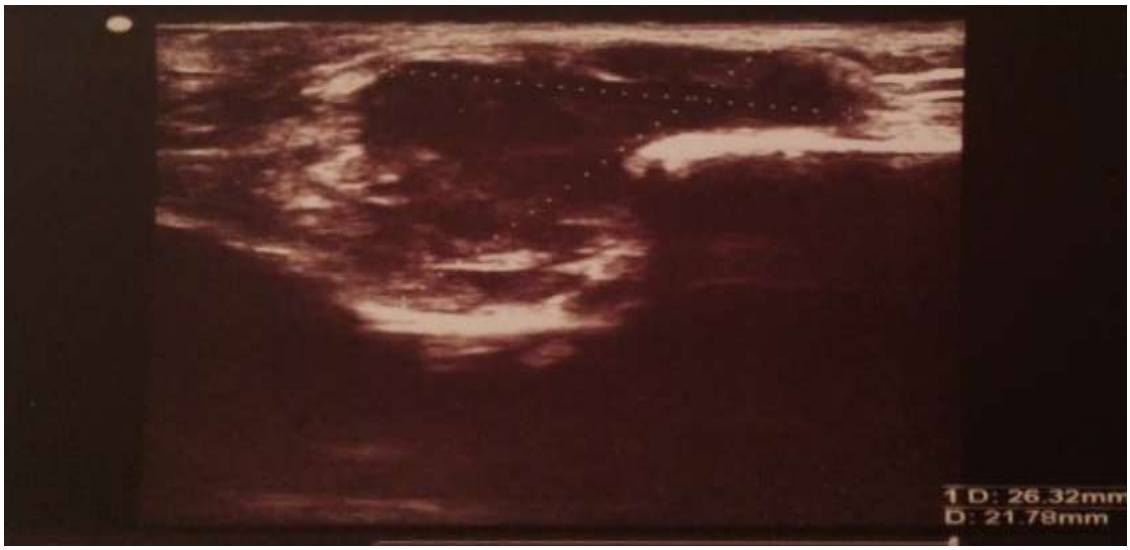

Fig. 3:- Ultrasound image of a QSI nodule.

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