Adenoid Cystic Carcinoma of the Breast: A Case Report

Abstract

Adenoid cystic carcinoma (ACC) is a basal-like subtype of mammary cribriform, solid, tubular, or trabecular architectural patterns. Epithelial and myoepithelial components make up the distinctive histologic pattern of ACC of the breast, which is a well-known tumor with the same nomenclature that originates in the salivary gland. In this report, we describe a rare case of ACC in the breast cribriform, trabecular growth pattern. Here we present a rare case of breast adenoid cystic carcinoma with a cribriform, trabecular growth pattern. Imaging studies are nonetheless extremely useful in ACC cases for screening and management planning but might be non-specific due to their variable imaging characteristics. The standard reference is pathological examination. Even though the pathological classification of ACC is still controversial, it is critically important in the decision of treatment options. However, the primary form of therapy for breast ACC patients is currently understood to be surgery. Because this pathological type is so uncommon, there is no clear direction in the selection of specific surgical treatments for this condition, which leads to variations in therapy.

Keywords: Adenoid Cystic Carcinoma, Breast, Pathological, Tumor, Screening

Introduction

ACC is a basal-like subtype of mammary cribriform, solid, tubular, or trabecular architectural patterns.1, 2 Epithelial and myoepithelial components make up the distinctive histologic pattern of ACC of the breast, which is similar to a well-known tumor with the same nomenclature that originates in the salivary gland.3, 4 Cribriform neoplasms such as carcinoma of the lung, prostate, stomach, and colon have been reclassified as more aggressive entities than previously thought.5, 6 Therefore, pathologists and oncologists may be affected in terms of practical prognostication by the discovery of a cribriform pattern.7, 8 Less than 4% of all breast cancers are invasive cribriform carcinomas, an uncommon histological subtype of invasive breast cancer.9, 10 Here we present a rare case of breast adenoid cystic carcinoma with a cribriform, trabecular growth pattern.11, 12

Case description

A 48-year-old premenopausal female, (gravida 5, para 5), medically free, presented to our hospital in 2016 complaining of right breast pain. There is no family history of breast cancer or any tumor or malignancy. On examination, no palpable masses or axillary lymph nodes, and no skin changes. Mild serous discharge with squeezing is noted.

Figure 1. Ultrasound showed multiple cystic lesions within the right breast, the largest one measuring about (1.5×1 cm), with thin walls and well-defined outlining, some of them showing fluid debris level, with no mural nodules inside.

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Annual follow-up was recommended.

The patient missed follow-up until January 2023, she came with the same complaint. Examination found a tender lump (2x1 cm) in the upper outer quadrant of the right breast.

On diagnostic mammogram and ultrasound was a BI-RADS-4 as shown in Figures 2 and 3 and recommendations were given for tissue diagnosis with ultrasound-guided core biopsy.

Ultrasound showed an Irregular solid hypoechoic mass with lobulated margins seen at the 3 o’clock position, 1 cm from the nipple. Another lobulated large cystic lesion with multiple internal echoes is seen at the position of 5-6 o’clock (Figure 3).

Similarly, showing right axillary lymph nodes with preserved fatty hilum as shown in Figure 4.

The patient underwent a tru-cut biopsy under aseptic measures and local anesthesia was applied and done from the solid mass lesion seen at 3 o’clock.
Histopathology examination shows predominantly cribriform, trabecular growth pattern with few formed spaces containing pink/mucoid secretions. The tumor has round to oval nuclei, clump chromatin, inconspicuous to occasional prominent nucleoli, and indistinct cytoplasm. In between, a few angulated tubular glands are also noted. Rare mitotic figures are seen. The tumor had weak estrogen receptor (ER) positivity, negativity for progesterone receptor (PR), and human epidermal receptor 2 (HER2) as well. The proliferation marker measured with Ki67 is 15–20%.

The patient underwent a right mastectomy with excision of axillary and sentinel lymph nodes. Fresh frozen for sentinel lymph nodes which were proven to be free of malignancy.

**Results and Discussion**

Less than 0.1% of all primary carcinomas of the breast are ACC, a rare subtype of invasive breast carcinoma.[13] As with other invasive breast cancer cases, the reported age distribution for patients with ACC of the breast varies from 38 to 81 years (with a median age of 60 years).[13] The majority of instances are in women, however, there have been a few cases documented in men on occasion.[10, 14]
In histology, neoplasms having an architectural pattern of development made up of straight-packed glands endowed with unevenly distributed lumina and without intervening stromal tissue are referred to as cribriform (from the old Latin cribrum, meaning sieve).[7] Invasive neoplasms that develop in several organs exhibit a cribriform pattern.[6, 10] Cribriform neoplasms such as carcinoma of the lung, prostate, stomach, and colon have been reclassified as more aggressive entities than previously thought.[10] Therefore, pathologists and oncologists may be affected in terms of practical prognostication by the discovery of a cribriform pattern.[7] Less than 4% of all breast cancers are invasive cribriform carcinomas, an uncommon histological subtype of invasive breast cancer.[10]

It is distinguished by angulated epithelial nests surrounded by a fibrosclerotic stroma and made up of mild to moderately atypical cells organized in a sieve-like arrangement.[10] Similar to our case, cribriform, trabecular growth pattern was predominant upon histopathology with few formed spaces containing pink/mucoid secretions. The tumor has round to oval nuclei, clump chromatin, inconspicuous to occasional prominent nucleoli, and indistinct cytoplasm. In between, a few angulated tubular glands are also noted. Despite the rarity of the presence of rare figures in mitosis, rare mitotic figures are seen in our patient. Unlike other invasive cribriform carcinomas, our case did not have microcalcifications.

