Case Report Open Access

Archives Of Breast Cancer

DOI: 10.32768/abc.1598047263-519

Adenoid Cystic Carcinoma of the Breast: Report of a rare Case and Review of the Literature

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ARTICLE INFO

Received:
25 June 2025
Revised:
11 November 2025
Accepted:
13 November 2025

Keywords:
carcinoma, adenoid cystic, triple negative, case report

ABSTRACT

Background: Breast cancer is the second most commonly diagnosed cancer and leading cause of cancer-related death among women in the United States. Adenoid Cystic Carcinoma (AdCC) of the breast is a rare subtype, comprising less than 0.1% of cases. Despite its triple-negative profile, it typically carries a favorable prognosis.

Case Report: A 55-year-old Hispanic female presented for routine screening mammography. Initial imaging revealed fibroglandular densities and architectural distortion in the right breast (BI-RADS 0). Further evaluation identified an irregular 0.9 × 0.9 × 1.0 cm mass at the 12:30 position and an 18 mm lesion at 12:00, 2.5 cm from the nipple (BI-RADS 4). Lumpectomy and sentinel lymph node biopsy confirmed AdCC (pT2 N0). Radiation therapy was recommended; systemic therapy was not indicated.

Conclusion: This case highlights the rare presentation and diagnostic features of breast AdCC, contributing to the limited literature on this uncommon malignancy.

INTRODUCTION

Adenoid cystic carcinoma (AdCC) of the breast is a rare malignancy, representing less than 0.1% of all breast cancer cases. [1,2] Although histologically similar to its salivary gland counterpart, breast AdCC exhibits distinct clinical behavior and generally follows an indolent course with favorable long-term outcomes. [3,4] Despite its classification under triple-negative breast cancers—lacking expression of estrogen receptor (ER), progesterone receptor (PR), and HER2—AdCC is biologically less aggressive than other triple-negative subtypes. [5,6]

Histologically, AdCC is characterized by a biphasic population of luminal epithelial and abluminal myoepithelial cells arranged in cribriform, tubular, or solid patterns. [7] Immunohistochemically, it typically shows negativity for ER, PR, and HER2, but is positive for myoepithelial markers such as p63, calponin, SMA, and CD117 (c-KIT), aiding in its distinction from histologic mimics. [8] Axillary lymph node involvement is uncommon, and the tumor usually exhibits low proliferative activity, often reflected by a low Ki-67 index. [9]

Given its rarity, evidence guiding optimal treatment is limited to retrospective studies and expert consensus. Surgical resection with negative margins, often followed by adjuvant radiation therapy, is the mainstay of treatment. Chemotherapy is generally not indicated due to the tumor's low-grade behavior and limited metastatic potential. [10]

We report the case of a 55-year-old woman diagnosed with conventional AdCC of the right breast. This case highlights the diagnostic challenges and clinical considerations involved in managing this rare entity and contributes to the growing body of literature on uncommon breast cancer subtypes.

CASE PRESENTATION

We present the case of a 55-year-old Hispanic female with a medical history of hypertension and

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pre-diabetes who was recently diagnosed with breast cancer. She had no personal history of breast malignancy, but reported a positive family history in her sister and maternal aunt. Her surgical history included a hysterectomy and unilateral oophorectomy.

The patient underwent routine screening mammography, which revealed scattered fibroglandular densities and an area of architectural distortion in the upper central quadrant of the right breast, initially categorized as BI-RADS 0. A subsequent diagnostic mammogram revealed persistent distortion at the 12:00 position, approximately 7 cm from the nipple, with an associated ill-defined area and an adjacent irregular mass measuring 0.9 × 0.9 × 1.0 cm at the 12:30 position. These findings were categorized as BI-RADS 4 (Figure 1), prompting further evaluation. An ultrasound-guided core needle biopsy was performed two weeks later (Figure 2).

