

Adenoid cystic carcinoma of breast: An unusual breast malignancy

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EPIDEMIOLOGY

Incidence is 0.1–1% of all breast cancers. Till date only 933 cases have been documented [1, 2]. Adenoid cystic carcinoma (ACC) is an unusual breast malignancy with treatment strategy under evaluation [3].

Age: The age of occurrence is 30–90 years of age. However, it is more common in 50–60 years females. Exceptionally, it is seen in young age also [4]. It is extremely uncommon in males (only eight cases have been documented as per the existing literature) [5–13].

CLINICAL FEATURES

Most patients present with a dominant breast mass tender to palpation [14, 15]. This associated pain has been suggested to be due to the contractile myoepithelial component of these tumors, as perineural invasion is not commonly seen in these lesions [16, 17]. All quadrants seem to be affected, with a particular trend for the periareolar region [16].

DIAGNOSTIC EVALUATION

1. *Fine needle aspiration cytology (FNAC):* It can be diagnosed on FNAC [18, 19]. The aspirates are cellular and show extracellular metachromatic

material surrounded by uniform cells with scanty amount of cytoplasm. Cytology reveals atypical epithelial cells with absence of bare nuclei mimicking as atypical epithelial hyperplasia [20] (Figure 1).

2. *The core biopsy:* It can be used as an effective diagnostic modality and reveal prominent solid, microcystic, and focally tubuloglandular patterns of growth with intact myoepithelial layer [20] (Figure 2).
3. *Immunohistochemistry:* On immunohistochemical evaluation, the tumor cells are immunopositive for CK7 expression (Figure 3) and myoepithelial cells are positive for p63 (Figure 4).
4. *Histological grading:* It is usually well circumscribed and with size ranging from 1–5 cm in [21]. According to previous workers, breast ACC can be graded on the basis of presence of solid areas in tumor microscopy and it was directly affecting the prognosis of the patients [22]. Proposed Grades were grade 1 (no evidence of solid areas); grade 2 (< 30% solid proportion); grade 3 (>30% solid areas) and the proposed treatment were local excision, simple mastectomy and mastectomy with axillary node dissection respectively.
5. *Mammography:* Although calcification may develop in these tumors detected rarely on mammography. In one series only, 4 of 22 patients detected radiologically [23].
6. *Metastasis:* Axillary lymph node metastases are rare in patients with ACC. Arpino et al. noted lymph node metastases in only 4 of the 182 cases collected from literature [4]. Distant metastases are uncommon with only 14 cases having been reported, but when they occur they tend to do so without prior lymph node involvement. Hence a routine axillary lymph node dissection is not recommended [4].
7. *Estrogen receptor/progesterone receptor:* ACC is generally estrogen receptor (ER) negative; in various series the ER was described as positive

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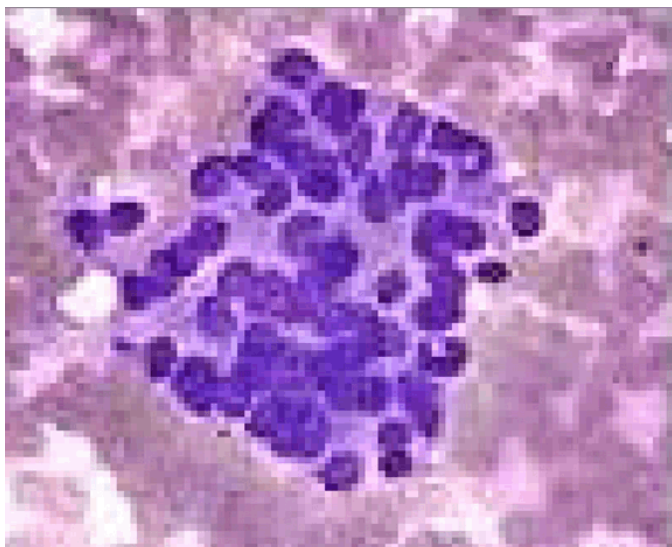


Figure 1: Fine needle aspiration cytology (FNAC) showing loosely cohesive ductal atypical epithelial cells atypical epithelial hyperplasia.

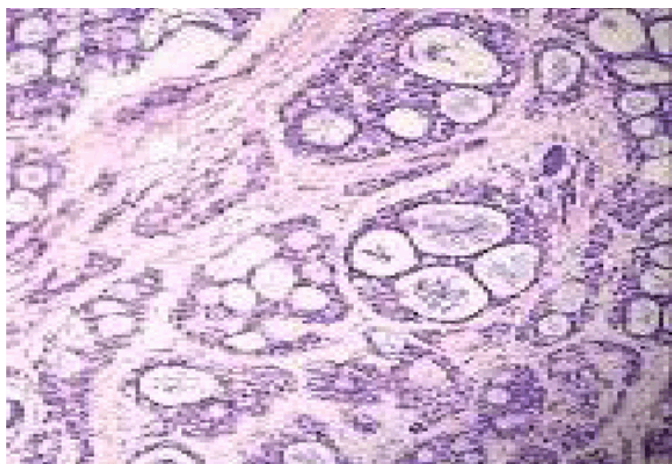


Figure 2: Prominent solid, microcystic, and focally tubuloglandular patterns of growth with intact myoepithelial layer (x10).

