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(CASE REPORT)

Adjuvant radiation therapy for adenoid cystic carcinoma of the breast: A case report and literature review

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Abstract

Adenoid cystic carcinoma (ACC) of the breast is an uncommon malignancy, accounting for only 0.1% to 1.0% of all breast cancers. Unlike more aggressive breast carcinomas, ACC is characterized by slow progression and a favorable prognosis. Early detection relies heavily on diagnostic mammography and breast ultrasound. Standard treatment includes either lumpectomy followed by radiation therapy or mastectomy alone. Although rare, cases of late recurrence and metastasis have been documented, underscoring the need for long-term follow-up. This review explores the existing literature and presents the case of a 79-year-old woman diagnosed with adenoid cystic carcinoma of the left breast, managed with lumpectomy and adjuvant radiation therapy.

Keywords: Breast cancer; Adenoid cystic carcinoma; Surgery; Radiation therapy

1. Introduction

Adenoid cystic carcinoma (ACC) of the breast is a rare subtype of breast cancer, comprising less than 1% of all breast malignancies [1]. It is classified as a variant of triple-negative breast cancer (TNBC), with an incidence of less than 0.1% annually [2]. While ACC predominantly originates in the salivary glands, cases have been reported in other anatomical sites, including the breast [2]. Compared to its salivary gland counterpart, breast ACC exhibits an indolent course and is associated with an excellent prognosis [3]. It primarily affects women aged 50 to 60, with only a few cases documented in men [2].

Histologically, ACC of the breast is characterized by a heterogeneous architecture, comprising solid, cribriform, and tubular-trabecular growth patterns in varying proportions [4]. Clinically, it often presents as a palpable mass, sometimes associated with pain due to perineural infiltration and myoepithelial cell contraction [4]. Despite being classified as TNBC, ACC differs from conventional TNBC due to its lower Ki-67 proliferation index [5] and the rarity of axillary lymph node involvement. Nevertheless, it remains associated with a risk of local recurrence and distant metastases, most commonly to the lungs [6].

Due to its rarity, ACC of the breast poses diagnostic challenges, making imaging crucial for early detection. Treatment typically involves either lumpectomy with adjuvant radiation therapy or mastectomy, with both surgical approaches yielding comparable survival rates [7]. The role of adjuvant systemic therapy remains uncertain and is considered on a case-by-case basis [7]. While case series and population-based studies have improved our understanding of this rare malignancy, further research is necessary to optimize treatment strategies and enhance patient outcomes [8].

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Our case is particularly noteworthy due to the exceptional rarity of breast ACC and the unusual bilateral presentation, making it a complex and unique clinical scenario.

2. Case Presentation

A 79-year-old multiparous woman with no personal or family history of breast cancer presented to our institution with bilateral breast malignancy.

She first noticed a self-palpated mass in her right breast three months prior to consultation, which progressively increased in size. On clinical examination, her performance status was 1. The right breast exhibited inflammatory changes with a large mass measuring approximately 10 cm, occupying almost the entire breast. No palpable nodules or inflammatory signs were detected in the left breast. Axillary and supraclavicular lymph node examination revealed a cluster of enlarged right axillary lymph nodes. The clinical tumor staging was cT4dN1M0, according to the 8th edition of the TNM classification (2017).

Mammography and ultrasound identified an ACR5 lesion in the right breast and an ACR3 lesion in the left breast, with a 1 cm intraparenchymal axillary lymph node.

Core needle biopsy of the right breast confirmed an infiltrating, poorly differentiated carcinoma, NOS, grade II SBR, with numerous lymphovascular emboli and an associated high-grade intraductal component. Immunohistochemistry (IHC) revealed a triple-negative profile (ER-, PR-, HER2- score 0) with a Ki-67 proliferation index of 10%.

A thoraco-abdominopelvic CT scan revealed a 6 mm pulmonary nodule in the left upper lobe and a right axillary lymphadenopathy measuring 20 × 15 mm, with no other distant metastases. A bone scan showed no evidence of secondary bone lesions. A biopsy of the left breast lesion (classified as ACR4b) revealed an intraductal proliferation with suspicious glandular structures, necessitating IHC confirmation.

The case was discussed in a multidisciplinary meeting, and neoadjuvant chemotherapy was recommended. The patient received dose-dense chemotherapy (four cycles of AC60 followed by four cycles of weekly paclitaxel), after which she underwent a right radical mastectomy with axillary lymph node dissection and lumpectomy of the left breast.

Histopathological examination of the right mastectomy specimen showed no residual invasive carcinoma. Lymph node dissection revealed two positive nodes out of ten, with evidence of therapeutic effect and subcapsular emboli (ypT0N1). The pathological response was classified as Chevallier Grade 1 and Sataloff TA/NC.

Examination of the left breast lumpectomy specimen revealed a solid papillary carcinoma with suspected invasion, prompting additional IHC studies. Subsequent immunohistochemical analysis confirmed adenoid cystic carcinoma with solid architecture, positive for P63, CK5/6, CK7, and CD117, and negative for hormone receptors and HER2. The Ki-67 index was 80%.

