

Corpus Journal of Case Reports (CJCR)

Volume 1 Issue 1, 2020

Article Information

Received date: June 04, 2020 Published date: June 23, 2020

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Keywords

Carcinoma; Squamous cell carcinoma; Outcome; Management

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Case Report

Primary Squamous Cell Carcinoma of the Breast: A Rare Case Report and Management Decisions

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Abstract

Background

Squamous cells are normally not found inside the breast. Therefore, a primary squamous cell carcinoma of breast is an exceptional phenomenon and the management of this type of disease is still debated. The prognosis is poor, and hence there is a clinical trend to offer multimodal treatment options of surgery, chemotherapy and radiation therapy.

Aim

Clinical outcome assessment of patient with squamous cell carcinoma of the breast and management decisions.

Materials and methods: we report case of primary squamous cell carcinoma of the breast T2N0Mx in a 78-years-old woman who underwent Modified Radical Mastectomy (MRM) plus adjuvant chemotherapy and hormonal therapy without radiation therapy and review literature for appropriate management decisions.

Result

With a follow up of 24 months, the patient is alive with no of local or distant recurrence.

Conclusion

Pure primary cell carcinoma of the breast is rare and aggressive disease, often treatment-refractory, our case show that the hormonal therapy after surgery and chemotherapy, allows to achieve a high local control. Multidisciplinary approach seems to be the optimal management for early stages in this rare disease.

Background

Primary pure squamous Cell Carcinoma (SqCC) of the breast is relative rare disease; according to the general Rules for clinical and pathological recording of breast cancer published by the Japanese's Breast Cancer Society, it comprises only 0.1% to 0.4% of all cases of breast cancer [1,2]. It is important to discriminate this entity from malignancy of the skin of the breast or metastasis of squamous cell carcinoma somewhere else in the body [3-5]. This type of tumor is theorized to develop from squamous metaplasia in ductal carcinoma cells [4]. The diagnosis is determined when 90% of malignant cells are of squamous type [6,7]. Clinical and radiographic characteristics are not specific for this tumor [8]. The tumor are described as aggressive, hormone receptor negative and treatment refractory with poor prognosis. Since the first recorded case 1908, few reports of primary SqCC of the breast have been published and clinical management has yet to be standardized [9].

Aim

To assess the clinical outcome of a patient with squamous cell carcinoma of breast who underwent postoperative chemotherapy and hormonal therapy.

Method and Results

A 78 year-old, postmenopausal women, with family history of breast cancer her daughter (IDC). Noticed a lump in her left breast gradually increase size without pain or nipple discharge, or dimpling of skin. She was a multiparous lady who had breast fed all her five children. Comorbidity with diabetic and hypertension under regular treatment. Excisional biopsy done 24/06/2018 show moderate differentiated squamous cell carcinoma. Let modified radical mastectomy with axillary clearance in July 2018. Surgical pathology revealed squamous cell carcinoma without glandular differentiation. Squamous metaplasia was noted in duct epithelium, supporting the primary squamous carcinoma and pathological stage T2N0MX. The tumor was ER PR/Her 2neu⁻. The patient received adjuvant chemotherapy with three cycles of 5-fluorouracil, Epirbucin and cyclophosphamide plus another three cycles docetaxel in Yemen completed in Dec 2018. Review operation specimen in India; left breast invasive carcinoma showing pronounced squamoid differentiation with squamous pearls in between area of IDC. Therefore, this is a metaplastic mammary carcinoma Tumor size 3x2.5x2.5cm, lymph vascular invasion absent, nipple and skin involvement: absent, axillary LN 0/18, DCIS scanty foci.

IHC Results

ER 40% weak heterogeneous nuclear intensity (Allord Score 6/8). PR negative, Cerb B negative (Score 0) and Ki67 40%, high path stage: T2N0 PET CT Scan 30/1/2019 no local or distant diseased noted.

All things considered:

- i. No role for adjuvant radiotherapy
- ii. 2: Hormonal therapy with letrozole tab 2.5mg daily for five years.



