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Brief Notes on Pure Congenital Melanoma

Dr. Najet Wanger*

Department of Surgical Oncology, Regional Hospital of Jendouba, Jendouba, Tunisia, Faculty of Medicine of Tunis, Tunis Elmanar University, Tunis, Tunisia

Abstract

Background: In 2-6% of women, ectopic breast tissue is found. Ectopic mammary tissue can go through the same degenerative processes as a eutopic breast in terms of physiological alterations. Since it only accounts for 0.3% of all breast neoplasms, ectopic breast cancer is a rare disorder that typically arises in the axilla.

Case study: We describe a unique case of a 57-year-old Tunisian woman who had a left-sided axillary lump that had been developing for almost a month. Axillary dissection was used to remove the mass-containing axillary ectopic breast tissue. A medullary multifocal carcinoma was found on pathology, and metastases were found in two lymph nodes. After six rounds of chemotherapy, she had local radiation. She had hormonotherapy and herceptin therapy. No signs of local recurrence or distant metastases have been seen after a 2-year follow-up.

Conclusion: If an axillary lump is found in ectopic breast tissue, the first diagnosis that should be explored is ectopic breast carcinoma. There are no specific recommendations for diagnosis or treatment. In order to prevent therapeutic delays, doctors should be informed of this condition. Due to the uncertain natural history of this uncommon condition, diligent patient follow-up is crucial after diagnosis.

Keywords: Congenital melanoma; Breast; Tissue

Introduction

2-6% of the population has ectopic breast tissue (EBT). It could develop anywhere along the milk lines' thoracoabdominal segment, which anatomically extends [1-4] from the axilla to the inguinal region. The axilla, though, is the most typical presentation location. EBT is vulnerable to all physiological and pathological changes, including cancer, that take place in the normal breast. Only 0.3% of breast neoplasms are primary EBC, making them extremely uncommon. Medullary carcinomas, on the other hand, make up a relatively small part of these uncommon tumours. We wanted to explain this strange phenomenon. In light of this, we present the case of a 57-year-old Tunisian woman who presented with an axillary lump that was later determined to be an invasive medullary carcinoma originating in EBT by histopathology.

Present a case

A 57-year-old post-menopausal Tunisian lady with poor socioeconomic status who was non-smoking, multiparous, and G6P4A2 arrived with a painless left axillary lump that had been developing for about a month. Unremarkable personal medical and surgical history was present. She [2-4] stated that she had no personal or family history of breast, uterine, or ovarian cancer at the time of the clinical examination. A 50-mm, hard, well-defined lump was found during a physical examination in the left axilla. It stuck to the skin like glue. The breast exam revealed no obvious anomalies, and neither axillary nor supraclavicular nodes were seen. The remainder of the somatic examination revealed no more anomalies. Both regular blood tests and tumor markers (CA15-3) yielded results that were normal. A typical bilateral mammography was done and showed no abnormalities. A dedicated mediolateral oblique mammogram of the ipsilateral breast was taken next, followed by an ultrasound of the left axilla, which showed a substantial, suspicious, hyperechoic tumour measuring 4 cm and projecting into the skin.

Materials and Method

The axillary mass was extensively resected. A medullary multifocal carcinoma with free margins and partial subcutaneous proliferation, positive HER status (score: 3+), low progesterone receptor expression, negative oestrogen receptors, and a Ki67 score of 80% were shown by histopathology to support the diagnosis of EBC.

In order to exclude out occult breast metastases, enhanced magnetic resonance imaging (MRI) was recommended. This test revealed no concurrent lesions. The results (Figure 1) of a thoracoabdominopelvic computed tomography (CT) scan showed no secondary localisation. So, we dissected the ipsilateral lymph nodes. On histologic investigation, 2 of the axillary lymph nodes were found to be positive. Following

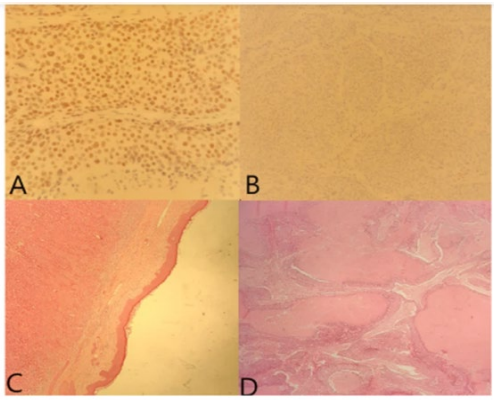

Figure 1: Low oestrogen labelling (*400); B no progesterone receptor labelling (*200); C subcutaneous carcinomatous growth (*40); D low oestrogen labelling comedonecrosis and carcinomatous tumours (*40).

