Case Reports

Primary pure squamous cell carcinoma of the breast: A case report and review of the literature

J.A. Makarem[1], *M.D.*; **J. Abbas**[2], *M.D.*; **Z.K. Otrock**[1], *M.D.*; **A.N. Tawil**[3], *M.D.*; **A.T. Taher**[1], *M.D.*; **A.I. Shamseddine**[1], *M.D.*

[1]Department of Internal Medicine, [2]Department of Surgery, [3]Department of Pathology and Laboratory Medicine, American University of Beirut Medical Center (Lebanon)

Summary

Pure primary squamous cell carcinoma (SCC) of the breast is a rare condition. The exact histogenesis of this malignancy is unclear. The rarity of the condition makes it difficult to draw firm conclusions on the course of the disease and the overall prognosis. We report a case of pure primary SCC of the breast occurring in a 62-year-old woman and presenting as an enlarged breast lesion with bleeding. We also review the literature for all cases of pure primary SCC.

Key words: Breast; Squamous cell carcinoma; Primary.

Introduction

Primary squamous cell carcinoma (SCC) of the breast is a rare condition. The pathological criteria required to establish a diagnosis of primary SCC of the breast are: (1) the tumor origin must be independent from the overlying skin and nipple; (2) the infiltrating component of the breast cancer is predominantly of squamous type (> 90%); (3) no other invasive neoplastic elements, ductal, mesenchymal or otherwise present in the tumor for pure SCC; (4) another site of primary SCC in the patient must have been excluded [1].

In pure SCC all malignant cells are squamous with keratinization. In fact there are only few pure squamous tumors. An extensive review of the English medical literature using the MEDLINE database from January 1966 through October 2004 revealed only 89 reported cases that fulfill the diagnostic criteria of pure SCC of the breast [1-19].

We report a case of primary SCC of the breast occurring in a 62-year-old woman and presenting as an enlarged breast lesion with bleeding. The histology of the tumor was purely squamous with keratinization. We have also reviewed the literature for all cases of pure primary SCC.

Case Report

A 62-year-old woman presented to the Emergency Department of the American University of Beirut Medical Center with massive bleeding from the right breast. Two months prior to her admission, she developed diffuse swelling of the right breast, which was progressively increasing in size without seeking medical attention. Later on, she noticed ulceration in the lower mid quadrant of her breast followed by spontaneous bleeding from the bed of the ulcer.

Clinical examination revealed a 2 x 2 cm ulcer over the lower mid quadrant of the right breast connected to a huge cavity in the breast tissue that was full of fresh blood and blood clots. The whole breast was severely swollen with skin erythema and ecchymosis around the cavity. There was no right axillary adenopathy. The left breast was normal. The bleeding from the right breast was continuous, despite packing the cavity and applying external pressure dressing. The patient became hypotensive and tachycardic and was transferred immediately to the operating room to control the bleeding. The operative findings revealed a large cavity full of blood clots with active bleeding from an ill-defined hard mass fixed to the chest wall. As a life saving measure, the patient underwent right simple mastectomy. The tumor was adherent to the chest wall for which a part of the periosteum of the ribs was resected (Figure 1).

Pathological examination of the right breast revealed infiltrating squamous cell carcinoma moderately differentiated with keratin formation arising from the breast parenchyma. The overlying skin was normal. All surgical margins were free from tumor (Figure 2). The chest wall was negative for carcinoma. Immunohistochemistry for estrogen and progesterone receptors was negative, and Her2-Neu was not overexpressed. Postoperatively, the patient had a complete metastatic work-up. Computed tomography (CT) scan of the chest, abdomen and pelvis, bone scan and mammography of the left breast were normal. There was no evidence of any extra mammary focus of squamous cell carcinoma. Right axillary lymph node dissection was dose two weeks post operatively. Twenty lymph nodes were retrieved and all were free of tumor cells.

