Adenoid Cystic Carcinoma of the Breast, a Rare Entity: Report of Two Cases and Review of Literature

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Abstract
Adenoid cystic carcinoma (ACC) is a rare special subtype of breast cancer, accounting for approximately 0.1% of all breast cancers. It is commonly seen in the salivary glands. The imaging findings of this rare tumor have not been well described in the literature. Histologically, ACC is a triple negative tumor with basal-like breast features (tumors that are devoid of estrogen receptor, progesterone receptor, and human epidermal growth factor receptor 2 expressions, and express basal cell markers). For the most part, ACC is low-grade, that is how they have a good prognosis, whereas there are a rare lymph node involvement and distant metastasis.

In the current state of knowledge, Treatment is either simple mastectomy or lumpectomy with radiotherapy. There is no consensus on optimal treatment for patients with ACC. Otherwise, the role of chemotherapy and hormonal therapy remains controversial.

Here, we report two cases occurred in female patients who presented with a palpable breast mass in the breast that turned out to be an ACC of the breast.

Keywords: Adenoid cystic, breast, carcinoma

Case Report

Adenoid cystic carcinoma of the breast (ACC) is an uncommon subtype of invasive breast carcinoma. It was initially described as a cylindroma by Bill Roth in 1856. [1] It accounts for less than 0.1%. [1] While common in salivary gland, this tumor also has been seen in other organs including nasopharynx, trachea, uterine cervix, skin, lungs, Bartholin gland and kidneys as well as the breast. [2] It affects mainly women aged 50 to 60. [3] Clinically, it is typically a slow-growing mass. The radiologic findings of ACC are often nonspecific. ACC is a triple negative tumor with basal-like breast features (negative for the estrogen (ER), progesterone (PR) receptors, human epidermal growth factor receptor 2 expressions (HER 2 neu), and express basal cell markers). [3]

Despite being triple negative breast cancer, its prognosis is favorable, as lymph node involvement and distant metastasis are exceptional. [1, 4] The treatment modalities of this rare entity still to establish. It mainly consists on surgery and radiotherapy. Chemotherapy and hormotherapy remain controversial.

Here, we report two cases of ACC, and we discuss the imaging findings of ACC of the breast, with the corresponding molecular characteristics and treatment modalities of this interesting entity.

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Case Report

Case 1 – A 63-year-old woman, with an unremarkable past medical history and no family history, presented with a two-months history of a palpable mass in her left breast. Breast examination revealed a palpable mass about 3 cm in diameter in the upper outer quadrant of the breast without lymph node and no skin changes.

Ultrasonography revealed a hypoechoic and well-defined mass, measured 3 cm and mammography demonstrated a non-homogenous and hyperdense lesion (Fig. 1). The histological exam of samples from a core biopsy of the mass showed an adenoid cystic carcinoma. Chest X-ray, ultrasound of the abdomen, and bone scintigraphy were performed and there was no evidence of distant metastases. The patient underwent a left lumpectomy with lymph node dissection. The histopathological examination revealed a 32 mm in the largest dimension consisted of glandular, tubular, and predominantly massive, cribriform patterns (Fig. 2). Tumor cells were moderately atypical. Two mitoses were counted in ten high power fields. The stroma was fibrous and inflammatory with the presence of focal hyaline deposits and without necrosis. This proliferation freely infiltrates the adipose tissue (Fig. 2).

Moreover, the margins and all lymph nodes were free of disease. The neoplastic cells were negative for ER, PR, and HER-2/neu expressions. On the basis of the morphological findings, the tumor is reminiscent of the appearance of ACC of the salivary glands. The patient underwent local radiotherapy with a boost at the dose of 64 Grey. No loco-regional recurrence or distant metastasis was found after 6-months of follow-up.

Case 2 – A 47-years-old woman without a medical history, presented with a lump in the left breast. Clinical examination revealed three painless, mobile lumps, up to 5 cm in greater diameter; with homolateral lymph nodes and no skin changes. The Mammography of the left breast showed a multiple, irregular, hyperdense mass with irregular spiculated margins, measuring approximately 5 cm in greater diameter. Ultrasonography revealed multiple confluent, round to oval, hypoechoicogenic mass. A microbiopsy was performed and histological examination showed the patterns of an ACC. The staging workup didn’t show any metastatic lesion. Since the lesion was multifocal, the patient underwent a left mastectomy with axillary lymph node dissection. The histopathology examination revealed an adenoid cystic carcinoma of the left breast with free margins. The neoplastic cells were negative for ER, PR, and HER-2/neu expressions. All lymph nodes were negatives. The patient received adjuvant radiotherapy. After 60 months of follow-up, the patient is alive and free of disease.

Discussion

The ACC affects women in the middle age from 50 to 63 years, although it has been reported in a few cases in men.[5, 6] Clinical presentation is the same as invasive carcinoma, it presents as a painful mass measuring from 7 to 100 mm, the mean size is 3 mm, and mostly multifocal as we reported in the second case. Unlike ACC of salivary glands, the pain is not related to perineural invasion.[5] In our cases the mass was unpainful. More than 30% of the ACC are asymptomatic and it is discovered only in radiological assessment.[6]

There is no specific location, both breasts are affected but some authors reported that the periareolar region is most affected.[6] Metastasis in the axillary lymph node is rarely reported.

In mammography, ACC appears as irregular mass or asymmetric developing density. Sometimes, the tumor mimics to a benign lesion, it appears as a well-circumscribed mass. Calcification is rarely reported, in a few cases, the tumor is without traduction in mammography.[1, 5–7] Ultrasound shows a heterogeneous or hypoechoic mass, irregular with minimal vascularity in color Doppler.[1, 5–7]
The nonspecific radiological appearances of ACC lead to practice a biopsy to confirm the diagnosis.\[^{[1]}\]

Due to its favorable prognosis, the treatment is generally based on breast-conserving surgery, followed or not by radiotherapy. Indications of radical surgery are the same as the other invasive carcinomas or when the tumor is multifocal and had a high-grade pattern.\[^{[7]}\]

Some authors demonstrated that lumpectomy with radiotherapy obtains favorable local control, that leads to avoiding useless mastectomy.\[^{[5, 6]}\]

Unlike the other triple negative breast cancers, the axillary lymph node metastasis is not usually found. The frequency doesn't exceed 2% according to the most authors.\[^{[6]}\] It isn't demonstrated a benefit of the lymph node dissection in a low-grade or nonsolid variant of ACC,\[^{[5]}\] that it leads to considering sentinel lymph node.

The histologic characteristic pattern of ACC belongs to the basal-like subgroup of breast cancers.\[^{[7, 8]}\] It has heterogeneous morphology and shows three varied growth patterns: Glandular, tubular and solid. Although, ACC has a particular biphasic pattern that consists of true laminate and pseudocystic spaces; true glands are lined by epithelial cells and pseudocysts are lined by myoepithelial cells. There is three grade based on the proportion of solid elements.\[^{[1, 5, 6, 8]}\]

The Immunohistochemical findings on ACC categorize as a basal-like subtype of breast carcinoma that characterized by negative hormone receptors (estrogen and progesterone), the absence of expression of Her2 and expression of one or more basal/myoepithelial cell markers.\[^{[8]}\]

The ACC has an excellent prognosis. The 10-year survival rate is 90%-100%. Distant metastasis is usually in the lung and rarely in other areas.\[^{[5, 7, 8]}\]

**Conclusion**

ACC of the breast is a rare subtype of carcinoma that is characterized by a good prognosis. The radiological assessment is nonspecific, the treatment is mostly surgical. Axillary nodes metastasis are rare.

**Disclosures**

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