Carcinoma en cuirasse: The body shield that has grave prognosis

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ABSTRACT

Carcinoma en cuirasse is a rare form of cutaneous metastasis. Like other cutaneous metastasis, Carcinoma en cuirasse signifies advanced malignant disease with poor prognosis and short survival. We report a 45 year old woman, known case of infiltrating ductal carcinoma of the right breast and had received treatment with neoadjuvant chemotherapy followed by surgery, presented with seven month history of progressive appearance of multiple nodular lesions on the chest. Diagnosis of carcinoma en cuirasse was made based on clinical and histopathological features. Cutaneous metastasis signifies the persistence of primary cancer after treatment, and dramatically alters the therapeutic plans. Due to the high incidence of cutaneous manifestations of breast carcinoma, every practitioner should be highly suspicious of any acute-onset, persistent, firm papulonodules, especially on the chest.

Keywords: Carcinoma en cuirasse, infiltrating ductal carcinoma, breast carcinoma, cutaneous metastasis.

INTRODUCTION

Cutaneous metastases can have variable clinical appearances and can mimic benign skin lesions. They are usually seen in patients with advanced disease, but they can be the presenting lesion.[1] Carcinoma en cuirasse is a rare form of cutaneous metastasis. It is most commonly associated with breast cancer, and it presents most commonly a few months or years with local recurrence after mastectomy, but it can also be a kind of clinical presentation of a primitive tumor. Here, the thoracic wall is studded with carcinomatous indurated lesions and exophytic nodules and the affected skin seems to resemble a breast plate (armor).[2] Like other cutaneous metastasis, Carcinoma en cuirasse signifies advanced malignant disease with poor prognosis and short survival.

CASE REPORT

A 45 year old woman presented with seven month history of progressive appearance of multiple asymptomatic nodular lesions on the chest. One and half year earlier she was diagnosed of infiltrating ductal adenocarcinoma of the right breast and had received treatment with neoadjuvant chemotherapy followed by a modified radical mastectomy and axillary lymph node dissection on right side.

She gave a history of development of small fluid filled vesicles over right infra axillary area seven months back which gradually became hard and dark colored, and began to spread, to involve the chest wall including the left breast, left infra axillary area, right supraclavicular area, and right shoulder. The lesions over right side chest wall began to ulcerate and started to ooze foul smelling discharge. She had been using over the counter medications like steroid creams, anti histamines, and antibiotics prior to her appointment with us.

On examination the patient was moderately built, with stable vital signs. Local examination revealed diffuse erythema, with multiple well defined hyperpigmented firm to hard plaques and nodules, with few areas of yellow crusts and purulent discharge noted over anterior chest wall extending to right supra clavicular area, right shoulder, right and left infra axillary area. (Figure 1-3). Biopsy of the skin lesion showed atrophic epidermis, and cells that are highly pleomorphic, with vesicular nuclei and prominent eosinophilic nucleoli, and abundant cytoplasm containing melanin pigment in the dermis suggestive of metastatic invasive ductal carcinoma. (Figure 4). The patient was attached to the services of tertiary care cancer center for further evaluation and management.

DISCUSSION

Cutaneous metastases of a primary internal malignancy are relatively uncommon, with an overall incidence ranging from 0.7 to 10.4% .[3,4] Their presence is influenced by the type and incidence of the primary tumor and the characteristics of the affected patients. The incidence of cutaneous metastases in patients with breast carcinoma is 23.9%.[4]

The most common sites of cutaneous metastases in breast carcinoma patients are the chest wall and abdomen, but they can occur at the extremities and in the head/neck region. Due the high incidence of breast carcinoma, BCCMs (Breast Carcinoma Cutaneous Manifestations) are the most common metastases seen by dermatologists. In clinical practice, cutaneous metastases show a wide range of clinical manifestations (Table 1).[5]

Table 1. Various clinical manifestations of cutaneous metastases of breast carcinoma.

Clinical aspect	Number of cases	% of Total
Nodular carcinoma	141	46.8
Alopecia neoplastica	36	12
Telangiectatic carcinoma	25	8
Malignant melanoma-like metastases	19	6.3
Carcinoma erysipelatoides	19	6.3
Subungual metastases	14	4.6
Carcinoma en curasse	13	4
Zosteriform metastases	11	3.6
Metastasis to eyelids	7	2.3
Paget- like metastases	4	1.3
Dermatomyositis	3	0.9
Granuloma pyogenic–like metastases	2	0.6
Multicentric reticulohistiocytosis	1	0.3
Sister Mary Joseph's nodule	1	0.3
Metastatic squamous cell carcinoma	1	0.3
Occult metastases	1	0.3
Infection-like acral cutaneaous metastasis	1	0.3
Clown nose	1	0.3
Targetoid cutaneous metastases	1	0.3
Nodule on benign intradermal nevus	1	0.3
Erythema annulare centrifugum- like	1	0.3
Dermatitis-like metastases	1	0.3
Calciphylaxis	1	0.3

