CASE REPORT

Cancer En Cuirasse; A Rare Primary Presentation of Breast Cancer: Report of a Case

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ABSTRACT:

Breast Cancer is the most common malignancy associated with Cutenous metastasis. Cancer en Cuirasse is the term used to describe a rare dramatic and extensive clinical presentation of Cutenous Metastasis from Breast Carcinoma that is very rarely manifest as the primary presentation of the underlying cancer. In the present paper, we report a rare condition of Cancer en Cuirasse in a 43-year-old lady who is totally unaware of her underlying pathology. The case story and diagnosis are presented and discussed along with recent management options and appropriate literature reference.

KEYWORDS: breast carcinoma,cutenous metastasis , advanced breast carcinoma, cancer en cuirasse

INTRODUCTION:

Cutaneous Metastasis (CM) is a phenomenon that can associate internal organ malignancies and it is often a sign of advanced tumor stage [(1)].

Breast cancer (BC) is not only the most common malignancy in women, but it is also the most common cancer that is associated with CM and account for about 70% of the sources followed by the colonic cancer, malignant melanoma and ovarian/cervical cancers in this order [(1)]. About 25% of women with breast BC will develop a sort of CM in the life time of the disease [(2, 3)].

Cancer en Cuirasse (*CEC*) is the term used to describe a rare dramatic and extensive clinical presentation of CM from BC [(4)]. Lymphatics invaded by cancer cells become obstructed resulting in extensive thickening, oedema and fibrosis of the dermis and subdermis of the chest wall [(5)]. It is usually seen as a local recurrence after mastectomy for BC but may rarely be seen as the presenting feature of an underlying BC [(4, 5)].

In the present report; we document a case of *CEC* in a 43-year-old lady who present for the first time with a frank extensive clinical picture of the disease. The case history and clinical findings are presented and the condition discussed with the appropriate reference to the literature.

CASE REPORT:

A 43-year-old multiparous lady presented for the first time to the private clinic in April 2011 with a chief complaint of an extensive itchy, painful and tender right breast with increasing difficulty in breathing. She admitted that she started a small lesion on her right breast 6 months ago for which she had been treated on outpatient basis with different kinds of oral and local therapies but progressed to her present state. She denies any personal or family history of malignancy and did not report such symptoms like fever, night sweats or weight loss.

On examination; there is an extensive prominently indurated raised fungating breast plate covering the entire range of a markedly retracted right breast and extending to the adjacent area of the chest.

Sharp demarcation was particularly striking over the entire right breast including the inframammary crease. Medially, the lesion is slightly crossing the left parasternal line. The sharply demarcated margin of the plate is seen fading in to erythematous plaques on all borders with few satellite lesions over the medial border of the left breast and the right axilla.

The right nipple could not be seen and reliable and thorough deep palpation of the right breast could not be performed. There is palpable right axillary lymphadenopathy. Apart from findings visible on the left breast; palpation of the breast and left axilla was unremarkable (Photograph 1).

Ultrasound and mammography were performed to exclude underlying abnormalities in the right

