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Case Report

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Accessory Breast Cancer: A Case Series

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Abstract

Accessory breast tissue (ABT) is ectopic breast tissue that occurs due to the failure of the embryonic mammary ridge to resolve during fetal breast development. This can occur anywhere along the milk line, with the axilla being the most common area of occurrence. Due to its pathophysiologic similarity with normal breast tissue, it is also susceptible to developing malignancies [1]. Primary accessory breast cancer accounts for 0.3-0.6% of all breast cancers [1]. The axillary area is the most commonly reported area of accessory breast carcinoma, accounting for 58% of reported ectopic cases [2]. Other reported areas include the parasternal line (18.5%), the subclavicular area (8.6%), the submammary region (8.6%), and the vulvar region (4%) [2]. Given the rarity of primary accessory axillary breast carcinoma, presentation and diagnosis can be challenging. Clinicians ought to be aware of this variance to ensure prompt management. Imaging plays an essential role in diagnosis. Treatment involves a multidisciplinary approach, including surgery, systemic therapy and radiation therapy as dictated by tumor biology and the stage of the disease. Here we report two cases of invasive ductal carcinomas arising in axillary accessory breast tissue.

Keywords: Accessory breast cancer; Axilla; Kajava classification; Invasive ductal carcinoma

Case Series

Case 1

A 49-year-old female presented for evaluation of a right axillary mass present for 1 month. On examination, a 2 cm mass was palpated in the right axilla, which correlated with mammographic and sonographic findings (Figure 1). A biopsy of the mass revealed a grade 2, ER/PR positive, HER2 negative invasive ductal carcinoma (IDC). The patient underwent ultrasound-guided, margin-negative resection of the right accessory breast with sentinel lymph node biopsy (SLNB). Final pathology confirmed a 1.5 cm IDC and 1 of 3 lymph nodes (LNs) positive for metastatic carcinoma (pT1cN1a). The patient was treated with adjuvant chemotherapy (docetaxel and cyclophosphamide) followed by tamoxifen and radiation with 5000 cGY (delivered in 25 fractions) to the right breast and right axilla covering the regional lymph nodes, including supraclavicular lymph nodes but not the internal mammary lymph nodes followed by boost to the right breast cavity in the right axilla of 100 cGy (delivered in 5 fractions) (Figure 2).

Case 2

A 47-year-old female presented for evaluation of a palpable and enlarging mass to her right axilla, which had been present for 12 months. On physical examination, the patient was found to have bilateral accessory axillary breast tissue type 4 on Kajava classification (Figure 3), along with a palpable 3 cm

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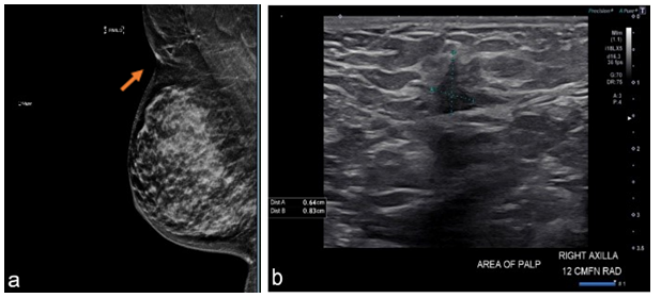

Figure 1 - Case 1: (a) MLO mammogram with presence of ectopic breast tissue in axilla (site of palpable abnormality is marked with a triangle; red arrow points to cancer). (b) Ultrasound of right axilla showing irregular hypoechoic mass measuring 0.6 x 0.8 x 0.5 cm.

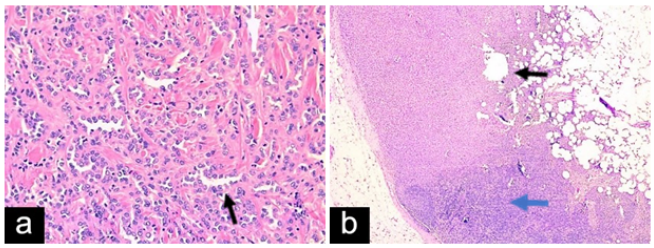

Figure 2 - Case 1: (a) Breast tissue with focus of invasive ductal carcinoma with lobular features, (H&E x 20). (b) Lymph node with metastatic carcinoma (H&E x 10). Tumor cells marked by black arrow. Blue arrow shows lymphoid cells.

