

## Primitive Adenoid Cystic Carcinoma of Breast: A Case Report

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### Case Report

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#### Article History

Received: 04.10.2018

Accepted: 17.10.2018

Published: 30.10.2018



**Abstract:** Adenoid cystic carcinoma (ACC) of the breast is a rare neoplasm. We report a case of 45-year-old female patient who consulted for a mastalgia. Clinical examination showed limited painful breast nodules. On ultrasound and mammography it was a well-limited mass without malignant signs. Core needle biopsy of the breast was done. Diagnosis was based on histopathological exam completed by immunohistochemistry. It's a triple negative phenotype. The patient was treated by mastectomy plus radiotherapy and remains alive in remission.

**Keywords:** Adenoid cystic carcinoma (ACC); Breast; Mastectomy; Radiotherapy.

### INTRODUCTION

Adenoid cystic carcinoma (ACC) of the breast, known formerly as cylindroma; is a rare malignant tumor. Histological, it is compared to salivary gland tumors of the same type, but it is distinguished by a slower evolution and a more favorable prognosis. It's represented in less than 0.1% of patients diagnosed with breast cancer [1].

Clinical signs are not specific and the paraclinical diagnosis based on ultrasound and mammography is difficult due to its rareness. Diagnosis is based on histopathological exam completed by immunohistochemistry.

We report a case of 45 years old female patient, who consulted for a mastodynia, the diagnosis leads to an adenoid cystic carcinoma of the breast.

### CASE REPORT

A 45-year-old woman, mother of three, allergic to penicillin, with no notable history, consulted on March 2017 for a right mastodynia. She was not on oral contraception with no reported family history of breast cancer.

Clinical examination showed a palpable mass, painful and mobile, in the right breast upper outer quadrant measuring 4cm\*4cm, and no nipple discharge. The skin of the breast was normal with one axillary lymph node.

Mammogram showed an asymmetric density in the right upper outer quadrant without microcalcifications. Ultrasound revealed an hypoechoic irregular nodule measuring 15mm\*8mm in the right upper outer quadrant which corresponded to a suspicious mass. The echogenicity was rather homogeneous and with posterior enhancement. Four indeterminate right lymph nodes were observed on the ultrasound of the axilla, without suspicious signs.

The mass was reported with a Breast Imaging-Reporting and Data System score of 4 according to ACR.

Core needle biopsy of the breast was performed. Histopathologic examination completed by immunohistochemistry showed interstitial collagen denaturation and myoepithelial hyperplasia and the diagnosis of adenoid cystic carcinoma was taken.

Staging scans did not reveal any definite distant metastasis. Her naso-endoscopy and magnetic resonance imaging (MRI) of the neck were normal. Extension assessment including chest x-ray and abdominal ultrasound was normal. We conclude to a primitive Adenoid cystic carcinoma of the breast.

A radical treatment was decided and a right modified radical mastectomy with axillary lymph node dissection was performed. The surgical margins were free from malignant cells. The operation was done on 01 April 2017 and was uneventful.

The histological study revealed an extensively infiltrative tumour grade II measuring 25mm\*14mm characterized by solid and cribriform nests, trabeculae of basaloid tumour cells intimately admixed with hyalinised and myxoidstroma. The mass was 7mm far from the deepest surgical limit. No ductal carcinoma in situ was seen. Lymph-vascular invasion was absent.

Immunohistochemistry showed a tumor negative for estrogen receptor, progesterone receptor, and Her-2 gene amplification (it's a triple-negative tumor ) with a dual population of intimately admixed luminal epithelial cells (CD117+, CK7+) and myoepithelial cells (p63+). The diagnosis of ACC was confirmed.

Adjuvant radiotherapy was performed. With a follow-up of 18 months, the patient is alive and in a situation of remission of her illness.

## DISCUSSION

Adenoid cystic carcinoma (ACC) is the most common malignant epithelial neoplasma of the salivary glands. ACC could be detected outside of the salivary glands in locations like the Bartholin's glands, uterine cervix, auditory canal, lacrimal glands, skin, lung, nasopharynx, and breast [2, 3]. Breast localization is exceptional; it represents only 0.1% of all breast cancers [4].

ACC of the breast is a rare malignancy with a considerable better prognosis than other subtypes of invasive breast cancer with rare invasion of the lymph node and infrequent metastases.

It mainly affects women from 30 to 90 years with the average age for development of this cancer is between the late 50's and mid 60's [5] but occasionally it develops in men as well. In our case, the patient was 45 years old.

Clinically ACC is presented as a well circumscribed palpable breast mass, often associated with mastalgia and nipple retraction [1].