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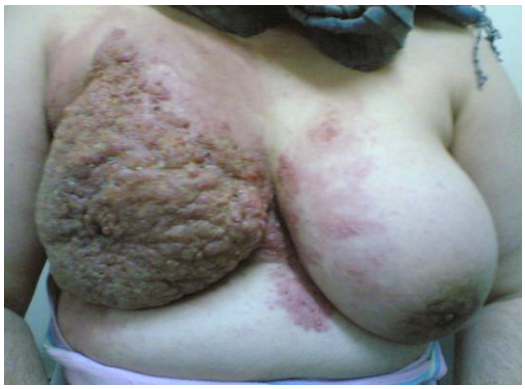
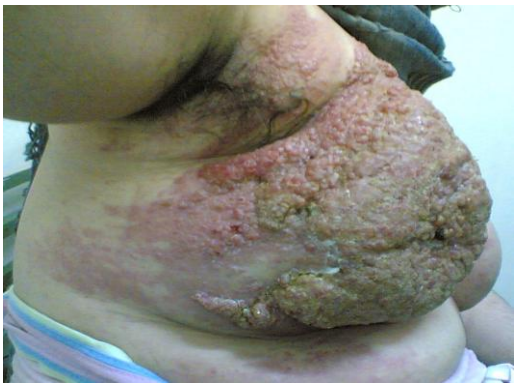
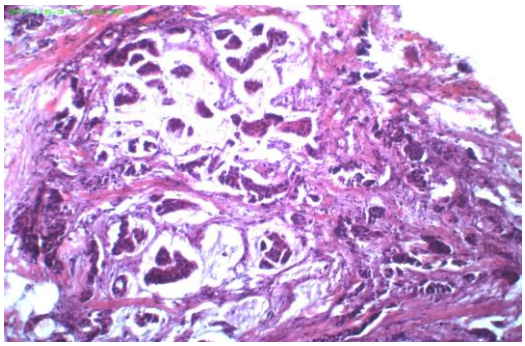
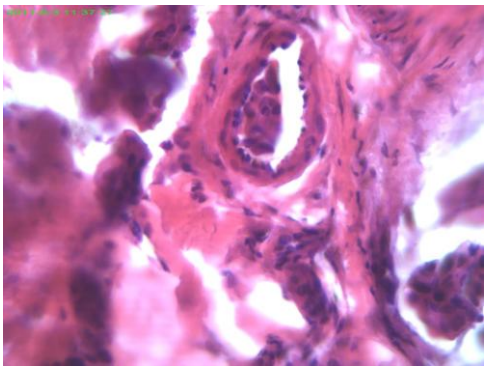
breast but both were of little help due to the significant degree of skin thickening and subcutaneous oedema. Chest x-ray was performed and the right chest field was partially obliterated by the lesion and no evidence of pulmonary metastasis can be reported.

Photograph 1: an anterior and right lateral view of the lesion over the right breast

An incisional biopsy performed and histological examination of the material showed clusters of malignant duct epithelial cells floating within lakes of mucin and infiltrating the dermis with obstruction of the lymphatic vessels. The picture is consistent with well differentiated mucinous adenocarcinoma of the breast (Photograph 2).

Photograph 2: Histopathology of the incisional biopsy of the right breast

The patient was referred to the oncology department for adjuvant therapy. The patient died one June 2011 because of septic complications.

DISCUSSION:

Phenomena attributed to metastasis of malignant cells from an underlying BC obstructing the dermal and subdermal lymphatics, not only include CM but are variable like the sign of "*Peau de Orange*", Lymphangiosarcoma, oedema of the arm and forearm in addition to fungating BC. All have been considered as signs of advanced BC [(6)].

The time between diagnosis of BC and resultant CM is variable although it usually occurs within the first three years after detection of the primary tumor [(6)].

CM from BC usually occurs in a body region near the primary tumor, most frequently on the anterior aspect of the chest wall, in some cases spreading to the neck, axilla and upper back. Generally; it is the consequence of direct extension, blood/lymphatic vessels dissemination and also surgical implantation with subsequent local recurrence after mastectomy [(7)].

Generally; there are eight well-described clinicopathological types of skin involvement by

CM from BC [8]. Those include Inflammatory BC, Telangectatic metastatic BC, Nodular metastatic BC, BC of the inframammary crease and the axilla (Intertrigo), Paget's disease of the nipple and the *CEC* [8, 9].

On the other hand; various morphological forms of CM from BC were described with the most frequently being solitary to multiple erythematous infiltrating papules and nodules [9] or a less common atypical variants that can mimic common processes like Erysipelas (Carcinoma Erysipeloides) [10], Herpes Zoster (Zosteriform metastasis) [11], Keloid of the chest wall [12], Radiation Dermatitis [13], Alopecia Areata (Alopecia Neoplastica), Lymphangioma Surcuscriptum, Cutenous Vasculitis (Carcinoma Telengectaticum), Basal Cell Carcinoma, Squamous Cell carcinoma and Intertrigo (Inframammary Carcinoma) [14, 15] .

