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Adenoid Cystic Carcinoma of the Breast: a Case Report with Imaging Findings

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Abstract : Adenoid cystic carcinoma is a uncommon variant of adenocarcinoma that ususally occurs in the salivary gland. In breast, adenoid cystic carcinoma is a very rare carcinoma accounting for less than 1% of the all breast carcinoma. It has an excellent prognosis with the lower incidence of distant metastasis and axillary lymph node involvement, and a benign looking or low suspicious findings on imaging. We report the imaging finding of a case of an adenoid cystic carcinoma of the breast in a 61-year-old woman.

Key Words : Adenoid cystic carcinoma, Breast neoplams

Introduction

Adenoid cystic carcinoma is a relatively rare variant of adenocarcinoma that usually occurs in the major and minor salivary glands but may occasionally arise in other organs including the breast, tracheobronchial tree, uterine cervix, larynx, and Bartholin's gland [1-5]. The origin of cell is obscure, but evidence suggests that it may be derived from the ductal epithelium and myoepithelium [6]. Pathologically, the presence of both cell types is necessary for the diagnosis of adenoid cystic carcinoma of the breast.

Adenoid cystic carcinoma of the breast accounts for 0.1-0.4% of all breast cancers. In contrast to extramammary adenoid cystic carcinoma, those arising in the breast have an favorable prognosis [7,8]. This unusual tumor of the breast displays slowly progressive growth and rarely metastasizes to the axillary lymph nodes. Distant metastases are uncommon. It is important to distinguish this tumor from other types of breast cancers as it

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has an excellent prognosis.

The imaging features of adenoid cystic carcinoma of the breast have scarcely been reported in the literatures to our knowledge. We report mammographic and utrasonographic findings of a case of adenoid cystic carcinoma of the breast.

Case

A 61-year-old woman presented with a palpable mass at right breast for one year. Her past medical history was not significant, and family history was negative for breast and ovarian cancer. Upon physical examination, we noted a non-tender, large mass with a relatively circumscribed margin in the upper central portion of the right breast. There were no skin changes nor was there nipple discharge. No palpable axillary adenopathy was noticed.

The mammograms revealed a 10 x 8 cm, well-demarcated, smoothly marginated, slightly lobulated, oval shaped, high-density mass in the upper central portion of the right breast [Fig. 1A]. No associated calcification were observed.

Ultrasound showed a large well-defined, slightly lobulated, heterogeneous mixed echogenic mass [Fig. 1B,C]. Color Doppler study showed some vascularity in the mass. We performed US-guided automated gun biopsy using a 14-gauge needle and histologic examination of the five core biopsy specimens revealed fibroepithelial tumor with atypical epithelium and myoepithelial proliferative lesion.

The patient underwent total mastectomy with sentinel lymph node biopsy.

Upon gross pathology, we did observe a 9 x 8 x 6cm sized, well-demarcated, round to oval shaped, solid mass [Fig. 1D]. Cut surface of the mass showed vague septation or lobular structure and pale tan to yellow, solid and granular. Microscopically, the tumor showed cribrifrom and invasive pattern. The tumor consists of gland-like component filled with pale basophilic cylindromatous component [Fig. 1E]. The tumor cells are positive for p63, CD117, and bcl-2 and negative for ER, PR, c-erbB-2, and p53. Sentinel lymph nodes were negative for metastases.

The patient remains well to date.

Discussion

Adenoid cystic carcinoma is a relatively rare tumor that constitutes 4-15% of all salivary gland tumors. It is usually found in the minor salivary glands, where it constitutes 25-31% of malignant neoplasms and is the most common malignant tumor [9]. This tumor constitutes 15% of tumors of the submandibular gland but only 2-6% of tumors of the parotid gland [10]. Despite the relatively benign histologic appearance and slow growth, adenoid cystic carcinoma of head and neck is characterized by a slow relentlessly malignant course. Repeated recurrences and distant metastases occur over many years. Repeated surgical excision and radiation therapy are treatment of choice.

