Case Report

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Adenoid cystic carcinoma – A rare triple negative breast carcioma with excellent clincial outcome

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Abstract:
A 65 year old female presented to Osmania General Hospital with a history of swelling in the right breast since 3 months. On palpation, the swelling was occupying the upper inner and outer quadrants. Clinically,a diagnosis of phyllodes tumor was made. Ultrasonography of breast swelling revealed a breast abscess. Fine needle aspiration cytology was attempted and a diagnosis of cystic lesion of breast was made. Simple mastectomy was carried out. Histopathological examination revealed a cystic tumor comprising of a cribriform pattern. Special stains and immunohistochemistry was helpful in final diagnosis of tumor as adenoid cystic carcinoma of breast.

Keywords: Adenoid cystic carcinoma, carcinoma of the breast, carcinoma, breast.

Introduction:

Adenoid cystic carcinoma (ACC) is a rare malignant tumor of the breast. They are most frequently noted in the salivary glands but are also common in the uterine cervix, skin, lungs, kidneys, esophagus and prostate [1]. These tumors occur in <0.1% of all patients diagnosed with breast cancer. The age distribution is from 19–97 years, and the condition is more common in the 50 to 60-year-age group. Typically, the tumors present as a sub areolar mass or as pain in the breast. The involvement of lymph nodes and distant metastasis are extremely rare.ACC of the breast shares the same histological characteristics with ACC of the salivary gland [2].

The prognosis of ACC of the breast is better in comparison to other pathological types of breast cancer and ACC of the salivary gland. High survival rates following mastectomy or breast protective surgery have been reported. ACC of the breast has a biphasic pattern. Histologically, it consists of small basaloid cells with a solid cribriform pattern or epithelial cells with a tubular growth pattern. Although the exact origins remain unknown, it is estimated that these tumors originate from the ductal epithelium or myoepithelium [3]. The presence of estrogen and progesterone receptors tends to be negative in these tumors [4]. ACC has an excellent prognosis, a low local recurrence and rare distant metastasis.

Case details:

A 65-year-old postmenopausal patient was admitted in department of general surgery (Osmania General Hospital, Hyderabad) with the complaints of swelling in the right breast since 3 months. The breast examination revealed a lump in the upper inner& outer quadrants, firm to cystic in consistency mobile. Clinically a diagnosis of phyllodestumor was made. On Ultrasonography a diagnosis of breast abscess was made. Fine needle aspiration cytology was attemptedon the cysticlesion of the breast and Simple mastectomy was carried out.

Gross: we received right Simple mastectomy specimen measuring 7×3×4cm. Nipple and areola grossly appear to be normal.CutSection showed a large cyst measuring 5×4cm and the Cyst wall thickness measured 0.5cm.(Fig1)

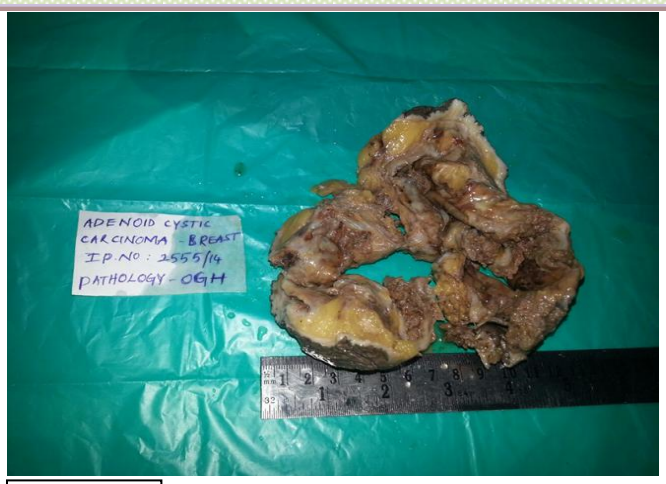

Figure 1

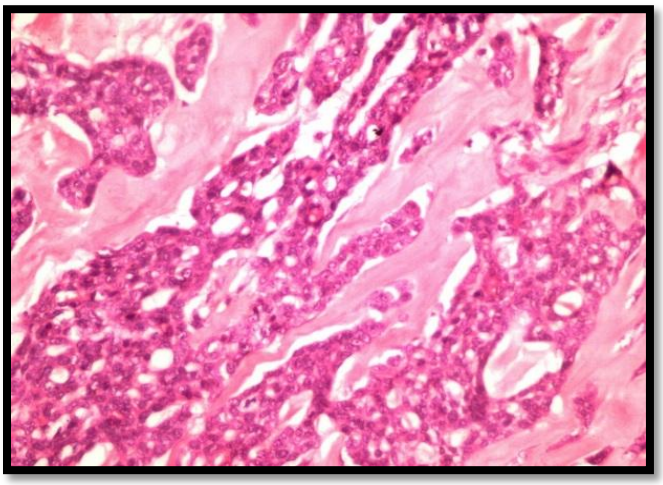

Figure 2b -40x

Fig 3:3a.P63-nuclear positivity inabluminal cells.3b. CD117-Cell membrane positivity in luminal cells 3c. SMA-Cytoplasmic positivity in abluminal cells3d.ERNegative.

