CASE REPORT

Adenoid Cystic Carcinoma of the Breast: A Rare Case Report

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Abstract:

Adenoid cystic carcinoma is an extremely rare subtype of breast carcinoma. Usually they are triple negative, but have favourable prognosis in contrast to their salivary gland counterparts. They have distinct morphological features, the solid component determining the grade of the tumour. They have low recurrence rate, may rarely metastasize after a long time after diagnosis, hence requiring a long term follow up. There is no consensus on the effective treatment. Breast conserving surgery with adjuvant radiotherapy appears to be effective. It is hence important to identify this distinct subtype of breast carcinoma.

Keywords: Breast Carcinoma, Adenoid Cystic Carcinoma, Prognosis

Introduction:

Adenoid Cystic Carcinoma (ACC) is a tumour, the occurrence of which is common in salivary glands, but can also occur in other sites such as nasopharynx, trachea, lungs, skin, kidney uterine cervix and breast [1]. Adenoid Cystic Carcinoma of Breast (ACCB) is a salivary gland type breast carcinoma and is an extremely rare subtype. Though, they are triple negative and show basaloid morphology, they are low grade tumours with indolent clinical course. They constitute less than 0.1% of breast cancer [2]. Though they resemble the ACC arising in salivary glandsin morphology, they have better prognosis. It is important to recognise and report this distinctive subtype as they have distinct clinical and morphological features and have excellent prognosis unlike other triple negative breast carcinomas which are known to have poor prognosis.

Case Report:

A 75 year old female patient presented with right breast lump which was present for the past 2 years. She had noticed increase in the size of lump in the last 3 months period. On examination, a lump measuring 3 × 2 cm, was present in the subareolar region of the right breast. Excision biopsy was done to rule out carcinoma. Excised mass was grey tan to pink, solid and showed well defined margins. Focal tiny cystic areas were also seen (Fig. 1).

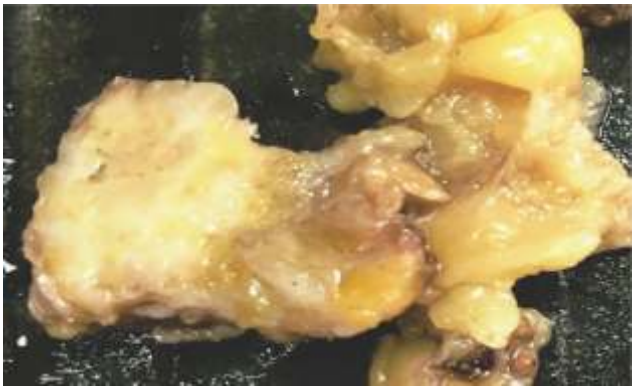

Fig. 1: Cut Surface of the Lump, Grey Tan to Pink, Solid with Well Circumscribed Margins

Microscopic examination revealed a neoplasm with cells exhibiting cribriform and solid pattern, glandular spaces containing basophilic secretion. The solid pattern was seen in <30% of the tumour tissue. The cells showed basaloid morphology. The tumour showed true glandular lumina as well as pseudo lumina containing eosinophilic material constituting cylindromatous component (Figs. 2 and 3). Invasion of the tumour into stroma and adipose tissue was seen. The surgical margins were however free of tumour. A diagnosis of ACCB, grade II was given. Immunohistochemical marker study was done. The tumour cells were (PR) negative, Her2 neu negative, with focal (ER) positivity. The tumour cells were positive forCD 117 (Fig. 4). Ki 67 score was 15%. The patient underwent modified radical mastectomy. There was no residual tumour, except for a tiny tumour embolus at the margin of the resection area. There was no evidence of lymph node metastasis. There was no recurrence during the follow up period of 1 year.

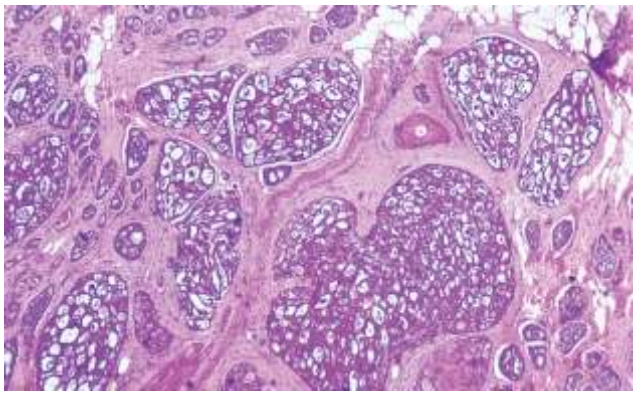

Fig. 2: Tumour Showing Predominantly Cribriform Pattern with Basophilic Secretions in their Lumina, Tumour Cells Appear Basaloid (× 100; H&E Stain)

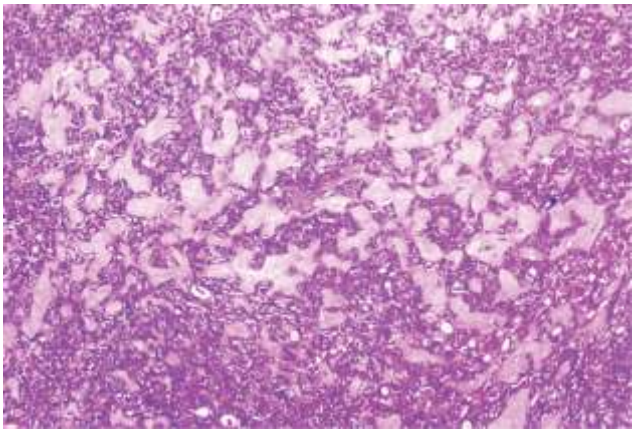

Fig. 3: Stroma within the Tumour Show Eosinophilic Material, the Cylindromatous Component (× 100; H&E Stain)

