



Letter to the Editor

An unusual cutaneous manifestation of inflammatory breast carcinoma

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Sir,

Inflammatory breast carcinoma (IBC) is an unusual but distinct form of cutaneous metastasis. It was originally described in 1924 in a review of 28 cases of breast cancers associated with inflammatory skin changes. These skin changes, characterized by red induration with distinctly marginated borders associated with skin metastases, were later described as “carcinoma erysipeloides” by Rasch in 1931 for their similarity to the infectious skin condition erysipelas. Thus, both the terms, inflammatory carcinoma and carcinoma erysipeloides, have been used interchangeably when describing skin metastases with inflammatory skin changes that showed histopathological features of invasion of dermal lymphatics by neoplastic cells.^[1]

Here, we report a patient who initially presented to the dermatology department with lymphangioma circumscriptum like lesions over the right breast which later turned out to be IBC.

A 46-year-old female presented to the outpatient department of dermatology with a 2-month history of asymptomatic papules and vesicles over the right breast with a recent onset of erythema over the entire breast. There was no prior history of topical or systemic treatment. On examination, there were multiple small grouped papules and vesicles, with a frog spawn appearance, over the upper half of right breast extending up to the infraclavicular area. The skin of the entire right breast was erythematous without any local rise of temperature or tenderness [Figure 1]. On palpation, the right breast was firmer compared to the left, but there were no palpable masses. The nipple on the right side was retracted. The left breast was normal on examination. There was no significant axillary or cervical lymph node enlargement. The clinical diagnosis of skin lesions was consistent with acquired lymphangioma circumscriptum. A skin biopsy was taken from the papulovesicular lesions which showed neoplastic cells arranged in nests within the dermis [Figure 2]. The individual cells had moderate amounts of eosinophilic cytoplasm and vesicular nuclei with prominent nucleoli. There were multiple tumor emboli in the dermal lymphatics [Figure 3]. The patient was referred to surgery department for evaluation of the underlying disease.

Breast imaging showed an irregular radiopaque lesion in the upper quadrant of the right breast with microcalcifications (BIRADS V). A trucut biopsy from the underlying breast tissue revealed invasive carcinoma of breast. Immunohistochemistry was positive for Her-2. The patient was referred to the oncology department considering the locally advanced nature of the disease. After four cycles of chemotherapy with adriamycin and cyclophosphamide followed by four cycles of docetaxel, modified radical mastectomy was done and it revealed invasive ductal carcinoma

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Figure 1: Papulovesicular lesions on upper medial quadrant of the right breast with surrounding erythema.

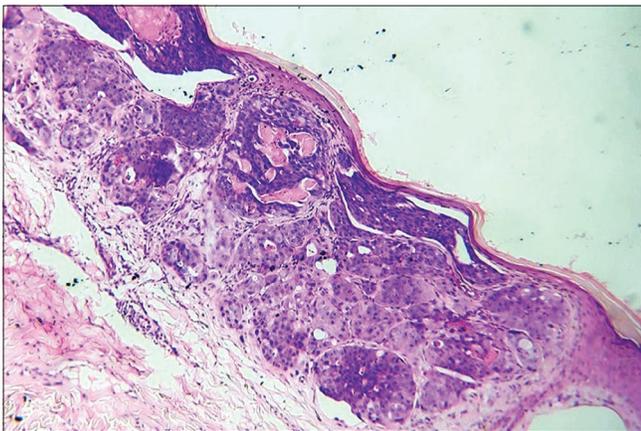


Figure 2: Neoplastic cell arranged in nests in dermis (H&E, x100).

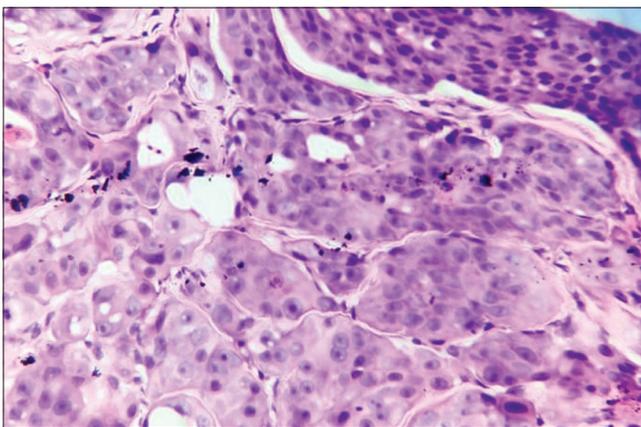


Figure 3: Malignant cells with moderate amount of eosinophilic cytoplasm, vesicular nuclei, and prominent nucleoli (H&E, x400).

with metastasis to axillary lymph nodes. Thus, we made a final diagnosis of IBC presenting with lymphangioma circumscriptum like lesions.