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Figure 1. — A gross specimen showing the resected tumor.

The patient was treated with adjuvant radiotherapy and she received a total dose of 60 Gy. Four months after surgery she is doing well with no signs of recurrence of her disease.

Discussion

Pure SCC of the breast is a rare pathologic condition. The exact histogenesis of this malignancy is unclear [1, 7]. Some authors suggest it may be due mainly to differentiation from ductal carcinoma [11]. It accounts for less than 0.1% of all breast cancers [10]. For instance there was not even a single case of pure SCC in a classical review of 1,000 cases of invasive breast carcinoma by Fisher *et al.* [20]. Furthermore in a study by Franceschi *et al.* [21] none of the 1,144 biopsies done for positive mammograms contained SCC. Also upon re-examination of about 4,000 breast cancer biopsies, three pure primary SCCs were found [10]. An extensive review of the English medical literature using the MEDLINE database from January 1966 through October 2004 revealed only 89 reported cases that fulfill the diagnostic criteria of primary pure SCC. We excluded from this review the cases that were not pure SCC and those whose histology was not clear enough to be labeled as pure type. Most of these cases have been reported as isolated case reports [12, 15, 18] and some are reported as case series [1, 3, 5, 7, 13]. It is worth noting that not all the cases reported in case series were pure SCC. For instance, in the study of Cornog *et al.* [11] which examined 24 cases of breast SCC, only two cases were of the pure type.

The existence of a primary tumor elsewhere must be excluded in primary breast SCC. Thus a complete workup of all probable primary sites should be conducted. This workup should include the oral cavity, bronchus, esophagus, renal pelvis, bladder, ovary, and cervix [6].

SCC tends to be somewhat larger at presentation than other types of breast carcinoma. The tumors are usually large (> 4 cm) at presentation and more than 50% of the tumors are cystic [3]. The mean age at presentation is 54 years. Metastases to axillary nodes are uncommon and in a recent series of seven patients with SCC, none had lymph node metastases [13].

Most of these tumors have been reported to be estrogen receptor (ER) negative [8]. According to Samuels et al. estrogen/progesterone receptor status tends to be negative [17] and therefore provides minimal assistance to further management of such cases [2]. This is expected of a tumor comprised solely of squamous cells [8].

Surgical biopsy is usually required to establish the diagnosis of SCC although it may be possible to recognize squamous carcinoma in fine needle aspiration cytology specimens [4, 19].

As there are fewer cases of primary SCC of the breast than conventional breast cancer, optimal treatment and prognosis are both unclear. The initial management will depend on the stage of the tumor, and there is general agreement that tumor size at diagnosis is an important prognostic indicator [19]. However, most primary SCCs

