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Case Report

# A rare case of recurrent adenoid cystic carcinoma of the breast with weak hormone receptor positivity ☆,☆☆

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ABSTRACT

Adenoid cystic carcinoma (ACC) of the breast is an exceptionally rare malignancy, accounting for less than 0.1% of all breast cancers. Despite its favorable prognosis, optimal management remains undefined due to its rarity and lack of consensus guidelines. We report a case of recurrent ACC of the breast, illustrating treatment challenges and the need for individualized management. A 64-year-old woman presented with a palpable mass in her left breast; imaging and biopsy confirmed ACC with very poor hormone receptor positivity. She started neoadjuvant chemotherapy but discontinued after 3 cycles due to severe neutropenia and lack of response. She then underwent breast-conserving surgery and radiotherapy. Three years later, she developed a local recurrence. Imaging and biopsy reconfirmed ACC and a subsequent total mastectomy achieved clear margins. Local recurrence of breast ACC can occur despite chemotherapy, surgery, and radiotherapy. Ineffectiveness of chemotherapy and recurrence after breast-conserving surgery suggest mastectomy might offer better local control. Hormone receptor positivity raises considerations for hormonal therapy, not standard for typically triple-negative ACC. The rarity of ACC complicates establishing standard protocols, necessitating personalized treatment plans based on tumor and patient factors. Recurrent breast ACC presents management challenges due to rarity and unpredictable behavior. More aggressive surgery and tailored treatment may improve outcomes. Further research is essential to develop evidence-based guidelines for managing this rare carcinoma.



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## Introduction

Adenoid cystic carcinoma (ACC) of the breast is an extremely rare malignancy, comprising less than 0.1% of all breast cancers [1]. It is more commonly associated with salivary glands and other exocrine tissues, making its occurrence in the breast unusual [2]. ACC typically affects women in their sixth to seventh decade of life and is characterized by a slow-growing, indolent course with a generally favorable prognosis [3]. Histologically, it presents unique features, often displaying a cribriform or tubular growth pattern [4]. While ACC of the breast is usually triple-negative—lacking estrogen receptor (ER), progesterone receptor (PR), and human epidermal growth factor receptor 2 (HER2) expression—cases with hormone receptor positivity have been documented, suggesting biological heterogeneity [5,6].

Due to its rarity, there are no established consensus guidelines for the optimal management of breast ACC. Treatment strategies are often extrapolated from protocols for more common types of breast cancer or ACC in other anatomical locations. Surgical excision with clear margins remains the cornerstone of treatment, but the decision between breast-conserving surgery and mastectomy is not well-defined and is typically made on a case-by-case basis [7]. The roles of chemotherapy and radiotherapy are also unclear, as ACC is generally considered less responsive to conventional chemotherapy regimens, and the benefits of radiotherapy are not conclusively proven.

Local recurrence can occur even after seemingly adequate treatment, and there is a lack of predictive factors to identify patients at higher risk. Because recurrence can occur, it's important to have long-term follow-up, which may also influence the initial surgical decision. The rarity of adenoid cystic carcinoma in the breast makes large-scale studies challenging, limiting the development of evidence-based guidelines.

## Case presentation

A 67-year-old woman presented to the breast clinic with a palpable mass in her left breast, which she had noticed 2 months prior. The mass was nontender, firm, and approximately 2 cm in diameter. She reported no nipple discharge, skin changes, or systemic symptoms such as weight loss or fatigue. Her medical history was significant for hepatitis C with progression to liver cirrhosis, and ulcerative colitis for which she underwent remote proctocolectomy with Barnett continent internal reservoir creation. There was no personal or family history of breast or ovarian cancer. On physical examination, a firm, mobile mass was palpable in the upper outer quadrant of the left breast. No axillary lymphadenopathy was noted. The overlying skin appeared normal, and there were no signs of nipple retraction or discharge. The right breast and contralateral axilla were unremarkable.

