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Case Review

Primary Apocrine Carcinoma of the Axilla: A Case Report, Review of Literature and Treatment Recommendation

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Introduction

Apocrine carcinoma (AC) of the sweat gland is an extremely rare neoplasm with around 50 cases reported in the literature so far [2,3]. It is usually a slow growing tumour, giving the clinical picture of a benign lesion but can sometimes progress aggressively to a malignant lesion. Clinically, it often presents as painless indurated nodule or plaque. Mostly it affects the apocrine dense regions like axilla and anogenital region but it can also affect the scalp, eyelid, ear, lip, chest, nipple, fingers, and toes. Apocrine carcinoma frequently affects the elderly population with a median age of 67 but can occur at any age [2,3,5]. There is no racial or gender predisposition noted [4]. A cytological diagnosis followed by histopathology is necessary for confirmation [10].

The incidence of primary AC is quite low at 0.0049 to 0.0173 cases/100,000 persons per annum [2]. Most AC are generally characterized by slow symptomatic progression and a slow growth rate and are not clinically suspected until a biopsy is done. Approximately 50% of the patients diagnosed with AC so far, had lymph node metastases at the time of diagnosis [9]. Only 14 cases of apocrine sweat gland carcinoma have been detected so far with distant metastasis to the lungs, liver, or bone [2].

Although apocrine gland carcinoma responds poorly to chemotherapy, adjuvant radiotherapy may be used in advanced local or regional disease [9]. The treatment of choice for localized AC is wide local excision with a margin of 1 to 2 cm, along with axillary lymph node dissection up to level III if nodes are positive clinically [2].

The prognostic factors for apocrine carcinoma are difficult to identify due to small number of cases, although the possible factors are tumor size, histological type, lymph node involvement, and distant metastasis [5].

We present the case of a 72-year-old male with apocrine invasive adenocarcinoma of the left axillary area with local lymph node metastasis.

Case Summary

A 72 year old male with comorbidity of hypertension, presented with a painless, itching pustule over left axilla in April 2016. The lesion was progressive in spite of local treatment with antibiotics and steroids. After few months the lesion acquired a size of around 4 cm which was irregular, superficial and slightly raised with pigmentation. It was not associated with any constitutional symptoms, such as fever, weight loss, night sweats, or loss of appetite. He had a family history of breast cancer in grandmother.

On initial investigation it was diagnosed as melanoma of left axilla. Slide review was done which revealed Paget's disease or superficial spreading melanoma. USG Breast and CXR (PA) was found within normal limit.

In view of progressive disease he underwent incisional biopsy (S - 1764/17) from the lesion which revealed extra mammary Paget's disease. Immunohistochemistry revealed atypical cells within the epidermis showing diffuse strong positivity for pan cytokeratin and focal positivity for GCDFP, and showing no expression for GMB45, Cytokeratin 20 and p63.

Subsequently wide local excision of left axillary skin lesion with left axillary lymphadenectomy up to level III was done. HPE(S-2235/17): Apocrine Carcinoma of Skin (predominantly intraepithelial in situ carcinoma, and a few small foci of invasive carcinoma invading up to the superficial dermis). Focal pagetoid spread of the in situ carcinoma into hair follicles seen. The largest focus of invasive carcinoma measured 4 mm in greatest dimension. Maximum depth of invasion of tumor into dermis 2 mm. Lymph vascular tumor emboli present without perineural invasion. All skin margins free of in situ or invasive carcinoma. All soft tissue margins free of tumor. Left sentinel axillary lymph nodes showed metastatic carcinoma without perinodal spread (2/3). Left axillary lymph node showed metastatic carcinoma without peri-neural spread (1/11). Immunohistochemistry: The tumor cells infiltrating the epidermis and dermis showed focal positivity for GCDFP and showed no expression for S-100, GMB45, cytokeratin 20 and p63. ER/PR receptor positive.

In view of metastatic spread to lymph nodes and to rule out distant metastasis a F-18 FDG whole body PET-CT scan was done which was suggestive of no distant metastasis.