Adenoid cystic carcinoma of the breast is a very rare neoplasm. It was first described in 1946 by Foote and Stewart [11]. The largest series continue to be the 21 cases reported in 1969 by Cavanzo and Taylor [12] and the 23 cases described in 1989 by Rosen [13]. The

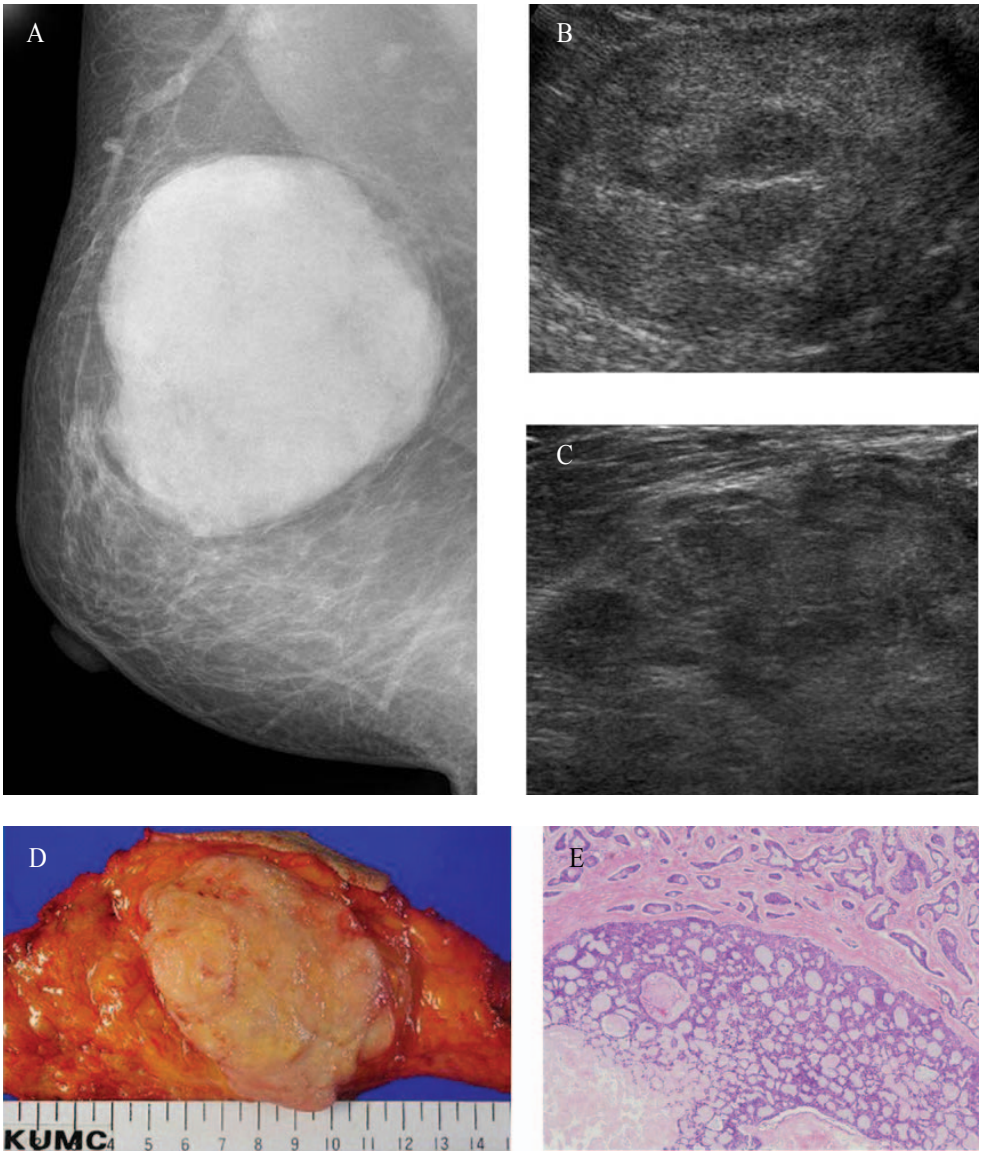

Fig. 1. Adenoid cystic carcinoma of the breast in 61-year-old women. (A) Right mediolateral oblique mammogram reveal 10 x 8 cm, smoothly marginated, lobulated, high-density mass on upper central portion of right breast. (B, C) Transverse and longitudinal US scan reveal well-defined, lobulated, heterogeneous, mixed density mass. (D) Gross pathology show well-demarcated, round to oval shaped, pale tan to yellow, solid and granular mass with vague septation and lobular structure. (E) Microscopically, tumor shows cribriform pattern (lower) and invasive pattern (upper). The tumor consists of gland-like component filled with pale basophilic cylindromatous component.

tumor demonstrates a strikingly characteristic microscopic pattern similar to that of adenoid cystic carcinoma of the salivary glands and accounts for 0.1−0.4% of all breast cancers. It displays slowly progressive growth and rarely metastasizes to the axillary lymph nodes. Distant metastases are uncommon and therefore has a favorable prognosis [7,8].