In 1943; Savatard L. announced that the term "*CEC*" was first coined by "Alfred Velpeau", a well-known French anatomist and surgeon who described this condition for the first time in 1838. The name given by Velpeau is based on the resemblance of the pathology to the steel breastplate of Cuirassier (the cavalry Soldier). He used it specifically to describe the diffuse sclerodermoid induration of the skin that characterised this unique clinical presentation [16]. However; other terms had also been given for this distinctive morphological variant of CM such as the "Armored Cancer" (from "encasement of Armor"), panzerkerb, Scirrhous Carcinoma and Acarcine Eburnee [17].

CEC has a distinctive clinical presentation. It typically starts with swelling, pitting oedema and scattered firm papules and nodules overlying an erythematous breast [18]. Those papules and nodules then coalesce in to a thickened leathery plate-like mass that may be pruritic, bleeding or having foul smelling discharge [19].

It is important to mention that the pathological process in *CEC* at an early stage might macroscopically be similar to the features of Inflammatory BC. Unlike Inflammatory BC which begins in the breast ducts and then spread and remains contagious with the skin, *CEC* is an actual metastasis of BC to the skin [20].

Histologically; the tissue in *CEC* is fibrotic with tumor cells in between collagen bundles in a linear pattern. The densely fibrotic matrix, combined with the decrease vascularity seen in this condition, make this pathology particularly resistant to systemic chemotherapy [21]. In this context; it is important to be differentiated from the condition called Breast Fibromatosis; the so called "Extra-abdominal Desmoid Tumor" which is an uncommon condition indistinguishable on clinical examination and imaging but histology shows the Fibromatosis process to have distinctive features of benign infiltrative proliferation of fibrous tissue [22]. *CEC* with distinctive fibrosis has also been called Scirrhous BC en Cuirasse [22].As far as the condition is a reflection of locally and probably systemically advance BC; investigations like Ultrasound, Mammography and Magnetic Resonance Imaging are not specifically helpful, first; the distortion of the breast structure and thickening of the overlying skin obliterating a clear image is not a diagnostic feature, second; the detection of any intramammary lesion by these investigations would not affect the course of diagnosis and treatment [23]. Chest X-ray might detect pulmonary metastasis and bone scan might show bony metastasis but has no clear indication in such advanced cases [24].Skin biopsy is often enough for revealing the histological type of metastasis which is usually similar to that of the primary tumor except for anaplastic carcinoma, in which the immunohistochemistry markers and ultrastructure examination may be particularly useful [25]. Pathologically; it is seen across all receptors subtypes and given the rarity of this disease, no tumor marker is known at the time being [26].Once diagnosed; the treatment of this problem is difficult and the outcome of various treatment trials is generally poor. The condition is definitely beyond any surgical procedure and systemic chemotherapy and radiotherapy are not proven helpful [26]. Adjuvant treatment modalities that have been tried include Intralesional chemotherapy especially with Capecitabine [27], radiotherapy, hormonal antagonists and HER2-targeated therapy depending on receptor expression [28]. Recently; data showed that electrochemotherapy can be effective and safe locoregional form of treatment in such patients. A success rate of 15-30% had been reported particularly in old age patients [29].

The prognosis of the condition is very poor and patients usually quickly succumb to local complications such as infection and sepsis or might die due to distant metastasis particularly to the lungs [30].

CONCLUSIONSAND RECOMMENDATIONS:

To the best of our knowledge; only few literature reports documented the rare incident of *CEC* presenting as the initial manifestation of

underlying malignancy of the breast. In our case; the patient was totally unaware about the seriousness of the underlying problem.

Early recognition of certain types of skin lesions as a possible sign of underlying or internal malignancy is vitally important to identify such a problem before advancing to a late untreatable stage. Skin biopsy for questionable odd dermatological lesions is highly recommended before starting empirical local and systemic therapy.

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