Breast malignancy is the most common cause of cutaneous metastases.^[2] Various morphological variants of cutaneous metastases secondary to carcinoma breast (CMBC) have been described. The most common being solitary to multiple erythematous infiltrated papules and nodules.^[2] Other less common variants included in a study of 164 cases by Mordenti *et al.* were carcinoma erysipeloides or IBC, carcinoma telangiectaticum, carcinoma en cuirasse, and alopecia neoplastica.^[3] Schwartz described eight distinct clinicopathological subtypes of CMBC: Nodular (the most frequent CMBC), Paget's disease, en cuirasse carcinoma, carcinoma of the inframammary crease, inflammatory carcinoma, telangiectatic carcinoma, metastatic breast cancer of the eyelid, and alopecia neoplastica.^[4]

IBC or carcinoma erysipeloides is an aggressive and infiltrative stage of malignancy that frequently presents with a myriad of non-specific symptoms such as fullness or heaviness of breast and erythema and edema in the overlying skin. It is characterized by rapid progression and poor prognosis. Younger women of all ethnicities with an average age of 55 years are most commonly affected. In majority of patients with IBC, there are no palpable tumors at presentation and the condition is often misdiagnosed as dermatitis or mastitis due to the overlying skin changes of erythema, edema, and peau d' orange appearance. IBC is usually diagnosed late and the mean 10-year survival after diagnosis is 18%. The overall survival of IBC patients is significantly less than that of non-IBC patients.

According to the American Joint Committee on Cancer (AJCC), 8th Edition (2017) AJCC Staging System for Breast Cancer, IBC is a clinicopathological entity characterized by diffuse erythema and edema (peau d' orange appearance) involving approximately one third or more of skin of the breast. The skin changes may be due to lymphedema caused by tumor emboli within the dermal lymphatics, which may or may not be obvious in small skin biopsy. The pathological finding of tumor in dermal lymphatics in the skin biopsy is not necessary for a diagnosis of IBC. A tissue diagnosis is necessary to demonstrate an underlying invasive breast carcinoma. A characteristic feature of IBC is its rapid evolution from first symptom to diagnosis in < 6 months.^[5]

Most of the cutaneous metastasis occur due to lymphatic spread of tumor cells.^[2] IBC occurs when malignant cells disseminate throughout the dermis and subcutis through the lymphatics. Carcinoma telangiectaticum is characterized by spread only through the superficial lymphatics and blood vessels. In nodular metastasis and carcinoma en cuirasse, malignant cells spread mainly through tissue spaces and to a lesser extent through lymphatics. Hematogenous spread occurs in alopecia neoplastica.^[3] In inflammatory carcinoma, tumor cells plug up many dilated lymphatics in the dermis in association with a perivascular lymphoplasmacytic infiltrate.

The erysipelas-like appearance is caused by capillary congestion. Carcinoma telangiectaticum is characterized by markedly dilated capillaries in the papillary dermis filled with tumor masses and erythrocytes.^[4] These features were absent in our patient. Carcinoma telangiectaticum preferentially affects the superficial blood vessels, while IBC affects lymphatics in superficial and deep dermis. Parkes-Weber-Bowles suggested that carcinoma telangiectaticum may represent a superficial variant of IBC.^[6]

IBC is not considered as a specific histological subtype of breast carcinoma. Most IBC are ductal carcinomas with high nuclear grade.^[4] Certain pathologic features can provide supporting evidence, but are neither necessary nor sufficient for a diagnosis of IBC. Dermal lymphatic invasion is one such feature; it can occur in all stages of breast cancer, but in IBC, dermal lymphatic emboli are often more numerous and larger in size. Two skin punch biopsies to identify dermal lymphatic invasion should be attempted whenever possible, but this finding is confirmed in only 75% of biopsy samples and is not required for the diagnosis.^[7]

IBC represents a diagnostic challenge, and delay in diagnosis is common.^[8] The international expert panel on inflammatory breast cancer has advised the following criteria supporting a diagnosis of IBC: Rapid onset (≤ 3 months) of edema, erythema (affecting $> \frac{1}{3}$ of the breast), warmth, presence of a palpable mass or adenopathy, nipple retraction/crusting/flattening, core needle biopsy-proven IBC, previously diagnosed mastitis unresponsive to a 7-day course of antibiotics, overall duration < 6 months, and skin punch biopsy revealing intralymphatic tumor emboli.^[9] Erysipelas, cellulitis, and dermatitis of overlying skin of breast are the usual presentations but atypical presentations can also occur. Diagnostic imaging, such as ultrasonography, mammography, and magnetic resonance imaging, is often unsuccessful in detecting breast masses and in 30% of incidentally detected cases, distant metastasis is seen.^[10] IBC tends to have a worse prognosis in comparison to with other types of malignancies of breast.

Our case presents some peculiarities in that, the initial lesions were papulovesicles; erythema and induration appeared later in the course of illness and also the erythema was diffuse rather than well-demarcated. Papulovesicular lesions are described in carcinoma telangiectaticum, but not in IBC, according to the best of our knowledge.^[6,11]

A subtle skin change may be the harbinger of a grave disease. The dermatologist must be aware of the skin changes that should alert him to the possibility of an underlying malignancy, as skin is aptly described as the mirror of internal diseases. In addition to classical presentations of metastatic breast carcinoma, the presence of an atypical skin lesion on breast warrants evaluation for underlying malignancy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

Dr Mary Vineetha is on the editorial board of the Journal.

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