Microscopy:

Multiple sections studied from simple mastectomy specimen revealed cystic breast tumour arranged in cribriform, psuedoglandular& tubular patterns. Biphasic cell pattern comprising of epithelial and myoepithelial cell components. The luminal spaces contained basophilic basement membrane like material (mucin).The tumour was seen to infiltrate the stroma which was variably desmoplastic. No necrosis& occasional mitotic activity was noted. Skin, nipple and areola were free from tumour. Tumour was infiltrating upto the deep resected margin.

Fig 2a, 2b shows cribriform, tubular pattern (H&E stain)

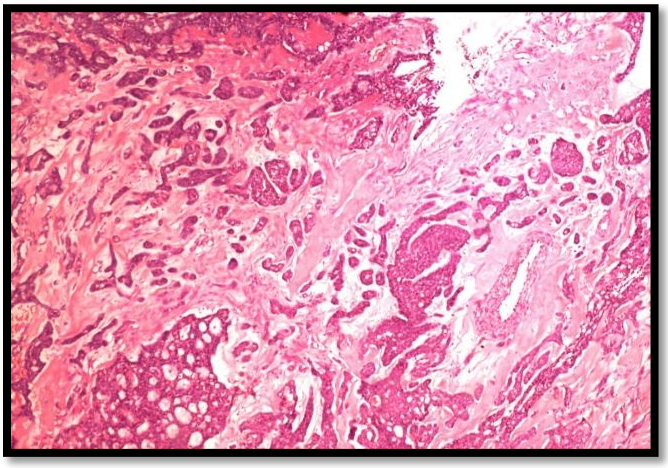

Figure 2a -10x

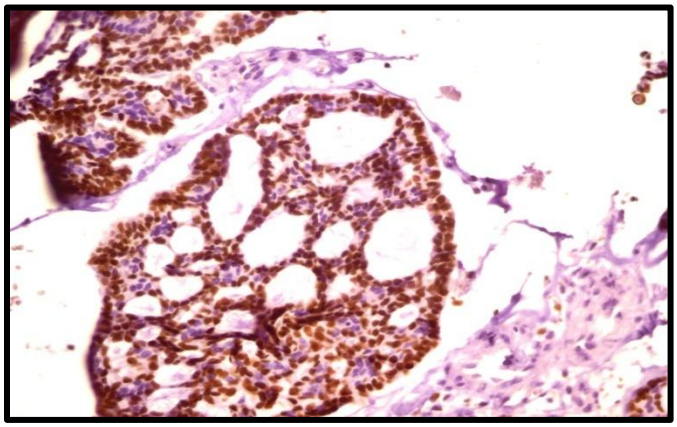

Fig 3a-P63

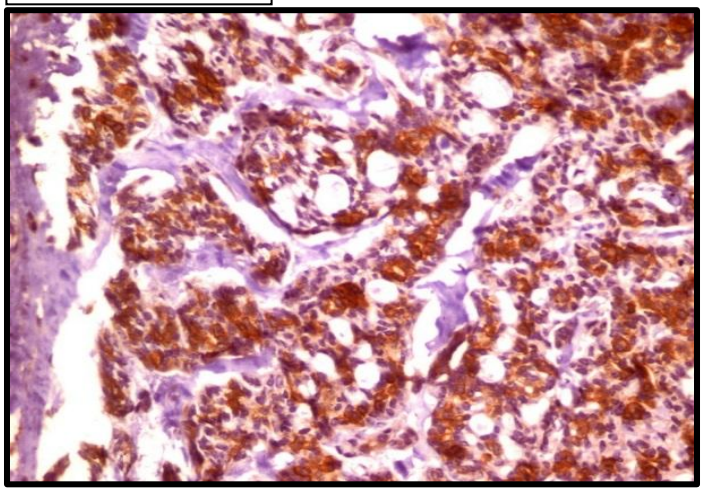

Fig 3b- CD117

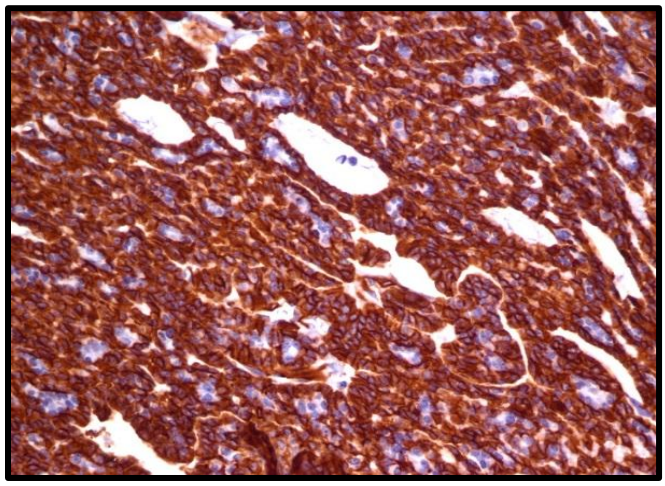

Fig 3c-SMA

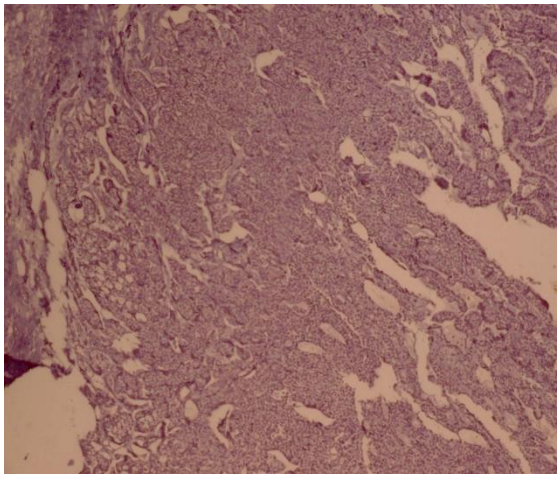

Fig 3d-ER

Discussion:

ACC occurs in one in 1 million females every year and was initially described as a cylindroma by Billroth in 1856. Breast ACC was first described by Geschickter in 1945 [5]. ACC has three varied growth patterns: Glandular, Tubular and Solid. The grade of the tumor increases with an increased rate of solid element [6].

Grade 1. Numerous glands and Cystic components, without solid components-require local excision. Grade 2<30% solid components-require simple mastectomy and Grade 3. >30% solid components-requires mastectomy and axillary dissection [7]. In our case, the patient underwent simple mastectomy and the tumour showed cribriform,psuedoglandular & tubular pattern.

ACCs are commonly detected in postmenopausal females and in the geriatric population. The left and right breasts are equally affected and there is no tendency for the occurrence to be bilateral. Although the tumor size is usually 2–3 cm, in our case it was 4-5 cm. The most frequent symptoms at presentation include the finding of a well-circumscribed palpable mass, and pain in the breast. Perineural invasion is extremely rare in ACC of the breast, which is common in ACC of salivary glands believed to be the cause for pain in tumour [8]. Our patient complained of breast mass without pain.