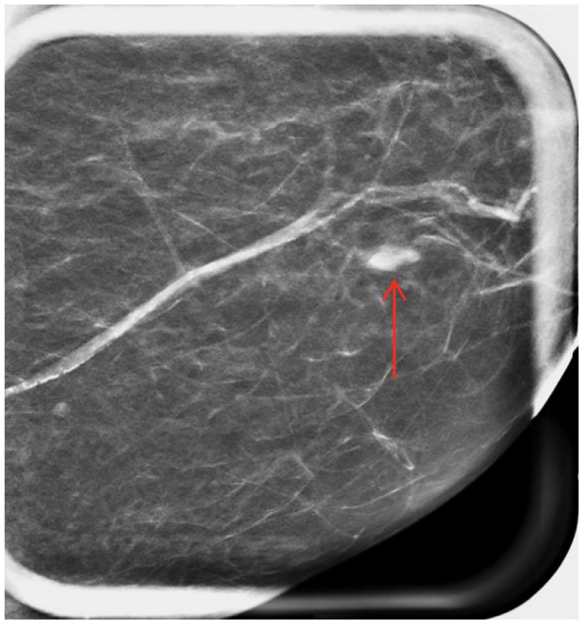

**Fig. 1 – Diagnostic left breast mammogram (cranial caudal view) with well-circumscribed, oval mass with partially obscured margins in the left breast, without microcalcifications (red arrow).**

### *Imaging studies*

A diagnostic mammogram revealed a well-circumscribed, oval mass with partially obscured margins in the left breast, without microcalcifications (Fig. 1). Breast ultrasound confirmed a 2.1 cm hypoechoic, solid lesion with microlobulated borders and minimal internal vascularity on Doppler examination (Fig. 2) at the 10 o'clock position 4 cm from the nipple. No suspicious axillary lymph nodes were identified.

An ultrasound-guided core needle biopsy of the left breast mass was performed, and post biopsy marker placed (Fig. 3). Histopathological examination revealed features consistent with adenoid cystic carcinoma (ACC) of the breast which has identical histomorphology to tumors seen in the salivary glands and includes basaloid cells arranged in tubules or with a solid and cribriform pattern. One of the characteristic findings of ACC is the presence of extracellular matrix. The tumor exhibited predominantly cribriform and solid patterns with extracellular matrix deposition (Fig. 4). Immunohistochemical staining showed that the tumor cells were positive for cytokeratin (CK) 7, CK5/6, and c-KIT (CD117). Notably, the tumor cells were weakly positive for estrogen receptor (ER) and progesterone receptor (PR), and negative for human epidermal growth factor receptor 2 (HER2) with a high Ki-67 index (97%).

### *Initial treatment*

Given the very poor hormone receptor positivity and to assess chemosensitivity, the patient was started on neoadju-

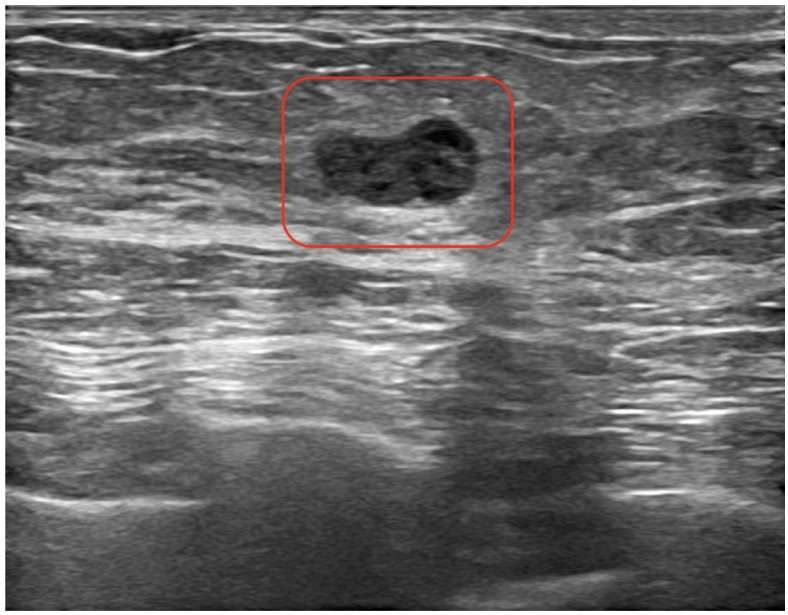

**Fig. 2 – Diagnostic left breast ultrasound demonstrating a microlobulated, hypoechoic, and solid lesion (red square).**