Postoperative radiation therapy was indicated. The patient received adjuvant radiation therapy to the whole left breast, the right chest wall, and the right supra- and infraclavicular lymph nodes, with a total dose of 42 Gy in 15 fractions (2.8 Gy per fraction).

At one-year follow-up, the patient remained in good general health (performance status 1). Clinical examination showed well-healed surgical scars with no signs of recurrence, no palpable nodules in the left breast, and no palpable lymphadenopathy. Mammographic surveillance revealed no evidence of disease recurrence, and no acute or late toxicities were observed.

3. Discussion

Breast adenoid cystic carcinoma (BACC) is an exceptionally rare subtype of breast cancer, accounting for less than 0.1% of all mammary malignancies [1]. It is a distinct salivary gland-like carcinoma characterized by intermixed gland-forming luminal cells and myoepithelial or basal cells, creating a pseudolumen filled with basement membrane material [9]. Initially described as 'Cylindroma' by Billroth in the 1850s, it was later renamed adenoid cystic carcinoma by Geschikter in 1945 [9].

BACC predominantly affects women in their 50s and 60s, although rare cases have been reported in men [7]. Clinically, it presents as a slow-growing, palpable mass, most commonly located in the upper outer quadrant or beneath the areola [10]. Breast pain occurs in approximately 14% of cases, likely due to perineural infiltration of tumor cells and myoepithelial cell contraction [4]. Bilateral involvement is exceedingly rare, and nipple discharge is uncommon [11].

Histologically, BACC exhibits cribriform, tubular-trabecular, and solid growth patterns, with the cribriform pattern being the most prevalent. The solid pattern—particularly its basaloid variant—is associated with higher malignancy potential [12]. Tumor grading is determined by the proportion of the solid component: Grade I has minimal solid areas, Grade II comprises less than 30% solid areas, and Grade III has over 30%, correlating with an increased risk of recurrence and metastasis [4].

Imaging findings for BACC are often nonspecific. On mammography, it may appear as an irregular lesion or a well-circumscribed, round mass, mimicking benign tumors. Ultrasound typically reveals a hypoechoic mass with angular or microlobulated margins and mild peripheral vascularity [13]. Contrast-enhanced MRI is useful for surgical planning, often demonstrating a well-circumscribed lesion with rapid enhancement but no washout [13].

Despite being classified as a triple-negative breast cancer (ER-, PR-, HER2-), BACC is associated with a favorable prognosis [14]. Lymph node involvement is rare, and distant metastases are uncommon. However, local recurrences and delayed distant metastases—predominantly to the lungs—have been reported up to 10 years post-diagnosis [6]. Zhang et al. documented a 5-year local recurrence rate of 14.3% in patients treated with surgery alone [8].

Surgical resection remains the cornerstone of treatment. BACC is often managed with lumpectomy followed by adjuvant radiation therapy or mastectomy, both of which yield comparable survival rates [7]. Axillary lymph node dissection is generally unnecessary due to the low incidence of nodal metastasis and is reserved for cases with clinically involved nodes [7]. In our patient's case, axillary lymph node dissection was performed on the right side, whereas no lymph node dissection was conducted on the left, in accordance with the low risk of nodal metastasis in BACC.

Adjuvant radiation therapy is commonly recommended in cases with nodal metastases or following breast-conserving surgery, as positive surgical margins increase the risk of local recurrence. A study by Sun et al. demonstrated that adjuvant radiation after lumpectomy improved the 5-year cancer-specific survival rate by 4.3% [15]. Similarly, another study indicated that postoperative radiation was associated with better survival outcomes compared to surgery alone without radiation [16].

The role of systemic chemotherapy remains uncertain, as survival rates do not appear to differ significantly between patients who receive chemotherapy and those who do not [17]. However, chemotherapy may be considered for high-grade tumors, lesions >3 cm, or cases with nodal involvement [14].

Given the rarity of BACC, no standardized treatment guidelines exist, and management is often individualized. Further studies are needed to optimize diagnostic and therapeutic strategies for this unique breast cancer subtype.

4. Conclusion

Adenoid cystic carcinoma of the breast, though rare, is associated with a favorable prognosis due to its low propensity for distant metastases and recurrence. Management primarily consists of local surgical resection, often complemented by radiation therapy, which plays a crucial role in minimizing the risk of local recurrence, particularly given its sometimes delayed onset. Although classified as a triple-negative histological subtype, this malignancy is distinguished by its favorable response to surgery, with mastectomy alone often being curative. Despite its rarity, early diagnosis is essential to achieving optimal outcomes. Long-term, rigorous follow-up, including regular mammography and ultrasound, is critical for the early detection of recurrence and ensuring optimal patient management.

Compliance with ethical standards

Disclosure of conflict of interest

All authors have no conflict of interest to declare.

Statement of ethical approval

This case report was conducted in accordance with ethical guidelines.

Statement of informed consent

The patient provided informed consent for the publication of this case report.

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