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A Rare Case of Adenoid Cystic Carcinoma of Breast - A Case Report

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Abstract

Background: Adenoid cystic carcinoma of the breast (ACC) is a rare tumor, comprising <0.1% of all breast cancers. It has a unique dual-cell pattern (both luminal and basal) and is indistinguishable from ACC arising from salivary tissue. It is a low-grade, triple-negative tumor with favourable prognosis, and rarely show metastasis.

Case Report: A 50years old female presented with right breast nodule. Initial imaging revealed BIRADS IVa. For further evaluation lumpectomy was done and histopathology report and Immunohistochemistry report confirmed Adenoid cystic carcinoma of breast.

Conclusion: This case highlights the rare presentation and diagnostic features of breast ACC, contributing to the limited literature on this uncommon malignancy.

Keywords: Adenoid Cystic Carcinoma, Tumor, Breast Nodule, Malignancy

Introduction: Adenoid cystic carcinoma of the breast (ACC) is a rare tumour, comprising <0.1% of all breast cancers. It is more commonly seen in older women, with Caucasian women being at greatest risk[3-5]. Most cases present as a painful, well circumscribed and palpable mass. In spite of being triple negative (ER, PR, HER-2 negative) it has favourable prognosis with 10- year survival rate ranging from 80% to 100%.[1-4, 7]

WHO classified it into low grade, intermediate grade and high grade on the basis of presence of percentage of histological types (Cribriform, Tubular, Solid).[2] Histologically, it has a dual-cell pattern (both luminal and basal cells) similar to ACC arising from salivary tissue.[1-5] Immunohistochemically, it typically shows negativity for ER, PR and HER2 but is positive for

myoepithelial markers such as p63, SMA, calponin and CD117(c-KIT).[1-4,7] It rarely metastasizes to axillary lymph node.[1,2,5,8] Tumor usually exhibits low proliferative activity, often reflected by low Ki-67 index.[2]

Given the rarity of this cancer, treatment guidelines have yet to be well established. Current treatment is focused on surgical resection with negative margins, often followed by adjuvant radiation therapy. Chemotherapy is generally not done due to limited metastatic potential of tumor.[5,8]

Case presentation: A 50years old female presents with right breast nodules. On examination it was firm to hard, single, involving areolar skin, non-tender, fixed measuring 2x1 cm in size.

Radiological Findings

Well-defined heterogeneous hypoechoic lesion, parallel in orientation, measuring ~35x11 mm is noted in the lower outer quadrant of right breast at 7-8'O clock position, ~1-2 cm away from nipple areolar complex with few echogenic foci. On colour doppler examination, mild internal vascularity is noted. Overlying skin appears retracted with focal thickening-BIRADS IVa.

Pathological Findings

FNAC was done from nodule. Smears showed high cellularity with features suggestive of Proliferative Breast Disease with Atypia (YOKOHAMA CATEGORY III). Nodule was excised and sent for histopathological examination. Grossly received single nodular skin covered tissue mass measuring (5x3x2.5) cm (Fig 1).

Cut section shows ill-defined firm mass measuring (3x2x1.5) cm with focal areas of haemorrhage, grey white to grey tan in colour, hard in consistency.