Adenoid cystic carcinoma arising in the breast occurs predominantly in women with a

mean age of from 50 to 64 years; it is rarely bilateral and has no predilection with respect to laterality [13]. Typically, adenoid cystic carcinoma presents as a mass that is occasionally tender to palpation. The tumor is rarely fixed to the overlying skin, nipple, or pectoral muscles [6]. Although any part of the breast can be involved, most adenoid cystic carcinoma are found in either the central or subareolar region. Despite this location, nipple discharge is rarely present [13]. Pain is a symptom of adenoid cystic carcinoma of the salivary gland, the cause being attributed to perineural invasion by the tumor. However, perineural invasion in adenoid cystic carcinoma of the breast is rare. Our patient complained a large, upper cental palpable mass and perineural invasion but no pain.

The mammographic features of adenoid cystic carcinoma have been scarcely reported in the literature [14,15]. A review of the literature shows that its radiological appearances are generally nonspecific and it can present as a benign-appearing, smooth, round or lobulated density or as an irregular mass. In a review of mammographical appearances with pathologic correlation by Santamaria *et al.* [15], mammography most often disclosed a circumscribed lobulated nodule, usually in the upper quadrant or in a peri-areolar region. They also found that masses that appeared ill-defined or partially ill-defined on mammography could usually be correlated with tumors showing microscopic invasion. Calcifications may be seen on histology but only rarely detected on mammography. Radiologically, our case appeared as a well-defined, smoothly marginated, large mass on mammographic examination. Calcification was not associated.

Only few case reports for ultrasonographic features of adenoid cystic carcinoma have been reported on the literature. Lee *et al.* [16], described as round hypoechoic nodule with a partially indistinct margin, difficult to distinguish malignant lesion. Our case appeared as a large, well-defined, oval shaped, slightly lobulated, heterogeneous mixed echogenic mass.

Pathologically, various growth patterns have been described in adenoid cystic carcinoma of the breast. This patttern have been described as glandular (cribriform), tubular, and solid (basaloid) types [17]. These three structural types of adenoid cystic carcinoma frequently occur in combination. The glandular type consists of epithelial cell rests permeated by numerous cylindrical spaces, some of which are occupied by a hyaline stroma. The tubular type consists of epithelium strands surrounded by a hyaline stroma, and tubular lumina are seen among these structures. The solid type consists of solid epithelium strands, often with central necrosis. In any subtype, the two main cell components of adenoid cystic carcinoma are modified fusiform myoepithelium cells and glandular cuboid cells [6,18]. Adenoid cystic carcinoma showed low proliferative activity and were usually ER and PR negative. The differential diagnosis of the glandular and tubular subtypes includes cribriform intraductal carcinoma, invasive ductal carcinoma with a cribriform pattern, papillary carcinoma, and medullary carcinoma, whereas adenoid cystic carcinoma of the solid type can resemble ordinary ductal carcinoma [13,18].

Mammographically, the differential diagnosis of adenoid cystic carcinoma is difficult. If the tumor appear as a fairly well-

defined nodular lesion, then the diffential diagnosis includes either a benign tumor, such as fibroadenoma, or a malignant circumscribed lesion, such as a medullary carcinoma, mucinous carcinoma, or some other type of sarcoma and metastasis. If the tumor appears as a focal distortion, then the differential diagnosis includes invasive ductal carcinoma. Regardless of apparance, a preoperative cytologic examination or core biopsy frequently suggests a diagnosis.

Patients with adenoid cystic carcinoma of the breast have an excellent prognosis because of the low incidence of distant metastases and axillary lymph node involvement [6]. However, of all adenoid cystic carcinoma subtypes, tumors with a solid component tend to be larger, and recurrence and metastases are more frequently seen. While the lungs are the most frequent site, other reported sites of metastasis include the liver, kidneys, lymph nodes and brain [19].

