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A rare case of teenage adenoid cystic carcinoma of the breast

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ABSTRACT

Adenoid cystic carcinoma (ACC) of the breast is very rare, accounting for only 0.1% of all breast carcinomas. Their rarity has made them difficult to study systematically with the most published series consisting of few cases or case reports; although, this existing evidence does suggest a good prognosis. ACC usually affects women, more in the sixth decade of life. However, our case has an exceptionally early onset of cancer at 16 years, the youngest ever reported in literature. Good local control can be obtained with lumpectomy and radiation or simple mastectomy for nearly all tumors. A routine axillary lymph node dissection is not recommended. We gave adjuvant radiotherapy to this patient in view of close margins and presence of lymphovascular emboli. She tolerated well with minimal side effects. Chemotherapy and hormonal therapy have no established benefit.

Keywords: Adenoid cystic carcinoma (ACC), adjuvant radiotherapy, modified radical mastectomy.

Adenoid cystic carcinoma (ACC) occurs in 0.1-1% of the malignant breast carcinomas and is of special interest because of its favorable prognosis and distinctive histological appearance [1-2]. It tends to occur mostly in postmenopausal women; more in the 5th to 6th decade of life. However, our case is neither postmenopausal nor in her fifties [3-4]. She is only 16 at the time of presentation and premenopausal, probably, the youngest ever reported in literature. This unusual age of onset prompted us to report this case.

CASE HISTORY

A sixteen year old girl presented with a history of lump in the right breast just behind the nipple-areolar complex which was associated with intermittent pain. She gave a past history of injury to the right breast 3 years back. She attained menarche at 15 years of age, had irregular cycles (3days/ 2 months). Family history was negative for malignancies. On clinical examination of the right breast, a single 3*3cm sub-areolar mass was palpated, hard to firm in consistency with restricted mobility. Left breast and bilateral axilla were normal. Systemic examination was also unremarkable.

She underwent a core needle biopsy and the histopathology report revealed that it was adenoid cystic carcinoma grade 2. She underwent Right modified radical mastectomy with level III axillary adenectomy. Reexcision was not possible for this patient. Tumor measured 4.5 cm in the greatest dimension. Nipple-areolar complex was involved by the tumor. Posterior resected margin is <2mm from tumor. Other margins are free from tumor. Lymphovascular emboli were present. The tumor infiltration with dermis is seen. Lymph nodes were negative for tumor metastasis. Immunohistochemistry

(IHC) was performed for the diagnosis and prognostication. Immunohistochemical markers like ER, PR and HER2/neu were negative. However, IHC marker CD117 (c-kit) was positive in tumour cells and consistent with ACC (**Fig. 1**).

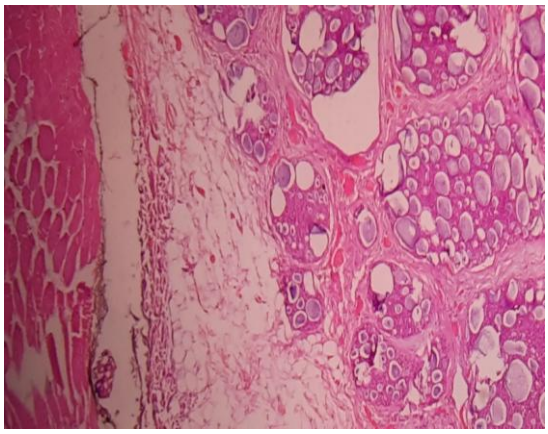

Figure 1 - Tumor composed of cells with scant cytoplasm and hyperchromatic nuclei in a cribriform tubular and pseudoacinar pattern along with luminal myxoid basement membrane material suggestive of adenoid cystic carcinoma