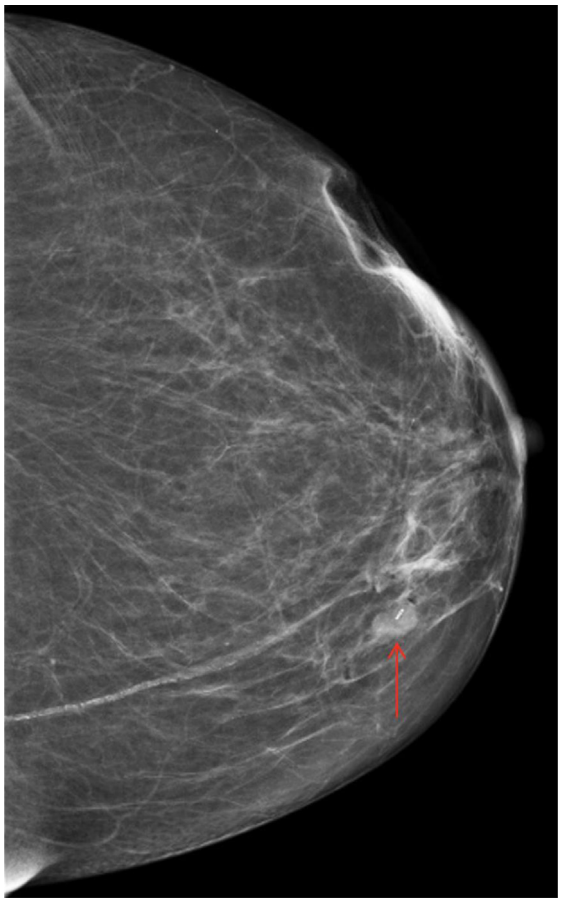

**Fig. 3 – Postbiopsy left breast mammogram (cranial caudal view) showing a lobulated nodular opacity with U-clip marker in place (red arrow). The nodule has well-defined margins without calcifications or obvious distortions.**

vant chemotherapy consisting of doxorubicin and cyclophosphamide. After three cycles, she developed grade 4 neutropenia and required hospitalization for febrile neutropenia. A follow-up assessment showed no significant reduction in tumor size. Due to the severe adverse effects and lack of response, chemotherapy was discontinued. The multidisciplinary team decided to proceed with breast-conserving surgery. The patient underwent a wide local excision with sentinel lymph node biopsy.

### *Adjuvant therapy*

Postoperative histopathology confirmed the diagnosis of ACC. Tumor size was 1.1 cm; margins were clear with the closest margin 6 mm from the tumor; no lymphovascular invasion was noted and four sentinel lymph nodes were negative for tumor. Adjuvant radiotherapy was recommended as is common for breast malignancies managed with breast conservation surgery. The patient received external beam radiotherapy to the left breast, totaling 5256 cGy in 20 fractions over 5 weeks. Hormonal therapy was considered due to the weak hormone receptor positivity. However, after discussing the uncertain benefit and potential side effects, the patient opted against hormonal therapy.

### *Follow-up and recurrence*

The patient was monitored every 6 months with clinical examinations and annual imaging studies. Three years after the initial treatment, she reported a new lump at the previous surgical site. Physical examination revealed a 1.5 cm firm mass in the left breast. Mammography and ultrasound confirmed a recurrent lesion at the site of the original tumor (Fig. 5), without evidence of regional lymphadenopathy or distant metas-

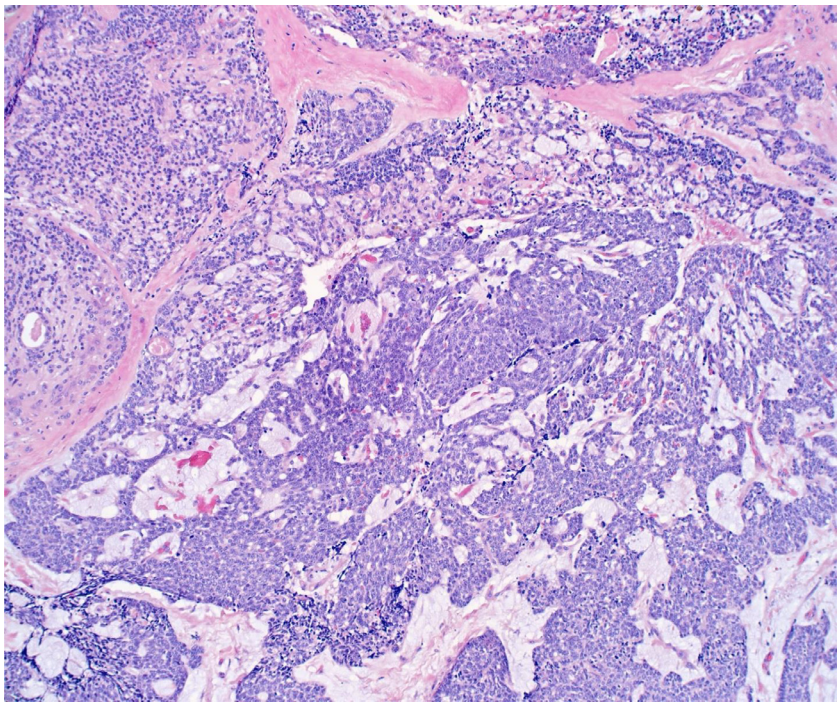

**Fig. 4 – H&E stain histopathology slide at 400x magnification demonstrating predominantly cribriform and solid patterns with extracellular matrix deposition which can be seen dissecting into the stroma surrounding the tumor.**