There is no consensus on optimal treatment for patients with adenoid cystic carcinoma of the breast as this tumor is such a rare diagnosis. Reported surgical treatment modalities range from a simple lumpectomy with radiotherapy to radical mastectomy. Little has been published to date on the role of adjuvant systemic therapy.

Summary

We have presented a case of adenoid cystic carcinoma of the breast in a 61-year-old women, including mammographic and ultrasonographic findings.

Reference

1. Cavanzo FJ, Taylor HB. Adenoid cystic carcinoma of the breast: an analysis of 21 cases. *Cancer* 1969;**24**: 740-5.
2. Hajdu SI, Huvos AG, Goodner JT, Foote FW Jr, Beattie EJ Jr. Carcinoma of the trachea: clinocopathologic study of 41 cases. *Cancer* 1970;**25**:1448-56.
3. Prempree T, Villasanta U, Tang CK. Management of adenoid cystic carcinoma of uterine cervix (cylindroma): report of six cases and reappraisal of all cases reported in the medial literature. *Cancer* 1980;**46**:1631-5.
4. Olofsson J, Van Nostrand AWP. Adenoid cystic carcinoma of the larynx: a report of four cases and a review of the literature. *Cancer* 1977;**40**:1307-13.
5. Addison A, Parker RT. Adenoid cystic carcinoma of Bartholin's gland: a review of the literature and report of a patient. *Gynecol Oncol* 1977;**5**:196-201.
6. Leeming R, Jenkin M, Mendelsohn G. Adenoid cystic carcinoma of the breast. *Arch Surg* 1992;**127**:233-5.
7. Lerner AG, Molnar JJ, Adam YG. Adenoid cystic carcinoma of the breast. *Am J Surg* 1974;**127**:585-7.
8. Zaloudek C, Oertel YC, Orenstein JM. Adenoid cystic carcinoma of the breast. *Am J Clin Pathol* 1984;**81**:297-307.
9. Nascimento AG, Amaral ALP, Prado LAF, Kligerman J, Silveira TRP. Adenoid cystic carcinoma of salivary glands: a study of 61 cases with clinicopathologic correlation. *Cancer* 1986;**57**:312-9.
10. Blanck C, Eneroth CM, Jacobsson F, Jacobsson PA. Adenoid cystic carcinoma of the parotid gland. *Acta Radiol Ther Phys Biol* 1967;**6**:177-96.
11. Foote Jr FW, Stewart FW. A histologic classification of carcinoma of the breast. *Surg* 1946;**19**:74-99.
12. Cavanzo FJ, Taylor HB. Adenoid cystic carcinoma of the breast: an analysis of 21 cases. *Cancer* 1969;**24**:740-5.

13. Rosen PP. Adenoid cystic carcinoma of the breast: a morphologically heterogeneous neoplasm. *Pathol Annu* 1989;**24**:237-54.
14. Bourke AG, Metcalf C, Wylie EJ. Mammographic features of adenoid cystic carcinoma. *Australas Radiol* 1994;**38**:324-5.
15. Santamarina G, Velasco M, Zanon G, Farrus B, Molina R, Sole M, *et al*. Adenoid cystic carcinoma of the breast: mammographic appearance and pathologic correlation. *AJR Am J Roentgenol* 1998;**171**:1679-83.
16. Lee MS, Kim MK, Kim EK, Park BW, Oh KK. Adenoid cystic carcinoma of the breast: a case report. *J Korean Soc Ultrasound Med* 2005;**24**:199-202.
17. Seifert G, Brocheriu C, Cardesa A, Everson JW. WHO international histological calssification of tumors: tentative histological calssification of salivary gland tumors. *Pathol Res Pract* 1990;**186**:555-81.
18. Sumpio BE, Jennings TA, Sullivan PD, Merino MJ. Adenoid cystic carcinoma of the breast: data from the Connecticut Tumor Registry and a review of the literature. *Ann Surg* 1987;**205**:295-301.
19. Arpino G, Clark GN, Mohsin S, Bardou VJ, Elledge RM. Adenoid cystic carcinoma of the breast: molecular markers, treatment and cliical outcome. *Cancer* 2002;**94**:2119-27.