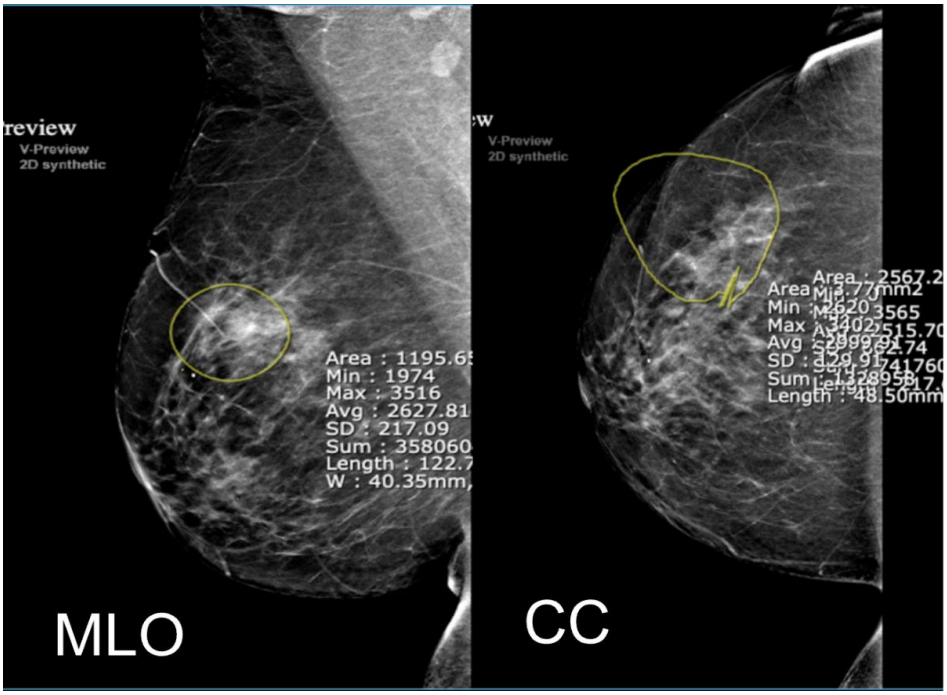

Figure 1. Diagnostic mammogram suggestive of BIRADS-4

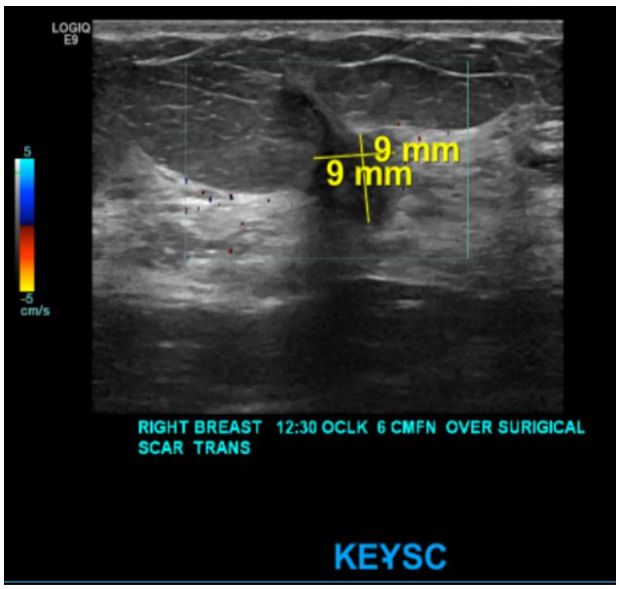

Figure 2. Breast ultrasound-guided core needle biopsy of the right 9mm x 9mm breast mass

Bilateral breast MRI identified an 18 mm enhancing mass in the superior aspect of the right breast at the 12:00 position, approximately 2.5 cm from the nipple, corresponding to the biopsy-proven malignancy (Figure 3). Additionally, multiple small nodules (2–4 mm) were seen scattered throughout the right breast parenchyma. These were radiologically benign-appearing and were not biopsied, but will be monitored through interval imaging. No axillary lymphadenopathy was present, and the left breast was unremarkable.

Histopathologic examination revealed conventional adenoid cystic carcinoma (C-AdCC), grade 1 (Figures 4–5). Hematoxylin and eosin-stained sections showed tumor nests with characteristic cribriform architecture, infiltrating the breast parenchyma. Pseudolumens contained eosinophilic basement membrane components, and the tumor cells exhibited uniform basophilic nuclei with no significant pleomorphism or mitotic activity. Immunohistochemical staining with p63 confirmed the presence of an abluminal myoepithelial

layeraround the cribriform structures (Figure 6), consistent with AdCC.

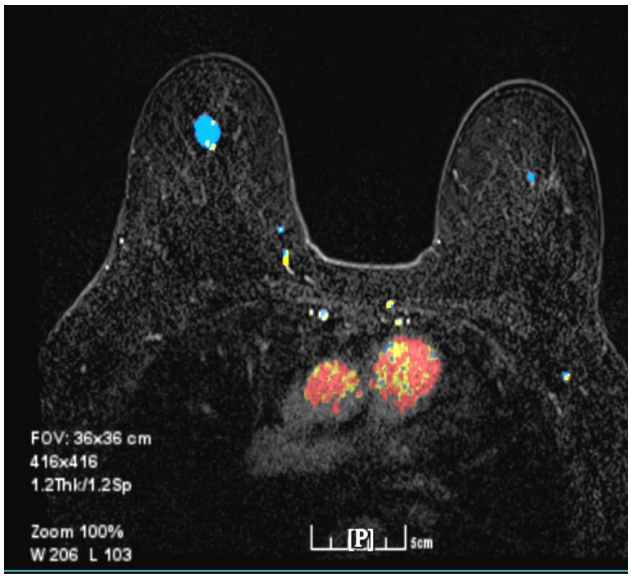

Figure 3. Bilateral breast MRI revealed an 18 mm mass in the superior aspect of the right breast at the 12:00 position, 2.5 cm from the nipple, corresponding to the known malignancy

