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Rhabdomyosarcoma of the breast – a rare malignancy

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

A **Suresh J. Bhosale**
E **Ashok Y. Kshirsagar**
F **Soniya R. Sulhyan**
B **Sanjitsingh R. Sulhyan**

Krishna Institute Of Medical Sciences and Research Center, Karad, India

Corresponding Author: Ashok Y. Kshirsagar, e-mail: drayk@indiatimes.com

Patient: Female, 60
Final Diagnosis: Rhabdomyosarcoma of the breast
Symptoms: Lump in axilla
Medication: —
Clinical Procedure: Mastectomy
Specialty: Oncology

Objective: Rare disease
Background: Primary nonepithelial malignancies of the breast include primary breast sarcomas, therapy-related breast sarcomas, the phyllodes tumors, and primary breast lymphomas. They account for less than 5% of all breast neoplasms.
Case Report: We report the case of a 60-year-old postmenopausal female diagnosed with rhabdomyosarcoma with infiltrating duct carcinoma. She was treated with modified radical mastectomy with axillary clearance and postoperative chemotherapy.
Conclusions: Primary rhabdomyosarcoma of the breast in adults is extremely rare. Rhabdomyosarcomas in adults account for less than 3% of all adult primary soft-tissue sarcomas.
Primary breast sarcomas usually present as large painless breast lumps with no associated skin and nipple changes or axillary lymphadenopathy; they are more aggressive and have more rapid growth than epithelial malignancies or benign breast lesions. The tumor can grow to large size, around 5.8 cm. Affected patients are typically women in their 50 s (ranging from 17 to 89 years), but it is also seen in men.
The treatment of primary breast sarcomas requires a multidisciplinary approach. Surgery remains the mainstay of therapy. Chemotherapy has no clearly defined role in primary breast or soft-tissue sarcomas.
The prognosis of primary breast sarcomas depends on the histologic grade and size of the tumor. They spread locally and hematogenously, but they are not usually associated with axillary lymphadenopathy.

Key words: rhabdomyosarcoma • breast • rhabdomyosarcoma breast

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Background

Primary nonepithelial malignancies of the breast include primary breast sarcomas, therapy-related breast sarcomas, the phyllodes tumors, and primary breast lymphomas. They account for less than 5% of all breast neoplasms [1].

Case Report

A 60-year-old postmenopausal female patient presented with a lump in her left breast that was first noticed 6 months ago and a lump in the left axilla first noticed 2 months ago. On examination, the left breast was larger than the right, with deviated nipple and peau d'orange present over the skin. A single 8×7 cm non-tender, firm, mobile lump was palpable in the left breast in upper and lower inner quadrants. The axillary mass was about 18×8 cm, and was hard and fixed. Systemic examination was within normal limits, with no clinical evidence of distant metastasis.

Hematological investigations, biochemistry profiles, and metastatic workup results were normal. FNAC of the lump was positive for malignant cells. The patient underwent modified radical mastectomy with axillary clearance (Figure 1). Histopathology found a rhabdomyosarcoma with infiltrating duct carcinoma. The tumor was positive for the immunohistochemical marker myogenin, thus confirming it to be rhabdomyosarcoma (Figure 2). The patient received 6 cycles of postoperative chemotherapy and recovered uneventfully. She is currently asymptomatic and free of any disease.

Discussion

Primary nonepithelial malignancies of the breast include primary breast sarcomas, therapy-related breast sarcomas, the phyllodes tumors, and primary breast lymphomas [1]. Pure nonepithelial primary malignant neoplasms of the female breast are rare, accounting for less than 1% of all breast malignancies [2]. Primary rhabdomyosarcoma of the breast in adults is extremely rare. Rhabdomyosarcomas in adults account for less than 3% of all adult primary soft-tissue sarcomas [3].

The clinical features of nonepithelial breast malignancies mimic those of breast neoplasms in many ways, but the prognosis and management of these types of tumors differ dramatically, thus it is important to distinguish between the two [1].

Primary breast sarcomas are malignant tumors arising from the connective tissue, accounting for less than 1% of all breast malignancies. The annual incidence of breast sarcoma is 44.8 new cases per 10 million women. The majority of breast sarcomas have no known cause. The established etiology for breast sarcomas is prior radiotherapy. Predisposing factors include:

Figure 1. Modified radical mastectomy gross specimen.

Figure 2. Rhabdomyosarcoma breast showing islands of myoepithelial cells with sarcomatous element with infiltrating duct carcinoma (Stain: Hematoxylin and Eosin [H and E], magnification: 10×).

genetic syndromes such as Li-Fraumeni and Gardner's syndromes, and neurofibromatosis type 1; environmental associations such as chemotherapeutic agents, arsenic compounds, vinyl chloride, and immunosuppressive agents; and human immunodeficiency virus and human herpes virus type 8 [1].

Primary breast sarcomas usually present as large painless breast lumps with no associated skin or nipple changes or axillary lymphadenopathy. They are more aggressive and have more rapid growth than epithelial malignancies or benign breast lesions. The tumor can grow to large size (up to 5.8 cm). Typically, the affected patient is a woman in her 50 s (age range 17 to 89 years), but it is also seen in men [1].

The treatment of primary breast sarcomas requires a multidisciplinary approach. Surgery remains the mainstay of therapy. Wide local excision with negative margins is adequate. If there is local recurrence, then salvage mastectomy should be done. Axillary dissection is not required, as primary breast sarcomas do not present usually with axillary lymphadenopathy; it is required in cases of palpable lymphadenopathy, carcinosarcoma or liposarcoma. Radiotherapy is controversial and does

not improve disease-free survival, but it may be useful in the treatment of high-grade lesions, larger tumors, and questionable or positive margins. Chemotherapy has no clearly defined role in primary breast or soft-tissue sarcomas [1].

Conclusions

The prognosis of primary breast sarcoma depends on the histologic grade and size of the tumor. Grading depends on

various pathologic features such as differentiation, mitoticity, necrosis, cellularity, and pleomorphism. Grades 1, 2, and 3 have 10-year overall survival rates of 82%, 62% and 36%, respectively. Depending on the size, the 10-year overall survival rates for tumors >5 cm, 5 to 10 cm, and >10 cm were 76%, 68%, and 28%, respectively. They spread locally and hematogenously, but they are not usually associated with axillary lymphadenopathy [1].

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