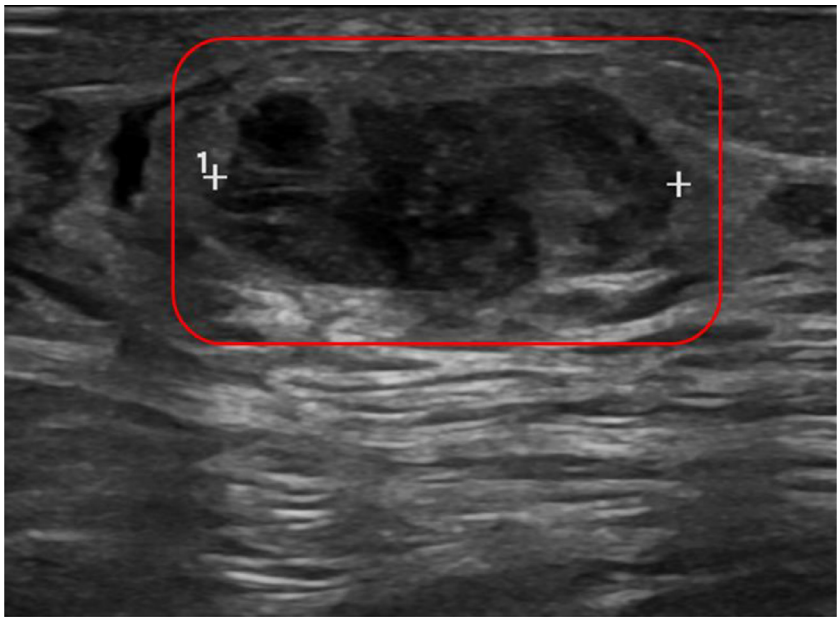

**Fig. 5 – Left breast ultrasound demonstrating a lobulated, wider-than-tall, hypoechoic mass reflecting a recurrent lesion (red box).**

tasis on computed tomography of the chest, abdomen, and pelvis.

An ultrasound-guided core needle biopsy of the new lesion was performed (Fig. 6). Histopathology confirmed recurrent ACC with similar morphological features to the initial tumor. Immunohistochemical staining showed negativity for ER, PR, and HER2 with high Ki-67 index (63%).

*Management of recurrence*

Considering the recurrence after breast-conserving surgery and radiotherapy, a total mastectomy was advised. The patient underwent a left simple mastectomy with sentinel lymph node biopsy without reconstruction. Histopathological examination of the mastectomy specimen confirmed ACC.

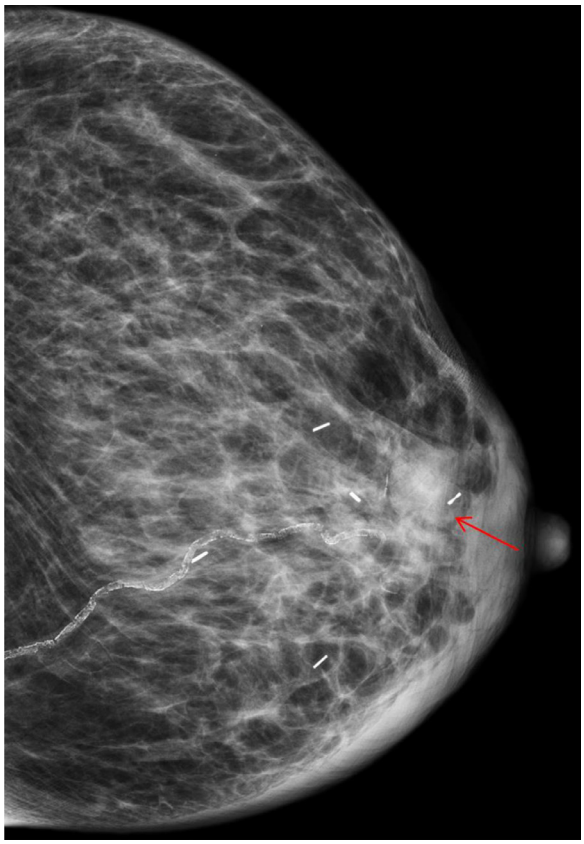

**Fig. 6 – Postbiopsy left breast mammogram (cranial caudal view) after reoccurrence demonstrating the recurrent lesion with hourglass clip just anterior to surgical clips marking the prior lumpectomy cavity (red arrow).**

The tumor was 2.0 cm in size with margins well clear at 2 cm. One sentinel lymph node was identified wans was negative for metastatic disease.