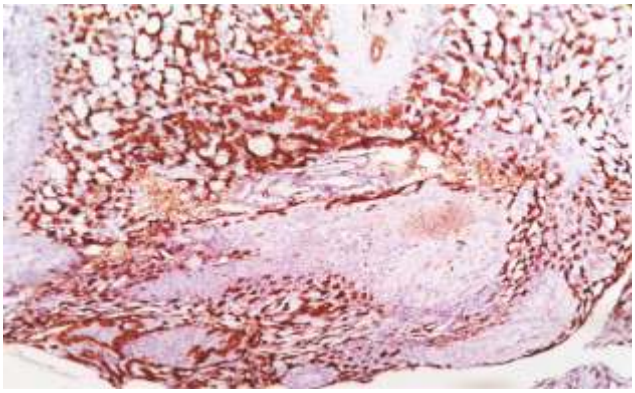

Fig. 4: Tumour Cells Exhibiting Reactivity to CD 117 (× 100; IHC Stain)

Discussion:

ACCB was first termed as 'cylindroma' by Billroth in 1856, later in 1945 was described by Geschickter [3-4]. These tumours are common in females, may also be seen in young men, mean age of presentation being 64 years (range: 25-80 years), 50% of them situated in the subareolar region [5].

Most of them occur as solitary lesions. The radiographic findings are nonspecific. A study by Khanfir *et al*. [6] detected only 18% of the cases on mammography. They appeared as irregular, microlobulated with uneven density, calcification being a rare finding [7]. Grossly, they are well circumscribed lesions, with pink-tan appearance and may be associated with micro cyst formation. They resemble the ACC of salivary glands with presence of dual population of cells, including luminal and myoepithelial basal cells, exhibit various patterns of architecture such as tubular, cribriform, trabecular, solid and basaloid [8]. In addition to true glandular lumina, they also show pseudolumina, may exhibit squamous and sebaceous differentiation. Two types of cells show preserved cell polarity and separate cell differentiation. It was proposed that this could be the reason for low grade, non-aggressive clinical behaviour of this tumour with reduced propensity for metastasis [9-10]. Histologic grading of ACCB is done based on the proportion of solid growth pattern. No solid pattern- G I, <30% - G II and >30% G III. Association of recurrence and metastasis was observed in G II and G III tumours. Solid pattern with prominent basaloid features is found to be associated with larger size, recurrence, metastasis and aggressive clinical course [11-13]. ACCB are usually triple negative, with absence of expression of ER, PR, and Her2 neu. Weak/focal ER positivity in some cases may be attributed to the positivity in normal breast lobules and ducts trapped within the tumour [14]. It is observed that ACCB can be associated with other lesions of the breast such as fibroadenoma, microglandular adenosis, invasive ductal carcinoma NOS [15-17].

Differential Diagnoses:

Differentiating ACC from Cribriform or tubular carcinoma, especially in core needle biopsiescan be difficult. Presence of dual cell population, ER, PR negative status, CD 117/c-Kit and / p63 reactivity of ACC can be of help in such cases. Collagenous spherulosis must be differentiated from ACC. Absence of mucosubstance in the lumina, lesion being at periphery and irregular margins, CD 117 negative expression favour collagenous spherulosis.

It is also important to differentiate solid variant of ACCB from small cell carcinoma (neuroendocrine carcinoma), solid papillary carcinoma and malignant lymphoma where immunohistochemical markers play an important role [13]. Molecular genetic studies have revealed an interesting fact that, though ACCB are triple negative, show a basal like phenotype, they differ from other basal like invasive breast carcinomas. They exhibit simpler patterns of gene copy number aberrations [18], they are low grade and show low frequency of genetic instability [19], aneuploidy [20] and exhibit an indolent clinical behaviour.

ACCB are morphologically similar to those of salivary gland. But, molecular studies revealed, decreased expression of leb-7b and overexpression of mik-24 in ACCB, in contrast to ACC of salivary gland. However, both are characterised by t(6;9)q22-23; p23-24 chromosome translocation which generated fusion gene MYB-NFIB, with overexpression of oncogene MYB [14,21].

When compared to Invasive ductal carcinoma NOS type, ACCB have low recurrence rate and favourable prognosis [22]. In contrast to their counterparts in salivary glands, they have

excellent prognosis. Axillary lymph node metastasis is rare (<2%). They are rarely aggressive and may show distant metastasis to lung, kidney, brain and bone [23-24]. Distant metastasis can occur without prior involvement of axillary lymph nodes [18]. Metastasis may be seen many years after the diagnosis of primary disease. Hence long term follow-up is important.

Mastectomy with axillary lymph node dissection in patients with positive sentinel node or Breast conserving treatment with adjuvant radiotherapy appears to be effective in terms of survival. After definitive treatment, ten-year survival rate is 86-95% [1].

Conclusion:

ACCB is a tumour with distinct morphology, excellent prognosis, low recurrence rate and may require long term follow-up, Hence, it is important to identify this distinct tumour.

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