Cancer en cuirasse is a rare cutaneous manifestation of breast cancer. This term was coined by Alfred Velpeau, a well-known French anatomist and surgeon, after describing this condition for the first time in 1838.[2] Carcinoma en cuirasse, also known as scirrhous carcinoma, is a form of metastatic cutaneous breast cancer, but it can also be a kind of clinical presentation of a primitive tumor.[6] The BCCMs can occur in form of nodules and/or papules or telangiectatic lesions in more than 90% cases and cancer en cuirasse accounts for only about 3-4 % of such lesions. Taylor and Meltzer first described 38 cases of this inflammatory metastasis involving the skin of the chest arising on the breast. Cancer en cuirasse most commonly appears as a local recurrence after a few months or years of mastectomy for breast cancer. More rarely, the condition can present as the presenting initial manifestation of breast cancer.[2] Cancer en cuirasse may initially appear as few small discrete cutaneous nodule/papules which progressively increase in size and coalesce to form a leathery indurated sheet (Figures 1 and 2) resembling a breast plate of armor.[6] Uncommonly, the lesion may appear as a subcutaneous nodule or yellowish marbling when it may be regarded as insignificant. Mullinax and Cohen have reported a case where cancer en cuirasse presented as keloids of the chest wall.[7]

Breast fibromatosis (also called extra-abdominal desmoid tumor) is an uncommon differential diagnosis of cancer en cuirasse.[8] It is indistinguishable on physical examination and imaging but histology shows fibromatosis to have distinctive features of benign infiltrative proliferation of fibrous tissue. Cancer en cuirasse develops continuitatem by dissemination via lymphogenous and hematogenous pathways.[9]

A biopsy of the skin helps in confirming the diagnosis of tumor. The pattern noted and the microscopic appearance often suggests the likely tissue of origin. The initial diagnosis can be made by examining frozen sections, but the final diagnosis should be reserved until permanent sections are included. Generally, the histologic features of the metastases are similar to the primary tumor, although metastases may be more anaplastic and exhibit less differentiation. Histopathological findings in Cancer en cuirasse include fibrosis with few neoplastic cells, sometimes exhibiting a characteristic Indian file pattern.[4]

Effective treatment depends on treatment of the underlying tumor. Palliative care is given if lesions are asymptomatic and the primary cancer is untreatable. This care includes keeping lesions clean and dry and debriding the lesions if they are bleeding or crusted. Hydrocolloid dressings may be used to help prevent secondary infection.[10]

The prognosis depends upon the type and biological behaviour of the underlying primary tumor[2] and is generally poor as the cutaneous metastases represent the advanced stage of the disease.

CONCLUSION

Due to the high incidence of cutaneous manifestations of breast carcinoma, every practitioner should be highly suspicious of any acute-onset, persistent, firm papulonodules, especially on the chest. The lesions must be appropriately investigated and treated aggressively as the prognosis is generally poor in such cases.

Figure 1

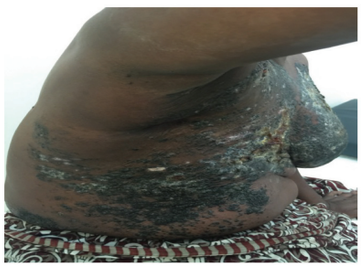

Figure 2

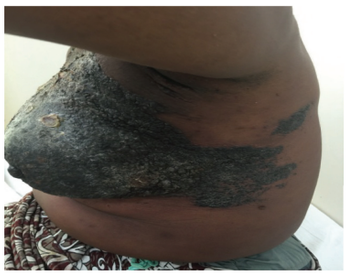

Figure 3

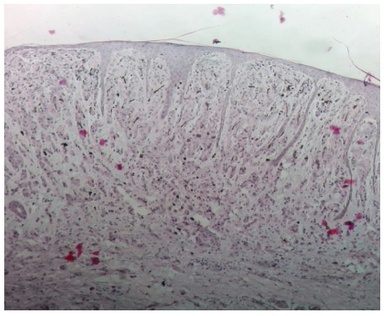

Figure 4

Figure 1: Hyperpigmented nodules and plaques interspersed with crusting noted over anterior chest wall resembling protective armour of soldiers.

Figure 2: Carcinoma en cuirasse seen extending to right infra axillary area and on to the back.

Figure 3: Carcinoma en cuirasse seen extending to left infra axillary area and on to the back.

Figure 4: Dermis shows ill defined tumour composed of cells arranged in nests, cords & singles. The tumour is seen abutting the epidermis. (H & E Stain, x100)

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