Adenoid Cystic Carcinoma of the Breast: A Clinical Case Report

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ABSTRACT

Introduction: Adenoid cystic carcinoma (ACC) is an uncommon tumor of the breast, accounting for approximately 0.1% to 1% of all breast cancers. It is characterized by rare lymph node involvement and distant metastasis, and associated with a favorable prognosis with excellent survival, despite its triple-negative status. In the current state of knowledge, results of breast-conserving treatment with postoperative radiotherapy seem to be equivalent to mastectomy alone, with respect to survival for ACC of the breast. Due to its rarity, there is no consensus on optimal treatment for patients with ACC. Otherwise, the role of chemotherapy and hormonal therapy remains controversial. Further clinical studies are required to compare treatment options for ACC. But, a long-term follow-up is very important and mandatory for affected patients, due to the late onset of local relapse and occurrence of distant metastasis. Case report: Here, we report the case of a patient who presented with a palpable breast mass in the left breast that turned out to be an ACC of the breast.

Key words: adenoid cystic, breast, carcinoma, surgery, treatment.

1. INTRODUCTION

The salivary gland-like tumor of the breast also called adenoid cystic carcinoma (ACC) is a rare neoplasm that comprises less than 1% of all breast carcinomas (1). It has favorable prognosis, as lymph node involvement and distant metastasis are uncommon. ACC is characterized by a biphasic pattern and histologically, it consists of proliferating glands and stromal/basement membrane elements (1, 2). While common in salivary gland, this tumor also occurs at the other sites such as in the nasopharynx, trachea, uterine cervix, skin, lungs, and kidneys as well as the breast. The prognosis for ACC of the breast is better when compared to those in other locations.

ACC of the breast occurs between 30 and 90 years of age and more common in women in their fifth and sixth decade of life (2). It typically presents as a slow-growing mass, often in the subareolar area while some ACCs have been associated with breast tenderness and pain. Although radiological appearances are often nonspecific, the diagnosis can be made by fine-needle aspiration biopsy (3). ACC is generally negative for the estrogen (ER), progesterone (PR) receptors and HER-2/neu (c-erbB2) (2, 3). In the present case, we report a patient with a diagnosis of ACC of the breast, and aim to present the treatment approach of this rare disease.

2. CASE REPORT

A fifty-one-year-old woman presented with a two months history of palpable mass in her left breast. Family history was negative for breast and ovarian cancer. Breast examination revealed a palpable mass about 5 cm in diameter in the lower inner quadrant of the left breast with no skin changes. Also, no palpable axillary lymphadenopathy was detected.
Figure 1. The cribriform pattern composed of cylindromatous microcystic spaces in which basophilic mucoid material was filled with (x5.7, HE; red arrows showing breast acini).

Ultrasonography revealed a lobulated, irregular, and hypoechoic mass and mammography demonstrated an asymmetric and non-homogenous hyperdense lesion. Pathological evaluation of the core biopsy specimen revealed invasive ductal carcinoma of the breast. Chest X-ray, ultrasound of the abdomen, and bone scintigraphy were performed and there was no evidence of distant metastases. Then, a left modified radical mastectomy was performed. The histopathological examination revealed a 45 mm in the largest dimension consisted of cribriform, tubular and solid patterns. The cribriform pattern which was the dominant type in the tumor was characterized by nests of cells with cylindromatous microcystic spaces in which basophilic mucoid material was filled with. The tubular pattern was made up of well-formed ducts and tubules with central lumina lined by inner epithelial and outer myoepithelial cells. In the solid areas, sheets of uniform basaloid cells lacking tubular or microcystic formation were observed (Figure 1). Seven mitoses were counted in ten high power fields. Moreover, axillary lymph nodes were negative for metastases and the neoplastic cells were negative for ER expression and HER-2/neu but positive for PR (70%). On the basis of the morphological findings, the tumor was diagnosed as breast salivary gland type carcinoma, adenoid cystic carcinoma. After the operation, the patient’s treatment was modified radical mastectomy (5-7). Local recurrence rates after modified radical mastectomy or lumpectomy are not rare and varying from 6% to 37%, more so in patients not receiving adjuvant radiotherapy (1, 5, 7). Metastases may be seen many years after the initial diagnosis and prolonged survival rates are reported in several series (Table 1).