Immunohistochemically, the tumor demonstrated a triple-negative phenotype—Estrogen Receptor (ER)–negative (0%), Progesterone Receptor (PR)–negative (0%), and HER2–negative (score 0). Tumor cells were diffusely positive for CD117 (c-KIT), a marker strongly associated with AdCC. The Ki-67 proliferation index was 25%, suggesting moderate proliferative activity.

Following multidisciplinary tumor board discussion, the patient underwent breast-conserving surgery, including lumpectomy with Savi Scout localization, clip placement, and sentinel lymph node biopsy. Final pathology revealed a 2.8 × 2.4 × 0.8 cm conventional adenoid cystic carcinoma, classified as triple-negative subtype and staged as pT2N0 according to the AJCC 8th edition. The tumor extended to within 0.28 mm of the inferior resection margin, with all other margins clear. No metastatic disease was identified in the sentinel lymph node.

Given the tumor's indolent histology, lack of lymphovascular invasion, and absence of nodal involvement, systemic chemotherapy was not recommended. The patient was referred for adjuvant whole-breast radiation therapy as part of her breast-conserving treatment plan.

At the time of this report, the patient is undergoing adjuvant radiation therapy and is scheduled for a follow-up breast MRI in six months to monitor the stability of the additional nodules. Continued surveillance will be coordinated through routine imaging and clinical examinations.

Table 1. Differentiating features between AdCC and DCIS.

Feature	Adenoid Cystic Carcinoma (AdCC)	Cribriform Ductal Carcinoma In Situ (DCIS)
Architecture	Cribriform, tubular, or solid with true luminal spaces ("punched-out" appearance)	Uniform cribriform spaces with rigid, cookie-cutter architecture
Cell Population	Biphasic: luminal epithelial and basaloid/myoepithelial cells	Monomorphic epithelial cells only
Nuclear Atypia	Mild; uniform round-to-angulated nuclei	Mild to moderate atypia; round nuclei with visible nucleoli
Mitoses	Rare	May be present depending on grade
Stroma	Often hyalinized or basophilic with basement membrane material	No prominent stromal reaction
Basement Membrane Material	Abundant; visible on H&E and PAS stains	Minimal or absent
CD117 (c-KIT)	Positive (highlights myoepithelial/basal cells)	Negative
p63	Positive (highlights myoepithelial/basal cells)	Myoepithelial layer surrounds ducts only
SMMHC, Calponin	Positive in basal/myoepithelial component	Positive only in peripheral myoepithelial cells
Estrogen Receptor (ER)	Typically negative	Usually positive
HER2	Negative	May be positive depending on DCIS subtype
Ki-67 Proliferation Index	Low to moderate (variable)	Variable, often low unless high-grade
Prognosis	Generally favorable; indolent behavior	Varies with grade; high-grade DCIS may progress to invasive carcinoma

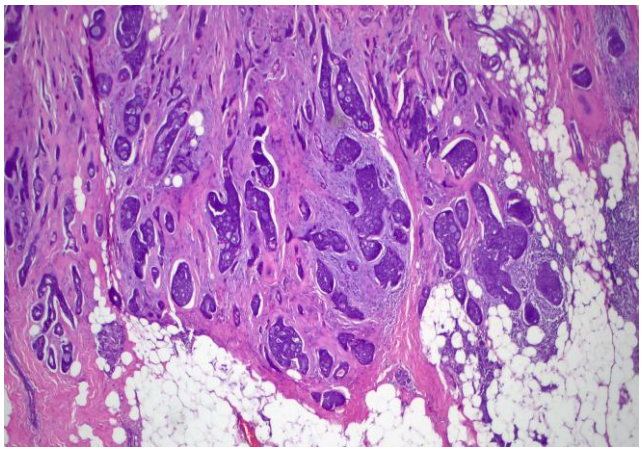

Figure 4. Low power immunohistology view shows multiple tumor nests infiltrating breast tissue