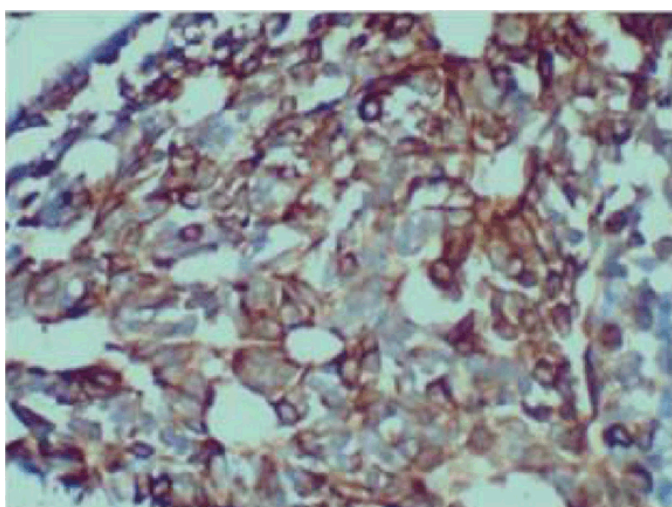


Figure 3: Tumor cells showing cytoplasmic positivity for CK7.

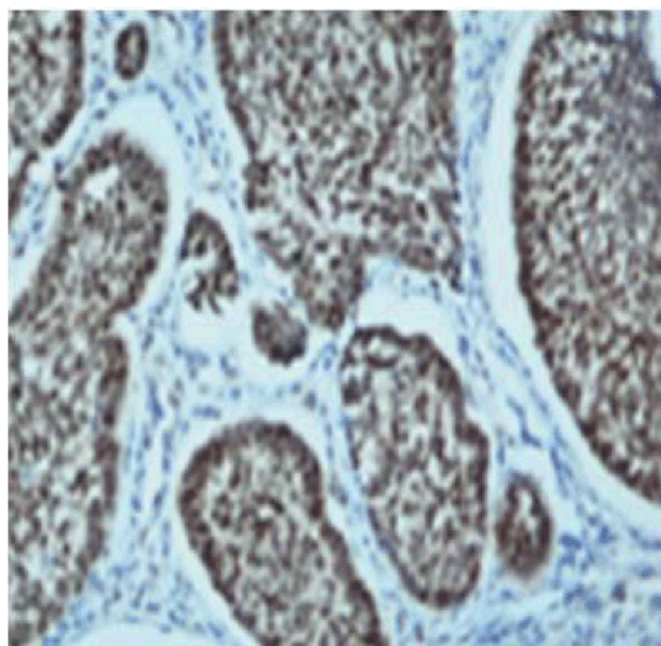


Figure 4: The p63 expression by myoepithelial cells.

in 0.7–28% of the cases [23, 24]. Progesterone receptor (PR) was positive in three of the 13 evaluated patients [23]. Hence, hormonal manipulation is of limited use as most cases are ER.

Treatment and Prognosis

Adenoid cystic carcinoma of the breast in females usually has very favorable biological characteristics for treatment [25–28]. It belongs to the basal-like subgroup of breast cancers [29]. Based on extensive molecular and genetic profiling studies, basal-like tumors are most often hormone receptor (ER and PR) negative and do not express human epidermal growth factor receptor 2 (HER2) [30, 31]. Adenoid cystic carcinoma has the excellent prognosis irrespective of IHC results [28]. The nodal and distant metastasis is lower as per existing literature [32]. It is because of these distinct clinic-pathologic features that set it apart from the other triple-negative breast cancers [33].

Surgical intervention is the basic treatment for ACC. The surgical treatment modalities include lumpectomy, wide excision, or MRM (modified radical mastectomy). The axillary clearance is not needed in majority of the patients. Lumpectomy with radiation or simple mastectomy has excellent results.

No clear guidelines are available for adjuvant chemotherapy or radiotherapy. According to previous studies (2011 St. Gallen International Expert Consensus), there is no need of adjuvant chemotherapy for ACC with absence of nodal metastasis. Radiotherapy is advised

in patients with treatment plan aiming conservation of breast or in patients with large tumor size with nodal metastasis [33]. Hormonal therapy or anti-HER2 treatment is rarely indicated as ACC usually does not express hormonal receptors and HER2/neu.

PROGNOSIS

Usually, the prognosis is excellent as compared to other breast malignancies and ACC of salivary glands [28, 34].

CONCLUSION

Adenoid cystic carcinoma of breast is a rare histological diagnosis. Early fine needle aspiration cytology/core biopsy and meticulous histopathological examination is mandatory in each and every palpable breast lump irrespective of age and clinical presentation to prevent untoward complications related to disease and treatment for the sake of accurate pathological diagnosis.

Keywords: Adenoid cystic carcinoma, Breast, Diagnosis and management

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S. B. Ingle – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

C. R. Hinge Ingle – Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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