Subsequently he was planned for Adjuvant Radiotherapy and received 45 Gy in 25 fractions over five week with Rapid Arc (VMAT) on Novalis-Tx linear accelerator. He has been on regular follow up with Tab Tamoxifen 20 mg once a day.

Discussion

Apocrine Carcinoma is a rare malignant cancer that arises predominantly in areas of high apocrine sweat gland. Till date, only around 50 cases of AC has been reported in the literature. The first of these reports was published in 1944 [5]. Most AC present as painless solitary nodule, 2-3 cm in size. Some patients also report pain, restriction of range of motion [12], ulceration and purulent discharge [6].

It is slightly more common in males as compared to females (M:F = 2.6:1). Clinical diagnosis of these neoplasm is unlikely due to the relatively slow progression of tumor growth and the relative lack of associated symptoms such as pain, swelling, or ulceration. Biopsy and histologic examination are required to reach the proper diagnosis [13].

Histologic examination of AC reveals characteristics features common to the apocrine glands. Typically, the carcinoma is composed of large cells with eosinophilic cytoplasm, hyper chromatic nuclei, mitotic figures, and apocrine decapitation secretion [13].

Apocrine carcinoma have different glandular patterns, some of these include papillary, cord-like, solid, or complex glandular patterns. Although one glandular type often predominates, multiple glandular patterns can be present in the same neoplasm. Vascular invasion correlates with a high incidence of lymph node metastases [10].

Although controversy still persists in diagnostic criteria of AC, decapitation secretion in eosinophilic epithelial cells are considered a key indicator of apocrine differentiation [8]. Consideration shall also be given to SLN biopsy and local lymph node dissection because up to 50 percent of the cases in the literature search had lymph node metastasis at time of diagnosis [13].

Paties., et al. proposed the following diagnostic criteria for AC: [2] Decapitation secretion, [2] PAS-positive diastase-resistant material in the cells or Lumina, and [3] Positive immunostaining for GCDFP-15 [11].

A number of reports have shown that staining for CD15 and lysozyme may help distinguish between AC and Eccrine carcinoma and that androgen receptor positivity is strongly associated with AC [7,14].

The standard treatment for AC is wide local excision and regional lymph node dissection up to level III for clinically positive nodes patients. The potential benefit of prophylactic regional lymph node dissection in clinically node negative patients is controversial in spite of the fact that approx. 50% of patients with AC at the time of diagnosis have regional lymph node metastasis. Some studies have shown that prophylactic lymph node dissection has no influence on survival or disease recurrence in AC patients. However, several case reports have found that SLN biopsy is useful in these patients. As with other skin cancers, SLN biopsy may be helpful in assessing regional nodal status and decision-making regarding lymph node dissection in AC [1,15].

The role of adjuvant chemotherapy is unclear and is not routinely offered, because AC is generally considered resistant to chemotherapy. However, some case reports have described favourable responses to various chemotherapeutic agents in patients with metastatic AC, thus warranting further study. Compared with chemotherapy, radiotherapy may cure the local disease and reduce the risk of relapse. A marked response to radiation therapy with a total dose of 50 Gy has been reported [2,16].

Chamberlain., *et al*. suggested that adjuvant radiotherapy should be considered if the disease has one or more of the following characteristics: 1) Large tumour size (≥5 cm). 2) Positive resection margin. 3) Moderately to poorly differentiated tumour, and 4) Vascular or lymphatic invasion [16].

Interestingly, AC often expresses ER and PR, unlike most primary apocrine breast carcinomas. The relatively frequent expression of these receptors may provide a rationale for anti-estrogen therapy, by using drugs such as tamoxifen [17].

Based on our literature review, we recommend the following for the treatment of localized AC: 1) A wide local excision with a 1- to 2-cm clear margin. 2) Regional lymph node dissection for clinically positive nodes. 3) SLN for clinically negative nodes. 4) Adjuvant radiation therapy and. 5) Adjuvant anti-estrogen therapy in patients with hormone receptor-positive neoplasm.

Primary apocrine carcinoma is a rare malignancy that arises from the sweat glands, for which diagnostic criteria and treatment guidelines are yet to be established.

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