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Case Report

Adenoid cystic carcinoma of breast: A rare case report

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ABSTRACT

Adenoid cystic carcinoma of breast is rare, accounting for less than 4% of all breast malignancy. We are presenting a case of 57 Year old Hindu female patient who presented with chief complaint of right breast swelling since 5 years associated with pain.
USG report showed well defined isoechoic to hypoechoic lesion without internal vascularity, with a possibility of fibroadenoma of right breast. FNAC of the lesion was also suggestive of fibroadenoma. The wide local excision was done, tissue specimen was sent for histopathological examination which was diagnosed as adenoid cystic carcinoma of right breast after immunohistochemical characterization. Immunochemistry markers show ER, PR HER 2: negative, P -63: Positive for myoepithelial cells, Ki 67: <10%, CD 117/ c kit: Positive for ductal cells.

1. Introduction

Adenoid cystic carcinomas (ACC) of the breast are rare tumors with unique histopathological features. In contrast to the other triple negative breast [ER, PR, Her 2 neu] breast carcinomas. ACCs is characterized by low expression of Ki 67, low malignant potential, slow progression and favorable prognosis.

2. Case Report

A 57 Year old retired female presented with chief complaint of right breast swelling since 5 years associated with pain. Patient was relatively asymptomatic before 5 years, then she developed swelling over right breast on lower outer quadrant of breast. The swelling was initially pea-sized then progressed to its current size 1 cm X 1 cm. Swelling is not associated with menstrual cycles or cyclic mastalgia, non-radiating, not associated with discharge from the swelling site or fever. After clinical examination and radiological investigations patient was diagnosed with right breast lump and was planned for wide local excision.

Grossly: The specimen measures 5 x 4 x 1.7 cm. On cut section it is solid and whitish.

2.1. Microscopic examination

Sections show infiltration by tumor composed of nests & strands of neoplastic cells showing cribriform and solid structures in different proportion having pleomorphic, hyperchromatic and vesicular nuclei with prominent nucleoli. Mitotic figures are present. The tumor was diagnosed as infiltrating ductal carcinoma and Impression of Ductal carcinoma was made and was further advised for Immunohistochemistry for confirmation and typing.

2.2. Special stain and immunohistochemistry report

PAS: Positive for intraluminal secretions
ER/PR/Her 2 neu: Negative
Ki 67: <10%

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P -63: Positive for myoepithelial cells

117/ c kit: Positive for ductal cells

Overall findings were reported as triple negative low grade carcinoma with cribriform pattern with possibility of adenoid cystic carcinoma.

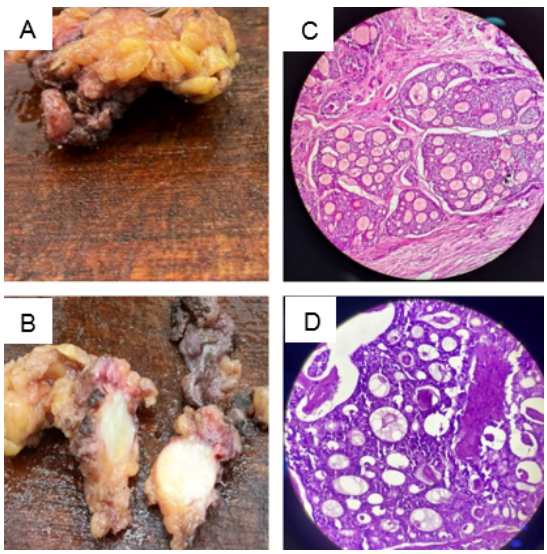

Figure 1: A: Gross picture of breast tissue; **B:** Cut section of breast tissue shows one whitish solid area; **C:** Dual population of basaloid and epithelioid cells, extensive cribriform architecture; **D:** Periodic acid Schiff (PAS): Intraluminal secretions are PAS positive.

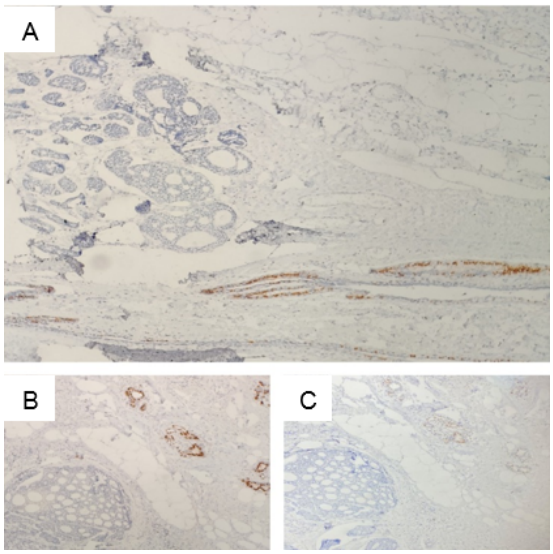

Figure 2: A: IHC for ER: Negative for ER with Positive internal control on bottom right side; **B:** IHC for PR: Negative for PR with Positive internal control on right side; **C:** IHC for Her 2 neu: Negative for Her 2 neu with positive internal control on right side.