Figure 3 - Case 2: (a) Clinical photograph of (a) bilateral accessory breast tissue in axillary region (b) right ectopic breast tissue with blue peritumoral dye injected (c) left normal ectopic breast.

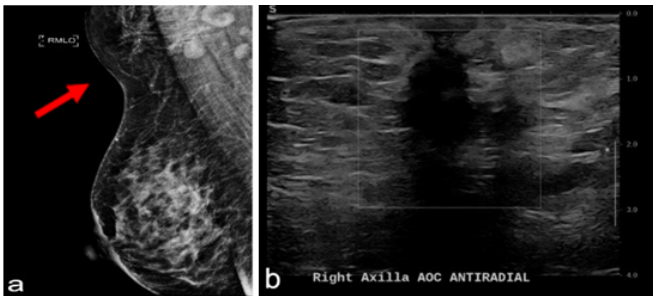

Figure 4 - Case 2 (a) MLO mammogram with arrow pointing to accessory breast tissue with spiculated mass. (b) Ultrasound of right axilla shows an ill-defined heterogeneous lesion measuring 3.4 x 1.2 x 1.4 cm.

mass in the right axilla with overlying skin thickening. Axillary lymph nodes were challenging to palpate due to the presence of accessory breast tissue. The patient's diagnostic workup included sonographic and mammographic evaluation, which revealed a suspicious right axillary mass (Figure 4), denoted as BI-RADS 4. Ultrasound-guided core needle biopsy of

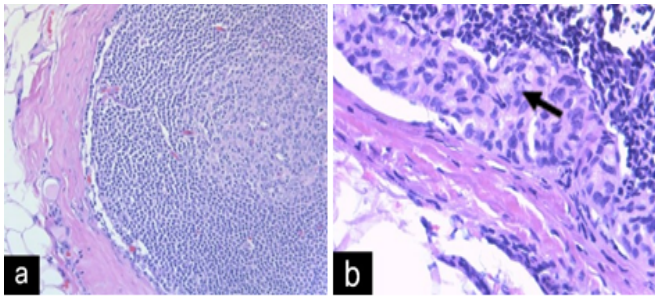

Figure 5 - Case 2: (a) Breast tissue with focus of invasive ductal carcinoma with lobular features (H&E x 20). (b) Lymph node with metastatic carcinoma (H&E x 40). Arrow marks tumor cells.

Table 1: Kajava's classification of supernumerary breast tissue

Kajava Classification	Supernumerary breast components
Class 1	Nipple, areola, and glandular tissue
Class 2	Glandular tissue and a nipple; no areola
Class 3	Areola and glandular tissue
Class 4	Glandular tissue only – Most common type
Class 5	Nipple and areola
Class 6	Nipple only
Class 7	Areola only
Class 8	Patch of hair

the mass revealed IDC with lobular features, grade 2, ER/PR positive, HER2 negative. The patient underwent margin-negative resection of the involved side with SLNB and excision of the contralateral axillary breast tissue. Periareolar injection of technetium Tc 99m tilmanocept and peritumoral injection of isosulfan blue dye were performed for sentinel lymph node identification. Two lymph nodes were identified as sentinel nodes, one with a high radioactive count but no blue dye and one blue node without a radioactive count. Histologic evaluation of the surgical specimen demonstrated a 2.5 cm IDC with lobular features on the right side and one of the two lymph nodes, the blue node, positive for metastatic carcinoma (pT2pN1a) (Figure 5). At the time of this writing, the patient was undergoing adjuvant systemic therapy (adriamycin and cyclophosphamide) and had plans to receive radiotherapy and endocrine treatment post-chemotherapy.

Discussion

During the fourth week of embryogenesis, breast tissue derived from ectoderm emerges from the mammary ridge. The ridges form bilaterally along the ventral surface of the body and run from the anterior axillary folds to the medial part of the inguinal folds. During normal development, all ridges, except those located in the pectoral area, regress. When the standard embryological regression fails, ectopic residual breast tissue develops under the hormonal influence at puberty.

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A classification system developed by Kajava in 1915 is used to describe the spectrum of presentations of ectopic breast tissue (EBT) [4]. Kajava's classification system differentiates supernumerary breast tissues into 8 different categories, as demonstrated in table 1 [1]. Both cases presented here fall into Class 4 of the Kajava system.

