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Case Report
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Ewing's Sarcoma Presenting as Breast Mass: A Rare Occurrence and Review of Literature

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Abstract

Introduction: Ewing's sarcoma typically involves the bony structures in adolescents and young adults. Extra skeletal sarcoma is a rare, aggressive, malignant soft tissue tumor with high recurrence rate. Ewing's sarcoma breast is a rare entity with less than 1% cases reported in literature.

Case Report: A fifty-five-year-old lady was referred to our institute with history of gradual onset painless breast lump for 1 year. Diagnostic ultrasound of bilateral breast identified a cystic lesion measuring 1.8 cm × 1.5 cm in lower inner quadrant of right breast. She underwent diagnostic excisional biopsy outside which showed malignant small round cell tumor. Post-excisional biopsy whole body 18-fluoro-deoxy-glucose Positron Emission Tomography showed no residual uptake with no evidence of metastatic disease elsewhere. Patient underwent a wide local excision of right breast and followed by adjuvant chemotherapy of 18 weekly VAC regimen.

Conclusion: Rarity of Primary breast Ewing's sarcoma poses a diagnostic dilemma and hence the treatment. The cornerstone of the treatment is local control with systemic therapy. Breast conservation surgery can be equivalent to mastectomy in local control in a good/average breast volume.

Introduction

Ewing's sarcoma /Peripheral Primitive Neuroectodermal Tumors (PNET) are rare, malignant small blue round cell tumor belonging to the Ewing's Family of Tumors (EFTs) characterized by the presence of translocation (11;22) (9q24; q12) and immunohistochemistry positivity for CD99 [1]. Ewing's sarcoma typically involves the bony structures in adolescents and young adults. Extra skeletal sarcoma is a rare, aggressive, malignant soft tissue tumor with high recurrence rate. Ewing's sarcoma Breast is a rare entity with less than 1% cases reported in literature [2]. To the best of our knowledge only 16 cases of extra skeletal Ewing's sarcoma has been reported in literature worldwide.

Case Presentation

A fifty-five-year-old lady was referred to our institute with history of gradual onset painless breast lump for 1 year. Diagnostic ultrasound of bilateral breast identified a cystic lesion measuring 1.8 cm × 1.5 cm in lower inner quadrant of right breast. She underwent diagnostic excisional biopsy outside which showed malignant small round cell tumor. Margins were not commented upon. Microscopic review was done which showed malignant round cell tumor with open chromatin with inconspicuous nucleoli. Mitosis was >10/10 hpf. Tumor cell showed focal positivity for CD99, Synaptophysin, S100, BCL2 FL1 and ki-67 (30% to 35%). Post-excisional biopsy whole body 18-fluoro-deoxy-glucose positron emission tomography showed no residual uptake with no evidence of metastatic disease elsewhere. Blocks review showed a malignant round cell tumor in sheet and nodules with immunopositivity for NKX2.2 and MIC2 and features consistent with Ewing Sarcoma. Case was discussed in MDT and planned for breast conservation surgery (Rewide local excision) followed by adjuvant therapy in view of unknown margin status. Patient underwent a wide local excision of right breast and followed by adjuvant chemotherapy of 18 weekly VAC regimen.

Discussion

Ewing's sarcoma or PNET belongs to a group of rare malignant neoplasm with blue round cell morphology commonly occurring in adolescent and younger age group. Primary PNETs have a predilection for the truncal and axial soft tissue, including the chest wall (Askin tumor), extremities and the paravertebral region [3]. More than 85% of Ewing's sarcoma is characterized

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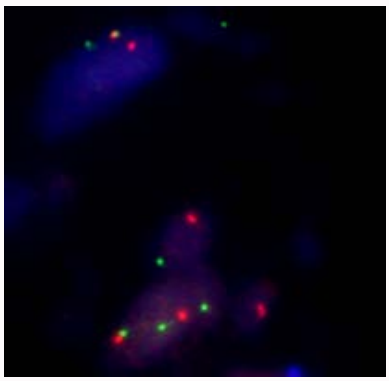

Figure 1: FISH performed using dual colour break-apart probes for EWSR1 gene showing break-apart signals indicating EWSR1 rearrangement.