Generally, cancerous nests have significant immunohistochemical positivity for cytokeratins, estrogen, and progesterone receptors, and lack the expression of myoepithelial markers including p63 and smooth muscle actin.[11] Nevertheless, our patient’s tumor had only weak estrogen receptor (ER) positivity, but negativity regarding progesterone receptor (PR) and human epidermal receptor 2 (HER2) as well.

Breast invasive cribriform carcinoma is divided into three types: pure invasive (when >90% of the entire lesion exhibits a cribriform pattern), invasive (when the cribriform pattern is combined with a component of tubular carcinoma that is 50%), and mixed (when 10–49% of the carcinoma exhibits a histotype other than tubular).[10] Our case’s lesion has shown to be pure invasive as it showed a cribriform pattern along with only a few tubular glands.

Both the left and right breasts are equally impacted by ACC of the breast, and tumors can develop in either breast quadrant. However, lesions are discovered in the subareolar area in roughly 50% of individuals.[16] According to reports, ACC typically manifests itself in the breast’s superior lateral quadrant or under the areola. Palpable masses are the most common reason for hospital admission for patients.[16] The mass is often single, and instances of numerous masses are seldom observed. Seven individuals in Alias et al. paper were reported to have a single tumor, and five of these patients’ tumors were all found in the superior lateral quadrant.[17] In our case, one lesion was discovered adjacent to the areola with only 1 cm and one in the superior lateral quadrant. The average ACC size is 3.0 cm, with a range of 0.7 to 12.0 cm.[18] The size of the lesion in our study was relatively similar to the other in the published literature (3x3 cm).

The pathological classification of ACC is under question.[15] Tumors with just tubular or cribriform cell structure are categorized into histological grade I, those with solid components between 30% and 70% are categorized into histological grade II, and those with solid components greater than 70% are categorized into histological grade III. The patient’s prognosis is poorer when the proportion of solid components is more prominent.[19]

The ACC’s imaging characteristics may vary. When an irregular tumor with an uncircumscribed margin appears, ACC has to be distinguished from other carcinomas. To prevent a missed diagnosis, a radiologist can easily identify it as BI-RADS-4 and order an additional biopsy.[20]

Similarly, in our case’s ultrasound, there was an irregular solid hypoechoic mass with lobulated margins and another lobulated large cystic lesion with multiple internal echoes. This led us to consider this lesion as BI-RADS-4 and proceed with tissue diagnosis with an ultrasound-guided core biopsy.

However, ACC lesions can present as a regular mass with a circumscribed margin. In Agafonoff et al. case report, the mammogram in their patient was noted to be BI-RADS-2 without direct or indirect signs of malignancy, likely in part due to the density of the patient’s breast tissue.[21, 22] An MRI was conducted due to the dense breast tissue and the inconclusiveness of mammography results. Except for T2 hyper-intensity in bigger lesions and T2 iso-intensity in smaller lesions, no consistent MRI characteristics have been shown. When it comes to identifying actual tumor characteristics and extent, MRI is more sensitive than mammography and ultrasound. Ultrasound usually demonstrates hypoechoic or heterogeneous mass with minimal vascularity similar to our case. Our case also has similarities and inconsistencies regarding MRI with Tang et al. paper of 9 mammary ACC cases who underwent MRI.[23] Of the largest masses had an extensive high T2-weighted imaging (T2WI) signal and hypointense internal septations, which demonstrated delayed enhancement. Dynamic enhancement illustrated washout kinetics. The 7 smaller masses appeared isointense on T2WI, and their internal septations were unenhanced. Among them, 5 demonstrated plateau kinetics and 2 demonstrated persistent kinetics. In our case, rapid enhancement and delayed washout (type 3 of the kinetic curve) was found. Nevertheless, all these findings indicated malignancy.

Therefore, imaging studies are nonetheless extremely useful in ACC cases for screening and management planning but might be non-specific. They could occasionally induce misreading due to their variable imaging characteristics. The standard reference is pathological examination.

The primary form of therapy for breast ACC patients is currently understood to be surgery. However, because this
pathological type is so uncommon, there is no clear direction in the selection of specific surgical treatments for this condition, which leads to variations in therapy. According to Ro et al., the surgical approach should be chosen based on the ACC grade. Grade I cancers should be treated with tumor lumpectomy, grade II tumors with mastectomy, and grade III tumors with mastectomy and lymph node dissection.\(^{[24]}\)

In our case, there were solid components, and it was considered a grade III tumor. Therefore, the patient underwent a right mastectomy with excision of axillary and sentinel lymph nodes. Thankfully, fresh frozen sentinel lymph nodes were proven to be free of malignancy.

**Conclusion**

Here we present a rare case of breast adenoid cystic carcinoma with a cribriform, trabecular growth pattern. Imaging studies are nonetheless extremely useful in ACC cases for screening and management planning but might be non-specific due to their variable imaging characteristics. The standard reference is pathological examination. Despite the fact that the pathological classification of ACC is still controversial, it is critically important in the decision of treatment options. However, the primary form of therapy for breast ACC patients is currently understood to be surgery. Because this pathological type is so uncommon, there is no clear direction in the selection of specific surgical treatments for this condition, which leads to variations in therapy.

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**Conflict of interest**

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**Ethics statement**

None.

**References**