Accessory breast tissue is rare, with a reported incidence between 0.4-6% [2], and pathologies of the tissue are even rarer. Diagnosis of accessory breast tissue in the axillary region can be challenging due to the broad range of pathologies that can manifest in the axilla, including but not limited to lymphoma, lipoma, adenocarcinoma of sweat glands, or hidradenitis suppurativa [1,9]. On average, women presenting with ectopic breast cancer are younger, with a reported median age of 53 years [2]. According to the National Cancer Institute, the median age of women diagnosed with breast cancer is 63 years [10]. In the cases presented here, both women were premenopausal, were symptomatic at presentation and had pathologic lymph node involvement. Screening for axillary breast tissue is similar to normal breast tissue. However, due to its location, axillary accessory breast tissue is usually not fully visualized on a standard mammogram; therefore, other screening modalities must be used [1]. Ultrasound and MRI are useful tools when assessing ectopic breast tissue and can help differentiate ectopic breast tissues from other pathologies [1,5,7]. In case 2, the patient's axillary masses were initially thought to be axillary metastasis from pectoral breast cancer. MRI helped in identifying the axillary mass as the primary breast cancer.

Similar case series of accessory breast carcinoma (ABC) have reported lymph node involvement in as high as 46% of cases at the time of diagnosis [2,6]. This observed advanced stage at presentation could be attributed to a delay in diagnosis, propensity of malignant degeneration of ectopic breast tissue, and proximity of the axillary ABC to axillary lymph nodes resulting in higher rates of node-positive disease. The difficulty in assessing axillary lymph nodes clinically in the presence of axillary ABT and reported high rates of node-positive disease has led some surgeons to carry out axillary lymph node dissection (ALND) on a routine basis. In a study by Maki et al. accessory breast carcinoma was most commonly treated with radical resection and axillary lymph node dissection [11]. This results in overtreatment in many cases where lymph nodes were found to be negative [8].

There are no National Comprehensive Cancer Network (NCCN) guidelines for managing ABC. Literature on ABC shows the management of ABC to be similar to pectoral breast cancer using a multidisciplinary approach, including surgery, systemic therapy, and radiation depending on the staging and tumor biology [12].

Performing SNLB in a clinical node-negative ABC is somewhat controversial due to the inability to accurately palpate axillary lymph nodes in the presence of an axillary mass, making it difficult to discern from lymphadenopathy and the potential altered lymphatic drainage of aberrant tissue. Lymph node mapping may help to accurately identify sentinel lymph nodes and guide the surgeon in performing SNLB vs. ALND. No current guidelines from the American Society of Breast Surgeons or similar entities exist on whether a tracer should be injected periareolar or peritumorally in the axilla when localizing the sentinel node in the setting of ABC. SNLB success may be affected in cases where lymphatics have been disrupted by prior surgery in the axillary region [9,11]. In the cases presented here, tracers were injected peritumorally and periareolarly, blue dye was injected peritumorally, and a radioactive tracer was injected in periareolar space, which aided in localization and performing of SNLB successfully.

The role and extent of radiotherapy for ABC have yet to be clearly defined. It is unclear if radiation is needed after the complete removal of ABC and if the pectoral breasts should be included in the radiation field. Zhang et al. share the belief that lymph node metastasis is more likely to occur early in in ABC near the axillary region and therefore radiotherapy at the tumor site has been recommended with or without axillary lymph node metastasis to increase local control and reduce the risk of local recurrence [13,14]. In our first case, the patient received radiation to her right axillary region, which housed the primary tumor, the ipsilateral disease-free right breast, and the supraclavicular region. At the two-year follow-up, the patient had no evidence of locoregional recurrence. Radiotherapy to the ipsilateral disease free pectoral breast remains an area of controversy.

Conclusion

Patients with accessory breast tissue should be educated by their healthcare provider about the risk of cancer in the area and advised to report any abnormalities promptly. Physical examination of accessory breast tissue should be included in yearly breast examinations to rule out abnormalities. In the presence of accessory breast tissue, any suspicious masses in the axillary region identified by clinicians or reported by the patients should be subjected to a diagnostic workup akin to breast masses.

Awareness of this condition and the potential for malignancy should be on the clinician's radar when patients present for evaluation of "swelling" in areas prone to hosting accessory breast tissue. Diagnosing ABC should include detailed clinical history, physical examination, imaging, and biopsy with histopathological studies.

Conflicts of interest

We have no conflicts of interest to disclose.

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