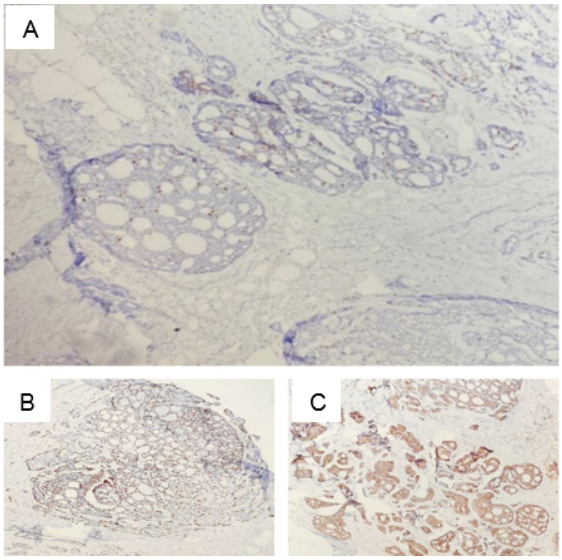

Figure 3: A: Ki 67< 10%; **B:** IHC for P 63: Positive for myoepithelial cells; **C:** IHC for CD -117: Positive for ductal cells.

3. Discussion

Adenoid cystic carcinoma is histologically indistinguishable from its counterpart in the salivary gland, characterized by a dual cell population consisting of epithelial and myoepithelial cells arranged in tubular, cribriform, and solid growth patterns, and is frequently associated with MYB::NFIB fusion.

Histologically, adenoid cystic carcinoma commonly takes on the appearance of nodular growth pattern but may also be infiltrative. Three subtypes have been defined based on architectural and cytological characteristics, including classic and solid-basaloid variants and adenoid cystic carcinoma with high-grade transformation.[1] The classic subtype often shows a mixed growth pattern, most frequently with a cribriform architecture, in which fenestrated nests of cells form myoepithelial cell–lined pseudolumina containing mucoid or basement membrane–like material, giving rise to a "Swiss cheese" appearance. The mucoid or basement membrane–like material is positive for Alcian blue and immunoreactive for type IV collagen. In the tubular growth pattern, tumor cells are arranged in conspicuous ducts and tubules containing mucin and basement membrane material, lined by inner luminal epithelial cells and outer myoepithelial cells. The cribriform areas may mimic invasive cribriform carcinoma, but the latter can be excluded by the dual cellular composition and the expression of estrogen receptor (ER) and progesterone receptor (PR) in the lesional cells. Expression of ER and PR is typically strong and diffuse in cribriform carcinoma, but is absent or focal in adenoid cystic carcinoma.[2]

Immunohistochemistry (IHC) is helpful in confirming the multi-lineage cellular components, as the epithelial

cells consistently express low-molecular-weight keratins (ie,CK7, CK8, AE1) whereas myoepithelial cells can be labeled by high-molecular-weight keratins (such as CK5/6), smooth muscle myosin heavy chain, and p63. c-kit (CD117) is usually positive with variable intensity, more prominent in the luminal component of the tumor. However, c-kit expression is not specific for adenoid cystic carcinomas, as a number of other breast tumors can also express this protein, including secretory carcinoma and adenomyoepithelioma, and the latter may demonstrate histomorphology similar to that of adenoid cystic carcinoma.[3]

The MYB::NFIB fusion gene has been identified in most classic adenoid cystic carcinomas and discovered in its rare subtypes as well.[4] Mutations in EP300, NOTCH1, and FGFR1 have been reported in adenoid cystic carcinomas with high-grade transformation. The latter lacks mutations frequently harbored by conventional triple-negative breast cancers, such as TP53 mutations.[5]

Classic adenoid cystic carcinomas typically demonstrate a triple-negative phenotype, although low ER and PR expression can be seen. These tumors show a favorable clinical outcome. Complete surgical resection is usually curative.[6–8]

4. Conclusion

ACCs are breast tumors with a unique histology, immunohistochemical profile, and a paradoxically good prognosis. Studies to date have been unable to identify the molecular mechanisms or precursor lesion for ACC. This information is critical to enhance our understanding of this triple negative tumor and add to our insight into the more aggressive basal type breast carcinomas, ultimately identifying possible therapeutic targets.

5. Conflict of Interest

None.

6. Source of Funding

None.

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