Table 1. Overview of adenoid cystic carcinoma of the breast.

| Reference          | Number of patients | Conservative treatment | Follow-up duration (months) | Lymph node involvement | Distant metastasis | ER/PR/Her2 status (positivity, %) | 5/10-year overall survival (%) |
|--------------------|--------------------|------------------------|----------------------------|------------------------|-------------------|----------------------------------|--------------------------------|
| Kulkarni et al (8) | 933                | 69.8%                  | 65.7 (median)              | 5.1%                   | —                 | 15/13/—                          | 88/—                           |
| Coates et al (9)   | 376                | 60.3%                  | 62 (mean)                 | 6.1%                   | 1.1%              | 12/10/—                          | 94/86                          |
| Ghobach et al (3)  | 338                | —                      | —                          | 1.7%                   | < 1%               | 10/8/—                            | 98/95                          |
| Thompson et al (10)| 244                | —                      | —                          | 4.9%                   | 2.9%              | —/—/—                            | 96/95                          |
| Khanfir et al (7)  | 61                 | 67%                    | 79 (median)               | 0%                     | 6.5%              | 3/5/—                            | 94/86                          |

Table 2. A summary of five major study in the literature related to the clinical characteristics of ACC of the breast.

The mean size of ACC is 3 cm (range, 1 to 12 cm) and it rarely (0.8% to 2%) causes axillary lymph node metastasis (6, 7). Nodal metastasis is rare and a routine axillary dissection is not recommended in this special type of cancer (5, 6). Distant metastases are uncommon and can occur without positive axillary nodes. Metastases are most commonly in the lung but also have been reported in liver, kidney, and bone (2, 8). Metastases may be seen many years after the initial diagnosis and prolonged survival rates are reported in several series (Table 1).

There is no consensus on optimal treatment for ACC of the breast because of its rarity. Surgical management options for treatment include lumpectomy, wide excision with or without radical radiation, or modified radical mastectomy (5-7). Local recurrence rates after lumpectomy or local excision are not rare and varying from 6% to 37%, more so in patients not receiving adjuvant radiotherapy (1, 5, 7). But adjuvant radiotherapy after local excision or lumpectomy decreases the local recurrence rates (6). Modified radical mastectomy is the most reported surgical procedures for ACC of the breast, simply because it used to be the standard treatment for common breast cancers (5). However, results

3. DISCUSSION

Adenoid cystic carcinoma (ACC) of the breast was first termed “cylindroma” by Billroth in 1856 and ACC...
of breast-conserving treatment that includes adjuvant radiotherapy seem to be equivalent to those of mastectomy alone, with respect to survival (5, 9). Mastectomy is recommended for invasive lesions when a cosmetically satisfactory excision is not possible, especially when the tumor has a high-grade pattern (6).

In contrast to other triple-negative breast carcinomas, ACCs have a relatively good prognosis and they are usually low-grade (3-5). In present case, the patient’s receptors status was followed as: ER-negative, PR-positive (70%), and HER-2/neu-negative. The modified radical mastectomy was performed which showed no axillary node involvement. The patient was not given radiotherapy; however, chemotherapy and hormonotherapy (tamoxifen) were administered postoperatively.

As shown Table 2, after definitive treatment, the 10-year overall survival rate is 86% to 95%, and lymph node metastasis is very rare, as well as distant metastases (3, 7-10). Data from the published literature indicate that ACCs of the breast have a very good prognosis after achieving local control, and the overall survival is not significantly different from the general population. These findings suggest that patients with ACC would derive little if any improvement in survival from adjuvant systemic therapy and thus can be spared the side effects and the cost of such therapy (1, 6).

4. CONCLUSION

With the information available today, the treatment of ACC of the breast is seen as to be mastectomy and addition of axillary lymph node dissection in patients with positive sentinel lymph nodes or breast-conserving treatment that includes adjuvant radiotherapy. Routine axillary lymph node dissection is not recommended due to the fact that the metastasis is less than 2%. Because of the excellent prognosis with ACC, the benefit to survival from systemic therapy is negligible. Due to the late onset of local relapse, as well as distant metastasis, a long-term follow-up is very important and mandatory for these patients. Further clinical investigations comparing treatment options for ACC are inevitable to define the optimal treatment.

- **Informed Consent:** Written informed consent was signed by the patient and responsible physician.

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