There is a review done by (KPNCR) Northern California Cancer Registry of the Kaiser Permanente Northern California Region showed the majority 77% presented with a palpable breast lump while 45.5% reported breast pain [6]. For our patient mastodynia was the only symptom.

These lesions are mainly slow-growing and normally measuring 2-3cm. they mostly occur in the subareolar region and nipple discharge is actually an uncommon symptom.

The mammographic aspects are identical to other neoplasm and not specific; however the suggestive element is the lack of microcalcifications [7, 8].

Mammographic diagnosis is difficult; just few studies have described the imaging characteristics of these nonspecific aspects, ranging from well-circumscribed mass to poorly defined contours, with more or less architectural distortion. ACC may manifest as a smooth round mass similar to that of a benign tumor or as an irregular mass on mammography. Microcalcifications are rarely seen on mammogram.

On ultra-sound, ACC usually presents as a hypoechoic heterogeneous mass with minimal vascularity on doppler imaging and may have an indistinct margin [9].

In our case, the tumour measured 15mm\*8mm on imaging, much smaller than its final histological size of 25mm\*14mm.

Differential diagnosis with the mucinous carcinoma, the fibroadenoma or the ICC can be difficult [10] and can sometimes be helped by ultrasound or MRI [11].

Shear wave elastography (SWE) is used more and more to measure the stiffness of breast tumors. It allows the quantitative measurement of the rigidity of the lesion in kilopascals in vivo and could be done during routine ultrasound exams.

SWE has a high sensitivity and specificity in the differentiation of malignant and benign breast tumor [12]. It was reported that some malignant breast tumors presented typical peritumoral stiffness on the colored elastic map [13].

Tozaki and Fukuma [14] showed that the increased stiffness at the lesion margin of the breast was a sign of malignancy.

The diagnosis could be made on microbiopsy. The histological aspect is comparable to that of cystic adenoid carcinomas observed in other locations based essentially on the presence of dual population of both epithelial/luminal and myoepithelial/basaloid (abluminal) cell. Three different cytoarchitectural forms could be seen (tubular, cribriform, and solid), and a mixture of them is commonly observed [15].

Immunohisto-chemistry can be helpful to distinguish other tumors from ACC [15]. ACCs are immunohisto-chemically triple-negative subtype for hormone receptors (RE-, RP-, Her2-) but positive for P63, CD117, Calponin, CK8/18, CK14, CK5/6 and S-

100. Nevertheless, its prognosis is much more favorable than that of triple negative infiltrating ductal carcinoma.

The optimal treatment of ACC of the breast has not yet been codified due to its low incidence. ACC of the breast is generally treated by breast-conserving surgery with or without radiotherapy [16].

Mastectomy is recommended for invasive lesions when a cosmetically satisfactory excision is not possible or when the tumor has a high-grade pattern.

Surgically, both options of mastectomy or lumpectomy with radiotherapy had been associated with equivalent survival in ACC [17].

Ro *et al.*, [18] have classified the breast ACC based on the histological grading and proposed three therapeutic strategies:

- a lumpectomy for grade I (absence of solid elements)
- a mastectomy for grade II (<30 % solids)
- a mastectomy with axillary dissection for grade III (> 30% solids)

Axillary lymph node involvement is exceptional, ranging from 0.8 to 6.7%, as well as distant metastasis, which affect mainly lung, skin and lymph nodes [5, 6].

Adjuvant radiotherapy may decrease local recurrence for patients with conservation surgery [2] and a recent study shows the beneficial effects of radiotherapy on overall survival and disease free survival.

Thus it was recommended for our patient to undergo radiation. Hormone therapy is not indicated due to the lack of consistent expression of hormone receptors.

There are few publications evaluating the benefits of chemotherapy but systemic adjuvant chemotherapy could be proposed for patients with tumours bigger than 3 cm and with axillary lymph node invasion [17] or distant metastasis but its role in breast ACC patients still be controversial.

For our patient, she had a mastectomy with axillary dissection and underwent adjuvant radiotherapy.

ACC is considered as a breast tumor with a low malignancy potential. The local recurrence rate is low according to literature studies. Metastases are rare on. The overall survival rate is favorable; Arpino *et al.*, reported rates of 100% at five years and 93.8% at ten years [5].

## CONCLUSION

Adenoid cystic carcinoma is a rare breast tumor that essentially affects menopausal woman. The clinical signs as well as the radiological aspect are not specific, ACC could mimic a benign lesion on ultrasound SWE may be helpful. The diagnosis could be made on microbiopsy. The treatment is based on surgery and radiotherapy. This tumor has a very favorable prognosis

**Conflicts of interest:** There are no conflicts of interest.

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