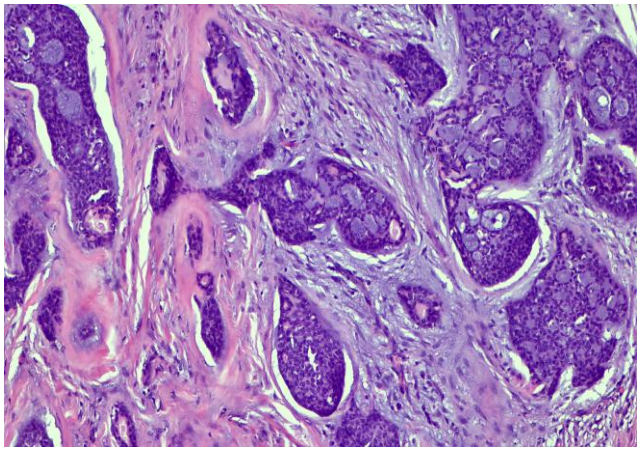

Figure 5. Medium-power view shows tumor with characteristic lumina filled with basophilic basement material and lined by small dark cells

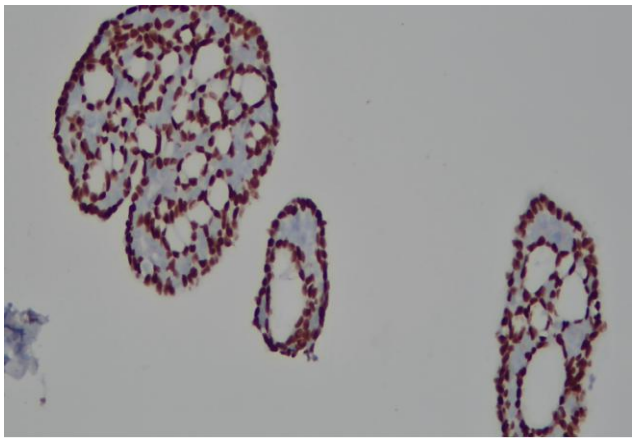

Figure 6. 5x image of P63 stain stain positive for myoepithelial cells and negative for epithelial cells

DISCUSSION

Adenoid cystic carcinoma (AdCC) of the breast is a rare malignancy, accounting for fewer than 0.1% of all breast cancers.[8] Owing to its rarity and often subtle imaging features, accurate diagnosis relies on integrating radiologic, histologic, and immunohistochemical findings. AdCC typically presents as a slow-growing, asymptomatic lesion, frequently detected during routine screening, as in our case. While MRI is valuable for lesion localization and surgical planning, its imaging characteristics—often a well-circumscribed, homogeneous hypoechoic mass—can mimic benign entities [9,10], making histopathological confirmation essential.

Histologically, AdCC closely resembles its salivary gland counterpart and is composed of biphasic cell populations: inner ductal epithelial cells and outer myoepithelial cells. This biphasic architecture can be highlighted by immunohistochemistry. The ductal component typically expresses CK7 and Cam 5.2, while the myoepithelial component shows positivity for markers such as SMA, S100, calponin, p40, p63, GFAP, and certain cytokeratins. These markers help distinguish AdCC from histologic mimics.

Three distinct architectural growth patterns are recognized: cribriform, tubular, and solid. [11]

Importantly, the 5th edition of the WHO Classification of Breast Tumors delineates three histologic subtypes of adenoid cystic carcinoma (AdCC) with distinct clinicopathological profiles:

1. Classic AdCC (C-AdCC): Characterized by cribriform and tubular growth patterns and low-grade cytology, this subtype follows an indolent clinical course.
2. Solid-Basaloid AdCC (SB-AdCC): This subtype exhibits solid nests, nuclear atypia, and necrosis, and is associated with a higher risk of recurrence and metastasis.
3. AdCC with High-Grade Transformation (AdCC-HGT): A newly recognized, aggressive variant defined by an abrupt transition from classic AdCC to an undifferentiated carcinoma, featuring loss of the characteristic biphasic morphology and increased mitotic activity.[12,13]

Our patient had C-AdCC, typified by a predominantly cribriform and tubular architecture with low-grade cytologic features, consistent with a favorable prognosis.

The cribriform architecture of AdCC may closely resemble ductal carcinoma in situ (DCIS), particularly the cribriform subtype. However, AdCC pseudolumens typically contain eosinophilic basement membrane components and are lined by small, uniform basophilic cells. In contrast, DCIS lumens are often empty and lined by larger cells with more pleomorphic nuclei. [14] Immunohistochemically, DCIS usually expresses ER and PR and lacks CD117 expression, whereas AdCC is classically ER/PR-negative and CD117-positive. [15–17] p63 staining further aids differentiation by highlighting the abluminal myoepithelial layer in AdCC. Refer to Table 1 for differentiating features.