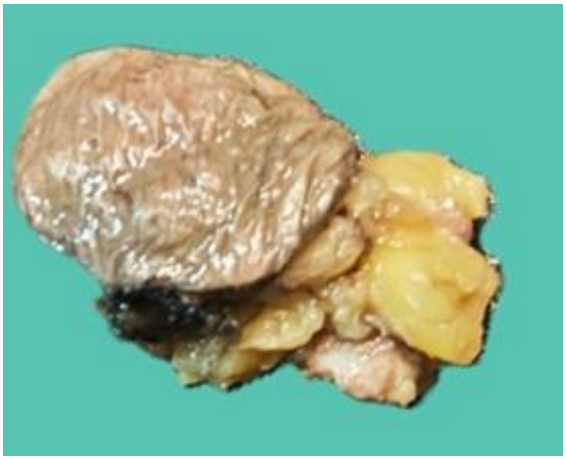

Figure 1: Gross lumpectomy specimen

Microscopy from multiple sections from breast nodule shows an ill-defined infiltrative dermal tumour comprised of cells arranged in solid nests and cribriform pattern (Fig 2). Individual cells appear basaloid, having hyper-chromatic oval nuclei and scanty indistinct cytoplasm (Fig 3). Periphery of the tumour islands exhibit deposition of basement membrane material. Mucin filled glandular spaces with focal areas of sebaceous differentiation seen. Surrounding stroma is fibrous with focal dense mononuclear inflammatory infiltrates. The features are of Malignant Epithelial Neoplasm, possibly Adenoid Cystic Carcinoma (Right Breast), (Solid-Basaloid type with focal Sebaceous differentiation).

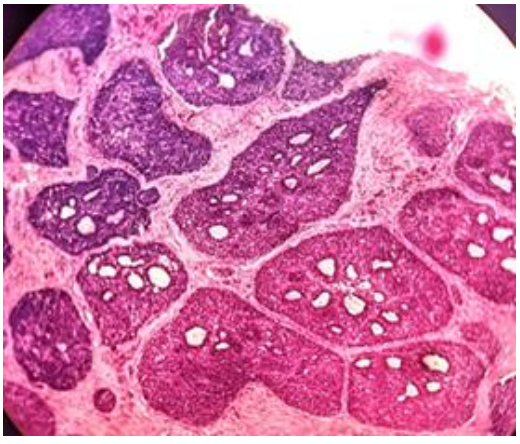

Figure 2: 10X H&E image showing tumor cells arranged in solid nest and cribriform pattern.

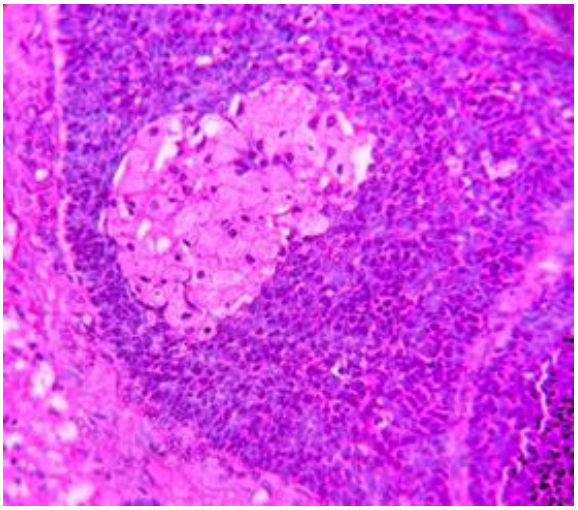

Figure 3: 40X H&E image showing dual cell population with characteristic pseudo cystic spaces in ACC.

Immunohistochemical staining with SMA confirmed presence of myoepithelial layer around the cribriform structures consistent with adenoid cystic carcinoma (Fig 4). Immunohistochemistry revealed diffuse strong cytoplasmic positivity for CK in luminal epithelial cells (Fig 5). Immunohistochemically, the tumor demonstrated a triple negative phenotype- Estrogen receptor (ER)-negative (0%), Progesterone receptor (PR)-negative (0%) and HER2- negative (score 0).

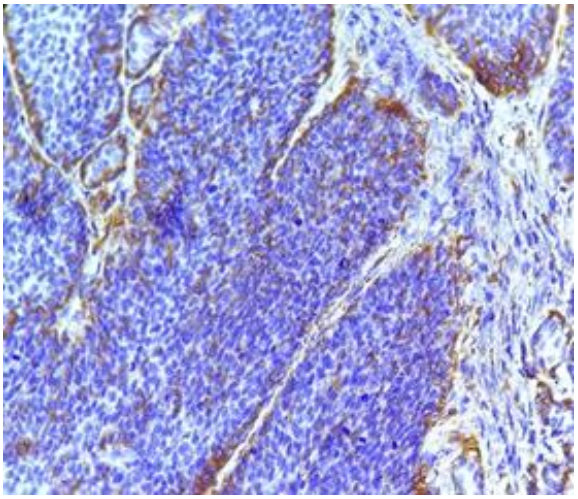

Figure 4: 40X image of IHC staining showing SMA positivity in myoepithelial cells.