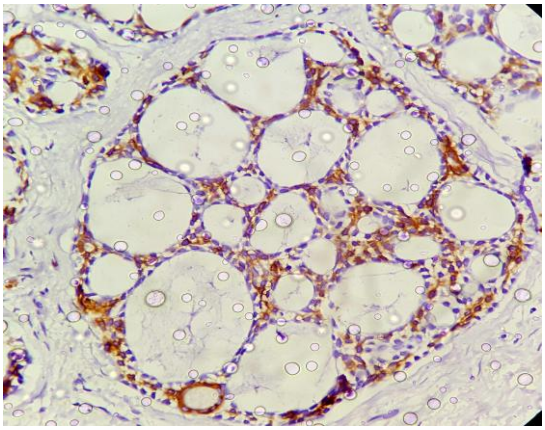

Figure 2 - IHC with CD117- Strongly positive in tumor cells

She underwent adjuvant radiation in view of close margins (<2mm) and presence of lymphovascular space invasion. She received 50Gy in 25 fractions to the chest wall alone using two tangential fields on a linear accelerator. She tolerated radiotherapy well with acceptable acute toxicities like grade I dermatitis with hyperpigmentation, grade I fatigue. On 6 month follow up, she was asymptomatic and physical examination was normal.

DISCUSSION

In recent years, the term "adenoid cystic carcinoma" has gained acceptance as the one preferred for the unusual malignant epithelial tumors that Billroth first described as "cylindroma" in 1856. Though quite uncommon, they may arise in a variety of anatomical sites including the paired salivary glands, lacrimal gland, mucous glands of the upper respiratory and digestive tracts, skin and the breast. In fact, ACCs of the salivary gland are characterized as aggressive tumors with a poor prognosis distinctly converse to the indolent behaviour and good prognosis associated with ACCs of the breast.

ACC occurs predominantly in women in their sixth decade but has been described between the ages of 30 to 90. However; our case may be the youngest reported in literature. The clinical presentation is usually a dominant subareolar breast mass tender to palpation, [5-6] as seen in our case. Most of the ACC are well circumscribed and firm. The diagnosis can be made on fine needle aspiration cytology. Histologically, ACC has a unique distinctive biphasic pattern that consists of true laminate and pseudo-cystic spaces, true glands are lined by epithelial cells and pseudo cysts are lined by myoepithelial cells [2,7]. Ro et al suggested that ACC of the breast can be graded on the proportion of solid growth of the tumor and this was correlated with prognosis [8]. Grades proposed were 1 (no solid element); grade 2 (< 30 % solid element); grade 3 (>30% solid element). The proposed treatment was local excision for grade 1 tumors, simple mastectomy for grade 2 tumors and mastectomy with axillary node dissection for grade 3 tumors.

The best surgical treatment for ACC of the breast has not been established. Local excision is followed by unacceptably high rates of recurrence [8]. The studies showed that ACC has low proliferation activity which may explain the low recurrence rate. Based on these findings simple mastectomy or lumpectomy followed by radiation treatment is thought to have a chance to achieve adequate local control of nearly all tumors [3,5]. Axillary lymph node dissection is rarely required because of the low incidence of spread to the axillary lymph nodes. The role of chemotherapy and hormonal manipulation is yet to be established. Addition of radiation to post mastectomy patient is rarely done; however, we did it in view of close margins and presence of lymphovascular emboli.

Because of its rarity, clinical trials comparing treatment options for ACC are needed to define the optimal treatment. Clinical studies suggest that breast ACC is associated with a favourable survival; thus, belying the expected poor prognosis suggested by the absence of hormone receptor expression [7,9]. Five-year relative survival was excellent among women aged <50 years (94.4%) and ≥50 years (99.0%) [4]. Regular long term follow up with chest x-ray and thorough physical examination is essential as there have reports of late onset of local relapse, as well as distant metastasis [10].

CONCLUSION

Adenoid cystic carcinoma of the breast is a special, oestrogen receptor, progesterone receptor, HER-2 negative and highly KIT-positive, basal-like breast carcinoma, associated with an excellent prognosis. Surgery is the main stay of treatment. Role of radiotherapy in breast conservation surgery has shown benefit in terms local control. A chest x-ray and thorough physical examination looking for local recurrence is all that is needed for follow-up.

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