Molecularly, AdCC of the breast often harbors the t(6;9)(q22–23;p23–24) translocation, resulting in a MYB-NFIB gene fusion. [19] Although this fusion was not tested in our case, it is considered a molecular hallmark of AdCC and may become a future therapeutic target. Elevated MYB protein expression on IHC further supports the diagnosis in many cases. Importantly, AdCC usually retains intact BRCA1 function and lacks the TP53 mutations typically seen in other triple-negative breast cancers. [18,19]

Despite its triple-negative status, breast AdCC paradoxically demonstrates an indolent clinical course and excellent prognosis—attributes uncommon among triple-negative breast cancers.[20] It generally displays low proliferative indices, with Ki-67 values often below 20%, though our patient had a slightly elevated index (25%), reflecting some variability in this marker's prognostic value in AdCC.[21] Additionally, lower p53 expression correlates with minimal nodal involvement and low metastatic potential, further supporting its favorable prognosis.

Management typically involves breast-conserving surgery with negative margins, followed by adjuvant radiation therapy. Systemic chemotherapy is usually unnecessary due to the tumor's low-grade nature and low risk of nodal or distant spread. [22] Reported 10-year survival rates for C-AdCC range from 90% to 100%.[6] Nonetheless, long-term surveillance is recommended, as late local recurrences—although rare—have been documented.

This case provides valuable insight into the diagnostic and therapeutic approach for conventional AdCC of the breast, emphasizing the importance of a multidisciplinary strategy. Although triple-negative by definition, breast AdCC differs substantially from more aggressive triple-negative subtypes, requiring tailored treatment and careful diagnostic consideration. Early recognition of its unique features—including biphasic histology, specific IHC profile, and relatively indolent behavior—ensures appropriate management and avoids overtreatment.

Informed consent was obtained from the patient for publication of this case.

CONCLUSION

This case highlights the importance of recognizing the unique imaging, histopathologic, and molecular features of adenoid cystic carcinoma (AdCC) of the breast—an uncommon subtype of triple-negative breast cancer (TNBC) with a distinctly indolent clinical course. Unlike typical TNBCs, which are often aggressive with poor prognoses, breast AdCC carries a favorable outlook, with low rates of lymph node involvement and distant metastasis.

Despite the reassuring behavior of AdCC, its diagnosis can be challenging. AdCC may mimic invasive ductal carcinoma radiographically and requires a high index of suspicion for accurate classification. Key diagnostic features—such as cribriform architecture, biphasic cell populations, CD117 positivity, and a triple-negative immunoprofile—should prompt thorough pathologic evaluation.

Awareness of these distinguishing characteristics is essential to avoid misdiagnosis and overtreatment. This case contributes to the limited literature on breast AdCC and underscores the value of integrating clinical, radiologic, histologic, and immunohistochemical data for accurate diagnosis and individualized management of rare breast cancer subtypes.

CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest. The authors do not have any financial relationship with the organization that sponsored the research.

ETHICAL CONSIDERATIONS

Informed consent was taken.

FUNDING

No funding was received for the preparation of this case report and the literature review.

DATA AVAILABILITY

No datasets were generated or analyzed during the current study.

ACKNOWLEDGMENT

None.

AI DISCLOSURE

The authors declare that no artificial intelligence–assisted technologies were used in the preparation, writing, or analysis of this case report.

AUTHOR CONTRIBUTIONS

All authors contributed significantly to the conception, drafting, and revision of this case report and literature review. [JS, NA, JV] contributed to the conception and design of the study and drafted the initial manuscript. [JS, NA, JV, NC, NM] performed the literature review and contributed to the critical revision of the manuscript. All authors approved the final version of the manuscript and agree to be accountable for all aspects of the work. Author contributions are classified according to the CRediT (Contributor Roles Taxonomy) as follows:

Conceptualization: [JS,NA,JV], Methodology: N/A, Investigation: [N/A], Resources: [N/A], Writing – Original Draft: [JS, NA], Writing – Review & Editing: [JS, NA, JV, NC, AA], Supervision: [NM, AA]

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How to Cite This Article

Sajdlowska J, Ahmadi N, Clarke N, Veltri J, Akmal A, Moulayes N. A Rare Case of Adenoid Cystic Carcinoma of the Breast: A Case Report and Review of the Literature. Arch Breast Cancer. 2025; 13(1):118-123.
Available from: https://www.archbreastcancer.com/index.php/abc/article/view/1153