***Corresponding author:** Dr. Najet Wanger, Department of Surgical Oncology, Regional Hospital of Jendouba, Jendouba, Tunisia, Faculty of Medicine of Tunis, Tunis Elmanar University, Tunis, Tunisia, E-mail: najet123@gmail.com

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locoregional radiotherapy, she underwent six rounds of intravenous, systemic, adjuvant chemotherapy based on 5-fluorouracil-epirubicin-cyclophosphamide (FEC) combined with Herceptin, as determined by a multidisciplinary committee. This treatment produced minimal side effects. After her chemotherapy was over, she continued taking tamoxifen for an additional five years as endocrine therapy. After a two-year follow-up, the patient is in good condition and there is no sign of a local recurrence or distant metastases.

Discussion and Results

We documented a sporadic instance of invasive medullary cancer that developed in EBT. In actuality, the prevalence of EBT in boys and girls varies from 1% to 3%. The most usual position is the axilla, as in our case, although other sites include the sternum, infraclavicular area, epigastrium, and vulva. EBT may be discovered in diverse places in up to one-third of patients. Similar to the breast tissue [5-9] in its anatomical position, the ectopic mammary tissue can undergo physiological changes related to menstrual cycle phases, pregnancy, and even the lactation stage. Similar to eutopic breast tissue, ectopic breast tissue goes through the same pathological processes. Ectopic breasts have been associated with fibroadenomas, fibrocystic changes, atypical ductal hyperplasia, phyllodes tumours, mastitis, and abscesses. Primary ectopic breast carcinoma (PEBC), which accounts for 0.3% of all breast malignancies despite being the most frequent cancer in women, is a rare occurrence. According to Evans et al. and research, 58% to 71% of all instances of PEBC are found in the axilla. The average duration to diagnosis is 40.5 months due to the low prevalence and misidentification of this illness. These lesions can be difficult to distinguish from benign or malignant axillary masses, such as skin tags, nevi, lipomas, and hidradenitis, and are commonly misdiagnosed. PEBC may manifest as an ulcerated lesion, as in our instance, or as ectopic breast tissue that looks normal. A subcutaneous tumour along the mammary line should signal the possibility of PEBC, and a histologic evaluation is required if there are any suspicious nodules present. It is usual practise to perform preoperative ultrasonography and mammography. In our case, we believed that performing an MRI was acceptable because, as recommended in the literature, it might be utilised to exclude a primary ipsilateral occult primary breast cancer or to facilitate surgical planning by determining the size and degree of involvement of the tumour. Histological evidence supports the PEBC diagnosis, and ductal carcinoma is identified as the typical subtype. However, more breast cancer subtypes have been discovered, including lobular, medullary, and papillary carcinomas. Marshall et al. reported the following distribution of histological types: 9.5% of lobular carcinomas, 79% of medullary carcinomas, and 9.5% of invasive ductal carcinomas. Medullary carcinoma is a rare and distinct subtype of breast cancer that accounts for less than 5% of all invasive breast malignancies, as was documented in our case. Orthotopic breast cancer paradigms should be used despite the lack of published medical literature on PEBC therapy or management recommendations due to the rarity and dearth of evidence.

Conclusion

If an axillary lump is found in ectopic breast tissue, the first diagnosis that should be explored is ectopic breast carcinoma. Once diagnosed, these patients should adhere to breast cancer staging and treatment recommendations. Patients with early-stage cancer may undergo radical excision, axillary lymphadenectomy, adjuvant radiation therapy along with hormone therapy, and/or chemotherapy, as needed. The uncertain natural history of this uncommon condition necessitates careful follow-up of these patients.

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Declaration of conflicting interest

No potential conflicts of interest were disclosed by the author(s) with regard to the research, writing, or publication of this paper.

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