### *Outcome and follow-up*

After surgery, the patient had prolonged drainage from the mastectomy site, and after drains were removed a recurrent seroma required replacement of a drain. Delayed resolution was felt to be due to prior radiation therapy. No adjuvant endocrine therapy, chemotherapy, or addition radiation is planned. Surveillance will be with clinical examination as no specific breast imaging is required after total mastectomy.

### *Patient perspective*

Throughout her treatment, the patient was actively involved in decision-making. She expressed a preference for less aggressive treatments initially but agreed to more extensive surgery upon recurrence. She emphasized the importance of quality of life in her choices and was satisfied with the care received and the outcomes thus far.

## Discussion

Adenoid cystic carcinoma (ACC) of the breast is an exceedingly rare malignancy, constituting less than 0.1% of all breast carcinomas [1]. It predominantly affects women in their sixth to seventh decades of life [3]. ACC is characterized by a slow-growing nature and a generally favorable prognosis, but local recurrence can occur even after initial treatment [3]. The rarity of this tumor presents challenges in establishing standardized management protocols.

The optimal surgical approach for breast ACC remains a subject of debate due to the lack of large-scale studies. Breast-conserving surgery has been performed with satisfactory outcomes in some cases, but there is evidence suggesting that mastectomy may offer better local control [8]. A review of cases indicates that local recurrence rates are lower following mastectomy compared to breast-conserving surgery. For instance, studies have reported recurrence rates of up to 20% with breast-conserving surgery, whereas mastectomy has demonstrated recurrence rates below 5%. Achieving clear surgical margins is crucial, as positive margins are associated with higher recurrence risk [9].

The role of adjuvant radiotherapy in breast ACC is not well-defined. Some reports suggest that radiotherapy may reduce local recurrence rates, especially after breast-conserving surgery [10]. However, the indolent nature of ACC and its low mitotic activity raise questions about the radio sensitivity of this tumor [10]. In the absence of definitive evidence, the decision to use radiotherapy should be individualized, considering factors such as tumor size, surgical margins, and patient preferences.

Chemotherapy is generally considered ineffective for ACC of the breast. The tumor's low proliferative index and distinct histological features contribute to its resistance to conventional chemotherapeutic agents [7,11]. Neoadjuvant chemotherapy has shown limited success in shrinking tumors or improving surgical outcomes as evidenced by the current case report, despite presentation with a high proliferative index tumor unique for breast ACC [7,11]. As a result, chemotherapy is not routinely recommended for ACC, except perhaps in cases with advanced or metastatic disease where treatment options are limited.

Hormone receptor status in ACC of the breast is typically negative, but cases with hormone receptor positivity have been documented [7]. The presence of estrogen and progesterone receptors raises the possibility of hormonal therapy as a treatment option. However, due to the scarcity of data, the efficacy of hormonal therapy in ACC remains uncertain. Further research is needed to determine whether hormone receptor positivity translates into a meaningful therapeutic target in this context.

The rarity of breast ACC necessitates an individualized approach to treatment. Multidisciplinary teams should consider patient-specific factors such as tumor size, location, hormone receptor status, and patient comorbidities when devising a management plan. Long-term follow-up is essential due to the potential for late recurrence. Regular clinical examinations and imaging studies are recommended to monitor for signs of recurrence.

## Conclusion

Adenoid cystic carcinoma of the breast is an extremely rare malignancy lacking standardized treatment guidelines. Local recurrence can occur despite initial breast-conserving surgery and adjuvant radiotherapy, suggesting that mastectomy may offer better local control. The ineffectiveness of chemotherapy observed aligns with evidence indicating limited responsiveness of ACC to conventional chemotherapeutic agents. Hormone receptor positivity, though uncommon in ACC, was present and may warrant consideration of hormonal therapy in similar cases. Individualized treatment plans based on tumor characteristics and patient factors, along with long-term surveillance, are essential in managing this rare carcinoma.

### *Ethical considerations*

Informed consent was obtained from the patient for all treatments and procedures. Consent was also obtained for the publication of this case report, and all efforts were made to maintain patient confidentiality.

## Patient consent

We confirm that we have obtained written, informed consent from the patient for the publication of this case report. The patient has been thoroughly informed about the details that will be published and understands the implications of the publication. The written consent is stored securely and is available for review by the editorial team upon request.

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