



ADENOID CYSTIC CARCINOMA OF FEMALE BREAST-AN INTERESTING PATHOLOGICAL DIAGNOSIS

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Abstract: Adenoid cystic carcinoma (ACC) of the breast is a very rare and indolent subtype of malignant breast tumors, accounting for less than 0.1% of all primary breast cancers, occurring in approximately 1 out of 1 million women each year. To date, there have been about 933 cases reported as per English literature. A 50-year old female presented with a right breast mass. Sono mammography revealed a large, well defined, solid appearing, lobulated mass. On fine needle aspiration, a differential diagnosis of metaplastic carcinoma, phylloides tumor, papillary neoplasm and primary chondrosarcoma of the breast was made and biopsy was advised. Partial mastectomy was done and a histopathological diagnosis of Adenoid Cystic Carcinoma of the breast was made. ACC is of special interest because of its favourable prognosis and distinctive histological appearance. Specifically, they are negative for ER, PR, and HER2-neu, yet on microarray genomic analysis, ACC is distinct from triple negative breast cancer. This tumor occurs predominantly in women in their sixth decade and usually presents as a tender breast mass, often in the subareolar area. The optimal treatment of ACC of the breast has not been defined, although the mainstay of treatment is surgery. Both partial mastectomy and modified radical mastectomy have been used, and the role of adjuvant radiation therapy is not clear.

Key Words: Adenoid cystic; breast; cylindroma

INTRODUCTION

Adenoid cystic carcinoma (ACC) of the breast is a very rare malignancy, the prevalence being less than 0.1% of all breast neoplasms¹. ACC of the breast is interchangeably called “cylindroma”. It was first described by Geschickter and Copeland in 1945². ACC affects mainly post-menopausal women; however, isolated cases have been identified in men and children. Most of the cases of ACC arising in the breast have been localized disease with a low rate of axillary lymph node involvement and distant metastasis. They have a better prognosis compared with other variants of breast cancer and ACC of the salivary glands.³ No specific guidelines for treatment have been established, due to the rarity of the tumor and large variations in the patterns of practice.⁴ Here we present a noteworthy case of ACC breast in a 50 year old female, the diagnosis of which was made on histopathologic examination and it presented with a distinct morphology on cytological examination.

MATERIALS AND METHODS

A 50 year old female presented with a painless mass in the right breast since 6 months. The mass was gradually increasing in size. On physical examination, a firm, well circumscribed, non-tender mass, measuring 5cm in maximum diameter was palpated in the upper inner quadrant of the right breast. The overlying skin, nipple and areola appeared normal.

Sonammammography revealed a large, well defined, hypoechoic, solid appearing, lobulated mass, measuring approximately 49mmx47mmx54mm in maximum dimensions with volume of about 65cc in the supero medial quadrant of the right breast. Thick capsule with mild distal enhancement was seen. Routine investigations of blood and urine were normal. No significant past history of prior breast disease or any medications or surgery and no family history of similar complaint.

On fine needle aspiration, blood mixed material was obtained. Smears revealed atypical cells arranged in a papillary manner intermixed with chondromyxoid material and hemorrhage. These cells were pleomorphic with opened up chromatin and moderate amount of basophilic cytoplasm, as seen in figure 1,2,3. A differential diagnosis of metaplastic carcinoma, phylloides tumor and papillary neoplasm of the breast was made and biopsy was advised.

RESULTS

Mastectomy was done and a breast mass measuring 15x8x6cm was received for histopathologic examination. Skin ellipse measuring 12x6cm was present. Nipple areola complex was unremarkable. Cut section showed a tumor, measuring 5x5x5cm, 9cm lateral to nipple areola. The cut surface of the tumor has a variegated appearance, greyish white to greyish brown in colour, with areas of necrosis, hemorrhage and few cystic areas. Microscopy showed tumor having a thick walled capsule, with the tumor cells arranged in lobules, suspended in fibrous tissue. These lobules showed glandular proliferation and mucin filled small cystic areas, as in figure 4,5. The tumor cells were pleomorphic with opened up nuclear chromatin and prominent nucleoli. A histopathological diagnosis of ACC breast was made.