by the presence of the typical translocation t (11; 22) (q24; q12) and immunophenotyping further establish the diagnosis of EWS/PNET which shows positivity for vimentin, Fli-1 and CD99 (Mic-2) [1]. Majority of patients with ES/PNET belongs to age group of 10 to 20 years. Several studies of adult EWS from Memorial Sloan Kettering, Royal Marsden have reported a median age of 24 to 27 years [4]. However, our patient presented in the unusual age of 55 years. Ewing sarcoma presenting as a primary breast lesion in older age group is very rare and poses a diagnostic dilemma. Differential diagnosis includes fibroadenoma, phyllodes tumor, carcinoma. Findings from ultra-sonography and mammography may vary. Similar cases in the literature have been reported and initially misdiagnosed as fibroadenoma, phyllodes tumor and mammary carcinoma. This diagnostic dilemma delays the treatment. In our patient a cystic mass with iso to hyperechoic content was initially identified on USG and was misdiagnosed as cystic lesion and therefore considered as benign. Excisional biopsy confirmed the diagnosis of small round cell tumor. Immunohistochemistry results further supplemented the diagnosis. This collective unusual presentation of site, age delays the diagnosis and hence the treatment. ES/PNET are aggressive tumor with high incidence of local recurrence and distant metastasis. Multi-modality treatment including local control with R0 resection followed by adjuvant systemic treatment is advised. Systemic chemotherapy improves the 5-year survival rate in localized PNET from 10% to 65% by preventing micrometastasis [5]. EWS/PNET of the breast is extremely rare. Sixteen cases of Ewing's sarcoma breast have been reported in literature to the best of our knowledge and had been managed by multimodality approach offering both breast conservation surgery and mastectomy. Out of 16 reported cases, three cases underwent mastectomy, followed by systemic chemotherapy in two cases. Seven cases underwent wide local excision/breast conservation surgery, followed by systemic chemotherapy in three

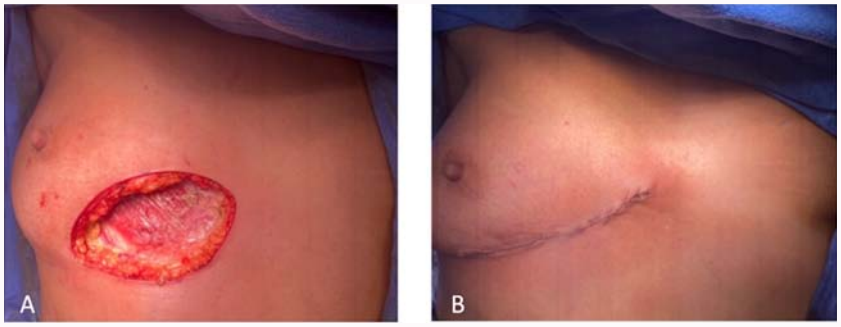

Figure 2 (A, B): Intra op images of breast conservation surgery.

Table 1: Summary of previous reported cases.

S.NO	CASE REPORT	AGE	PRESENTATION	SIZE	TREATMENT
1.	Tamura et al. [6]	47	LUMP	2.1 × 1.8	MASTECTOMY
2.	Maxwell et al. [7]	35	LUMP	1.8	LUMPECTOMY+ CHEMOTHERAPY
3.	da Silva et al. [8]	35	LUMP	12 × 7.5	CT+RT
4.	Ko et al. [9]	36	LUMP	2.5 × 2	LUMPECTOMY
5.	Vindal and Kakkar [10]	26	LUMP	3 × 2	WLE+CT
6.	Kwak et al. [3]	49	AXILLARY MASS		CT
7.	Dhingra et al. [11]	26	LUMP	3.5 × 3	WLE
8.	Suebwong et al. [4]	46	LUMP	4	CT+RT
9.	Majid et al. [12]	30	LUMP	7-RIGHT, 5-LEFT	CT
10.	Mahajan et al. [13]	50	LUMP	10 × 14	MASTECTOMY+CT
11.	Ranade et al. [14]	61	LUMP	6 × 6	CT+RT
12.	Meddeb et al. [15]	43	LUMP	3 cm	Breast Conservation Surgery + CT
13	Kim et al. [16]	35	Recurrent lump	2 × 2 cm	MRM + CT+ RT
14	Popli et al. [17]	14	lump	9.5 × 7 × 5	WLE
15	Srivastava et al. [18]	25	lump	11.6 cm × 9.2 cm × 6 cm	NACT+ WLE
16	Ikhwan et al. [18]	33	Metastatic		CTx

cases and NACT in one case. On follow-up, these patients remained disease free. Remaining other six cases received either chemotherapy alone or chemo and radiotherapy combined. In cases reported by Suebwong et al. [4] and da Silva et al. [8] treated by CTRT combined patients developed local and systemic relapse suggesting surgery remains the cornerstone for local control. So far in the literature no definite treatment of breast Ewing's sarcoma has been mentioned. Role of radiation therapy in the treatment of PNET is unclear, however it can combine with surgery to achieve local control. If a margin negative resection is achievable then breast conservation surgery (Wide local excision) is equivalent to mastectomy. Our reported case was managed with breast conservation surgery followed by adjuvant treatment. On follow-up patient remained disease free. Our case has shown that small tumors with good breast volume can be managed with breast conservation surgery and similar results can be achieved as mastectomy followed by adjuvant treatment.

Conclusion

Rarity of Primary breast Ewing's sarcoma poses a diagnostic dilemma and hence the treatment. The cornerstone of the treatment is local control with systemic therapy. Breast conservation surgery can be equivalent to mastectomy in local control in a good/average breast volume.

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