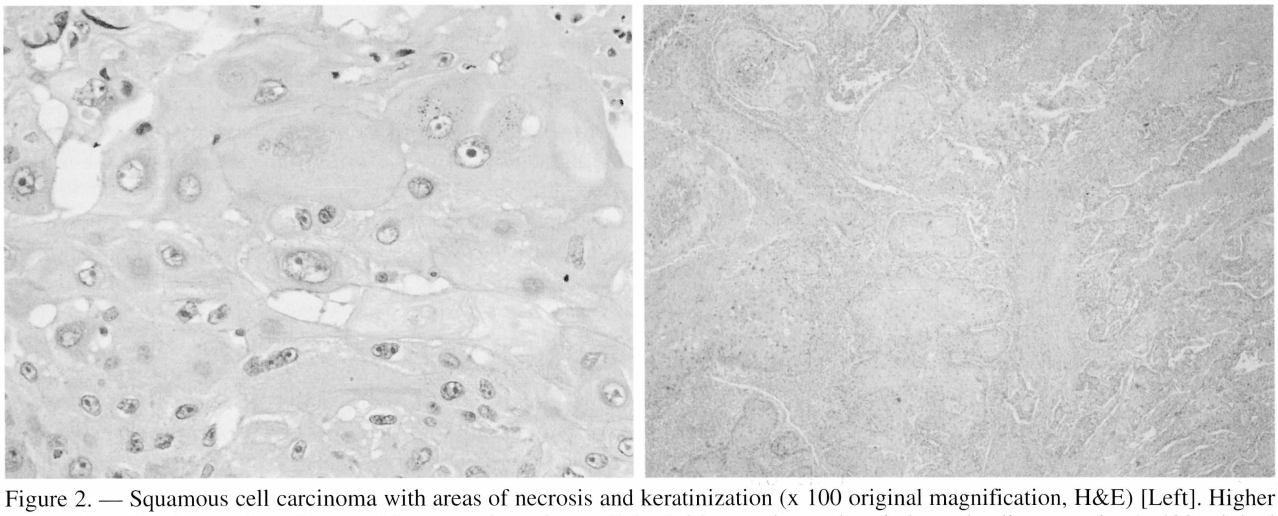

Figure 2. — Squamous cell carcinoma with areas of necrosis and keratinization (x 100 original magnification, H&E) [Left]. Higher power view showing nuclear pleomorphism, prominent intercellular bridges and cytoplasmic keratohyaline granules (x 400 original magnification), H&E [Right].

of the breast are large and thus if surgery is elected as primary management, the size of the tumor will often dictate mastectomy as the treatment of choice. There are insufficient data to dismiss breast conserving surgery as a therapeutic option.

It could be argued that axillary lymph node dissection be omitted except in cases with palpable nodal disease. This is supported by the finding that most cases of primary SCC do not have nodal involvement [13]. However, given the rather low morbidity associated with axillary dissection, some physicians might favor including node dissection in the treatment of these patients.

Not only the pathogenesis but also the clinical behavior and malignant potential of this type of tumor are unknown [13]. Age and tumor size seem to be important prognostic factors. The presence of lymph node metastasis at the time of diagnosis is also a marker of poor prognosis [3]. The prognosis of SCC is a controversial issue, with some studies suggesting that it does not seem to be different from that of breast cancer in general [11, 22] and other studies showing a more indolent clinical course although with significant propensity for local recurrence [3, 10]. This behavior, however, does not exclude the possibility of metastatic disease, namely lymph nodes and lung, even at the time of initial diagnosis [3, 22]. The largest single series, reported by Wargotz and Norris, included 22 cases of pure SCC. In this series, the cumulative 5-year survival was 63% [3]. It should be noted that there is great clinical, pathologic and therapeutic heterogeneity among different studies. Also the rarity of the condition makes it difficult to draw firm conclusions on the course of the disease and the overall prognosis.

The role of neoadjuvant chemotherapy is still controversial. A case of breast SCC reported by Takahashi *et al.* [14] showed progression of the disease while on neoadjuvant treatment with Adriamycin (doxorubicin) and Taxotere (docetaxel). In contrast, a remarkable response has been reported in a patient who received neoadjuvant cisplatinum and 5-flourouracil [16] but this has never been documented in other studies. The role of a cisplatinum-based regimen in the neoadjuvant and adjuvant settings needs to be evaluated in future studies.

Postoperative adjuvant therapy for SCC of the breast has generally been carried out in the same way as therapy for more common types of breast cancer [9, 10]. Wargotz *et al.* [3] indicated that prophylactic postoperative radiation therapy for lymph node metastasis did not prolong survival. The role of adjuvant radiation therapy remains unclear in the absence of clinical trials. Thus, further studies are required to determine whether primary pure SCC of the breast is to be treated as a separate entity or as a common type of breast cancer.

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Address reprint requests to:
A. SHAMSEDDINE, M.D.
Division of Hematology/Oncology
Department of Internal Medicine
American University
of Beirut-Medical Center,
P.O. Box 113-6044
Beirut 1107 2802 (Lebanon)