The patient was evaluated at approximately 3 months after completion of therapy and then 3-6 months after words. Follow- up evaluation consisted of interval history and physical examination. Additionally, tumor marker including Carcino Embryonic Antigen (CEA), carbohydrate 15.3 (CA15.3) were assessed during the follow-up, always staying in the normal range. With a follow of 24 months, the patient is alive with no evidence of local or distant recurrence.

Discussion

Pure SqCC of the breast is a rare and generally aggressive malignancy constituting and oncology do not mention pure SqCC in their classification of malignant breast tumor. It is a rare condition and concerned to arise through metaplastic change of ductal carcinoma cells [7]. The concept of disease continuum with varying degrees of squamous metaplasia was supported by Stevenson et al. who conclude that SqCC mostly represents an extreme from squamous metaplasia within adenocarcinoma [7]. An alternate theory is that arise directly from the epithelium of the mammary ducts. The SqCC of breast is generally large (>4cm) at diagnosis and cystic in 50% of case [3], likewise our case was clinically T2 at presentation. Contrary to large primary tumor size there is lower rate of lymph node metastasis at presentation compared with similar lesions of in filtering duct carcinoma (22% Vs 40-60% IDC) [10]. Similarly, our patient had on disease in spite of extensive lymph node sampling. Unlike IDC, there significant incidence of distance of metastasis even without lymph node involvement in primary SCC of breast. Treatment is planned on a case-by-case basis, as specific treatment guidelines have not been well defined as a result of lack of data, the issue of whether to prescribe adjuvant treatment for SqCC of breast, remains unsolved [4]. Some contribution can be derived from the review by MD Anderson group of clinical pathologic features, management and outcome of SqCC of the breast in series of 33 patients.

The prognosis of this type of breast cancer is still regarded as somewhat controversial, although many studies suggest that is an aggressive disease that behave like a poorly differentiated breast carcinoma [11]. In a series of 32 patient with breast SqCC, Hennessy et al. reported 26% relapse free survival rate at five years with localized disease. The median overall survival was 37 months (range 12-108 months) with 40% surviving at five years. Median survival from the time recurrent disease was recognize was 14 months (range 2-86 months) [9]. SqCC of the breast is reported to be resistant to standard chemotherapy, for in vase ductal carcinoma (cyclophosphamide, methotrexate, 5FU and Adriamycin) as reported in studies using stander agents for neoadjuvant treatment to downstage the disease. Dejager et al. reported that cisplatin-based chemotherapeutic regimen commonly used for SqCC of primary organs other than the breast was effective for SqCC of the breast [12]. Our patient treated with (Cyclophosphamide, Epirubcin, 5FU) three cycles and weekly docetaxel for 9 weeks with good response locally and systemic control. Radiotherapy has an important role in management in view of locally advanced disease at presentation. There is insufficient evidence about the activity of endocrine therapy for SqCC, but it is reasonable to use endocrine treatment for patients with ER-and/or PR SqCC. The frequent expression of epidermal growth factor receptor (EGFR) in this disease may constitute a potential therapeutic target to be exploited. Possibly in association with platinum-based treatment [13]. Endocrine therapy is often not possible because most tumors are negative for hormone receptor markers over 90% of SqCC are estrogen and progesterone receptor negative and cases of Her 2/neuro positive SqCC are few [14,15]. Our patient was immunohistochemistry ER^+ / and PR^- / Her^- 2 negative. This makes our case with ER positivity rare, was successfully treated with hormonal therapy. Future studies should establish further treatment guidelines for SqCC.

Conclusion

Pure primary squamous cell carcinoma of breast is a very rarely and aggressive disease. Clinical and radiographic characterizes are not specific except for the presence of cystic lesion, and usually present as large primary tumor with low lymph node involvement and are hormone receptor negative. In our case use of hormonal therapy will be change the outcome. Biologic studies to determine the reasons for chemotherapy resistance and to find other treatment targets can be undertaken. Clinical trials including large series of these rare tumors are needed to increase our knowledge and improve patient's outcome.

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