DISCUSSION

The term “Cylindroma” was proposed by ‘Billiroth’ who concluded that the tumor was composed of intertwined cylinders of stroma and epithelial cells. Ewing first mentioned Adenoid Cystic Carcinoma of the salivary glands and this term was applied to breast tumors by Geschickter in 1945. 933 cases of ACC have been described as per literature up till now⁵.

ACC occurs in adult women, the mean age distribution being 25-80 years of age⁶. The usual presentation is as a palpable, discrete and firm mass. Grossly, tumors with low grade features on histology tend

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to be smaller, mean size being 1.6cms, compared to high grade tumors, an average of 3.5cms in size. Mostly, ACC are circumscribed or nodular grossly although an invasive growth pattern is seen microscopically. Microcystic areas are seen in about 25% cases. Perineural invasion and lymphatic tumor emboli are extremely uncommon.

ACC shows a proliferation of both, glandular (Adenoid Component) and basement membrane (“pseudoglandular”) components.

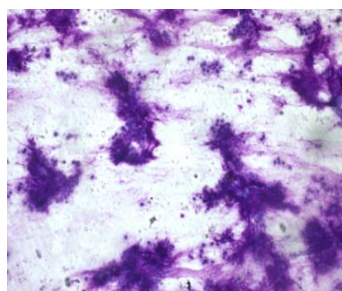


Fig.1: Hematoxylin & Eosin:10x

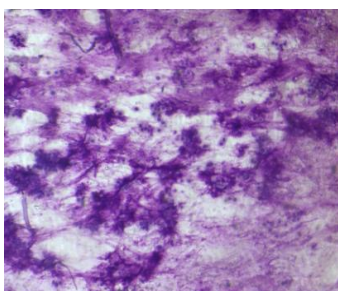


Fig.2: Hematoxylin & Eosin :10x

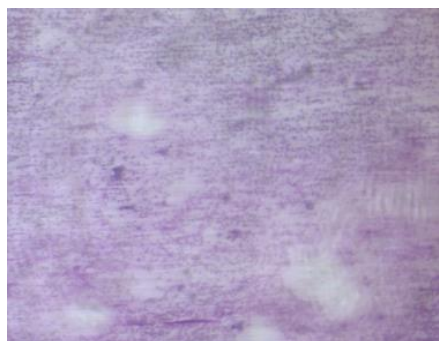


Fig.3: Hematoxylin & Eosin:10x

Figure 1,2,3: H&E stained FNA Smears - showing atypical cells arranged in a papillary manner intermixed with chondromyxoid material and hemorrhage (10x).

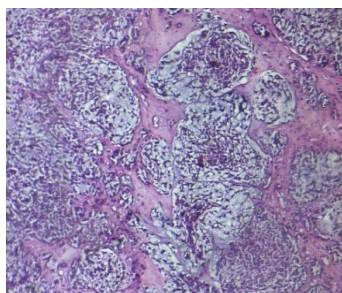


Fig.4: hematoxylin& Eosin:10x

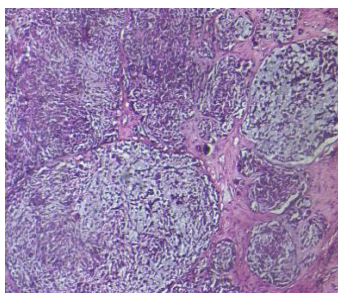


Fig.5: Hematoxylin & Eosin:10x

Figure 4, 5: H&E stained smears of histopathology: 10x- show tumor cells arranged in lobules, suspended in fibrous tissue. These lobules show glandular proliferation and mucin filled small cystic areas (10x).