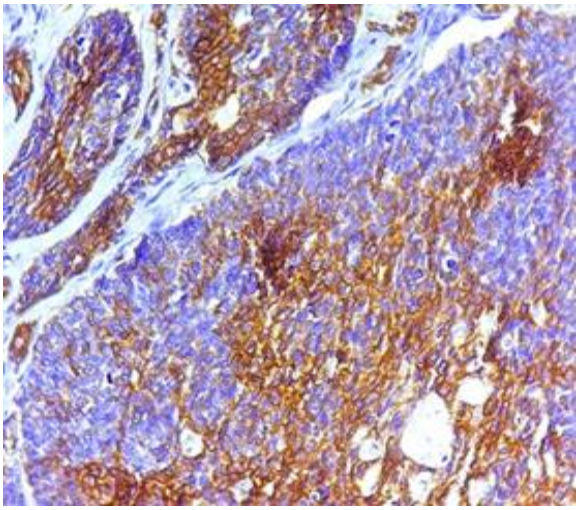

Figure 5: 40X image of IHC staining showing diffuse cytoplasmic positivity of CK in luminal epithelial cells.

Discussion

Adenoid Cystic Carcinoma is a rare tumour comprising of <0.1% of all breast cancers. It is more commonly seen in older women, with Caucasian women being at greatest risk.[1-4,7]

ACC presents as palpable mass in outer quadrants of breast, sometimes associated with pain. Mammogram or breast ultrasound shows mostly hypoechoic solid mass.[7] Histologically, it is characterised by presence of dual cell pattern (both luminal and basal cells), similar to those of salivary gland tumour.[1-5] It should be correctly distinguished from other differential diagnosis of Collagenous Spherulosis (CS) on basis of formation of ductal epithelium, as in CS ductal lumina of various sizes are evident. Other differential diagnosis, Cribriform Carcinoma must also be ruled out, as cribriform pattern is seen in both.[1,9] In this case an immunohistochemistry should be done to highlight biphasic architecture. The ductal component typically expresses CK and Cam5.2, while the myoepithelial component shows positivity for markers such as SMA, S100, calponin, p40, p63 and certain cytokeratin.[2]

Three distinct architectural growth patterns are recognized: cribriform, tubular, and solid.[2]

Importantly, the 5th edition of the WHO Classification of Breast Tumor delineates three histologic subtypes of adenoid cystic carcinoma (ACC) with distinct clinicopathological profiles:

1. Classic ACC (C-ACC): Characterized by cribriform and tubular growth patterns and low-grade cytology, this subtype follows an indolent clinical course.
2. Solid-Basaloid ACC (SB-ACC): This subtype exhibits solid nests, nuclear atypia, and necrosis, and is associated with a higher risk of recurrence and metastasis.
3. ACC with High-Grade Transformation (ACCHGT): A newly recognized, aggressive variant defined by an abrupt transition from classic ACC to an undifferentiated carcinoma, featuring loss of the characteristic biphasic morphology and increased mitotic activity. [2]

Our patient had Solid-Basaloid ACC.

ACC is typically negative for ER, PR and HER2Neu but having favourable prognosis with a 10-year Survival rate ranges between 80% to 100%.[1-4,7] Lymph node involvement or metastasis is rare, so mostly nodal resection is not done and only lumpectomy is done.[5,8]

Conclusion

This case highlights the importance of recognising the unique imaging, histopathologic and molecular features of adenoid cystic carcinoma of breast – an uncommon subtype of triple-negative breast cancer. Unlike typical triple-negative breast cancers, which are often aggressive with poor prognosis, breast ACC carries a favourable outlook with low rates of lymph node involvement and distant metastasis. Although it has favourable prognosis and have low risk of mortality, it still needed to be properly differentiated for proper management.

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