The nipple and the areola in these cases appears to be normal. Mammography generally reveals a well-circumscribed lobulated mass that presents with extremely rare microcalcifications.These are associated with hypoechoic lesions on ultrasonography.

ACC is generally negative for the estrogen, progesterone receptors and Cerb-B2 (Her 2 Neu).P-63 is positive inabluminalcells. In the present case the tumor was ER, PR&HER-2neu immunonegative. There is no consensus regarding the optimal treatment of ACCs of the breast due to the rare occurrence of these tumors. Surgical approaches range from local excision to mastectomy [9]. Recurrence rates ranging from 6–37% were reported following local excision. Since extremely few recurrences were reported following mastectomy, numerous clinicians recommend mastectomy for a diagnosis of ACC of the breast. The role of radiotherapy following breast conservation surgery remains unclear. Data on the role of radiotherapy in female breast ACCs are limited. Axillary lymph node involvement is extremely rare, occurring in an average of 0–2% of ACCs of the breast. Axillary dissection should not be clinically performed except in the presence of nodal metastasis [10]. Additionally, sentinel lymph node sampling can be performed if the tumor is >3 cm, has a high grade or contains additionally other invasive types of breast cancer. The role of adjuvant therapy and hormonal therapy is controversial in patients with ACC of the breast [11]. The 5-year survival rate for ACCs of the breast is reported to be 85–90%, with a 100% disease-free survival rate. Despite these results, ACC is reported to present with local recurrences and distant metastases. The most common site for distant metastasis is the lung followed by the liver, kidneys and brain [12].

Conclusion:

Adenoid cystic carcinoma of the breast are extremely rare neoplasms of the breast and have an extremely good prognosis. Although no consensus exists regarding the optimal treatment, breast

conservative surgery and radiotherapy are recommended. The affected patients require close follow-up due to the rare, but possible, occurrence of distant metastasis. The present case has not reported for any complications on recent follow up.

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