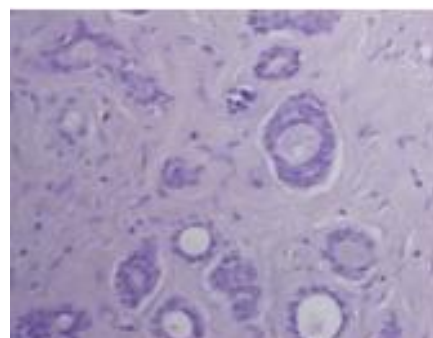


Fig.6: Immunohistochemical findings in adenoid cystic carcinoma of the breast: Estrogen receptor: negative. Original magnification × 100.

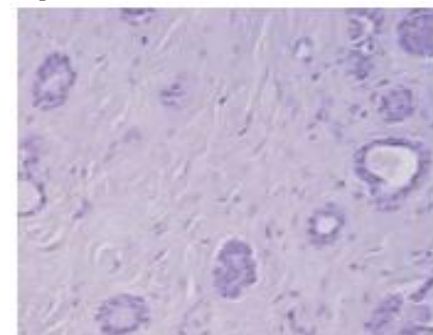


Figure 7: Immunohistochemical findings in adenoid cystic carcinoma of the breast, Progesterone receptor

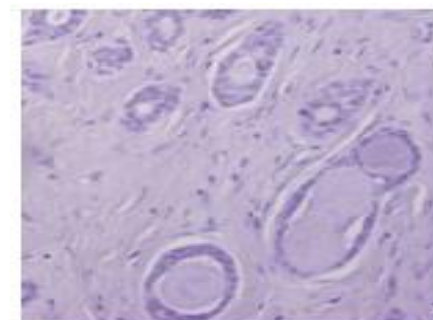


Figure 8: Immunohistochemical findings in adenoid cystic carcinoma of the breast, Human epidermal growth factor receptor: negative. Original magnification × 100.

If adenoid glandular clusters and nodules of cylindromatous material are present, a cytological diagnosis of ACC can be made. In the present case, a differential diagnosis of metaplastic carcinoma, phylloides tumor and papillary neoplasm of the breast was made on cytological examination.

On histopathologic examination, various microscopic patterns have been described as cribriform, solid, tubular, reticular and basaloid. Depending on proportion of solid growth within the lesion, three grades have been described⁷. Grade I comprises of cases with no solid elements, grade II includes cases with less than 30% solid elements and grade III cases comprise of more than 30% solid component. Other types of carcinoma can be present along with ACC, although in this case, no other lesion was present.

More than 90% of ACC examined by Immunohistochemistry were reported to be negative for estrogen and progesterone receptors and HER2-neu, as in this case, seen in fig.6,7,8 yet on microarray genomic analysis, ACC is distinct from triple negative breast cancer.

The prognosis of patients treated surgically by mastectomy has been curative in majority of cases, according to various studies. The 10-year survival rate for patients with ACCs of the breast has ranged from 85% to 100%^{8,9}. Patients with metastasis generally have pulmonary involvement and negative axillary lymph nodes. Recurrence in the breast has been reported after local excision alone. Mostly ACC are grossly circumscribed, as in this case but 50% tumors can extend microscopically into the surrounding tissue.

The treatment modalities of ACC vary widely from case to case and practice trends and include wide excision or quadrantectomy, radiotherapy and mastectomy. Axillary dissection is indicated if nodal involvement is suspected; systemic adjuvant chemotherapy also plays a role in high grade lesions.

CONCLUSION

ACC of female breast is therefore of special interest due to its rarity, favorable prognosis and distinctive histological appearance. Most information on ACC of the breast is derived from individual case reports and clinical case series. Treatment guidelines have not been established due to the rarity of the tumor and large variations in the patterns of practice. The radiologic appearance is generally nonspecific, and little has been published on the mammographic and ultrasonographic features of ACCs. The histopathologic features of ACC of the breast are similar to those of ACC originating from salivary glands and are distinct from those of ductal and lobular carcinomas of the breast. To conclude, a thorough history and physical examination, followed by vigilant screening of breast masses, coupled with mammography, fine needle aspiration cytology, histopathology and immunohistochemistry is imperative for detecting ACC and improving patient care.

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