

CASES FOR DIAGNOSIS

Erythematous Papules on the Arm of a Mastectomy Patient

A. Lucas and I. Betlloch

Servicio de Dermatología, Hospital General Universitario de Alicante, Alicante, Spain

Clinical History

A 75-year-old woman was seen for a 2-day history of asymptomatic rash on the right upper limb. Of interest in her past history was a mastectomy with axillary lymphadenectomy performed in 1990 for breast cancer. A recurrence in 1995 was treated with chemotherapy, radiotherapy, and tamoxifen. At the time of consultation, the patient was not on any pharmacological treatment, and was only undergoing sessions of lymphatic drainage.

Physical Examination

There was edema affecting the whole right upper limb, and multiple erythematous papules of 4-6 mm in diameter were present from the shoulder to the fingers, on both the anterior and the posterior surfaces of the arm (Figures 1 and 2). There were no signs of inflammation or induration of the mastectomy scar on the right breast and there were no palpable lymph nodes. The patient denied fever or other systemic symptoms.

Additional Tests

Complete blood count, basic biochemistry, and an echo-Doppler of the right upper limb were all normal.



Figure 1.



Figure 2.

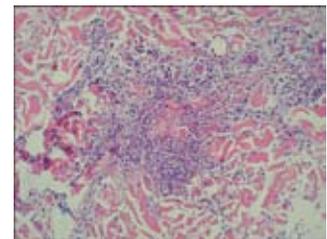


Figure 3. Hematoxylin-eosin, $\times 200$.

Histopathology

Biopsy showed preservation of the epidermis with mild edema of the papillary dermis and an inflammatory infiltrate formed mainly of neutrophils that affected the full thickness of the dermis, with nuclear dust and no vasculitis (Figure 3).

What was the Diagnosis?

Correspondence:
Ana Lucas Costa
Servicio de Dermatología
Hospital General Universitario de Alicante
C/ Pintor Baeza, s/n
03010 Alicante, Spain
lucas_ana@gva.es; analucos@yahoo.es

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Diagnosis

Atypical Sweet syndrome on postmastectomy lymphedema.

Clinical Course and Treatment

The patient was treated with paracetamol and the lesions resolved spontaneously within a week.

Comment

Sweet syndrome, or acute febrile neutrophilic dermatosis, was first described by Sweet in 1964.¹ It is characterized clinically by edematous, erythematous nodules or plaques on the face, neck, chest, and limbs, associated with fever and leukocytosis with neutrophilia. Histologically there is a variable degree of edema and an intense neutrophilic inflammatory infiltrate that affects the full thickness of the dermis, but with no vasculitis. The cause is unknown, though it is thought to be due to a hypersensitivity reaction to certain antigens. It has been associated with neoplasms, infections, autoimmune diseases, pregnancy, trauma, and drugs.² Sweet syndrome is typically divided into 3 groups: idiopathic (80%-90% of cases), paraneoplastic (hematological or solid tumors), and drug-induced (described particularly after the administration of granulocyte colony-stimulating factor).³

Many atypical clinical and histological forms of Sweet syndrome have been described; its appearance on an upper limb with postmastectomy lymphedema is not widely reported. In a review of the literature, we have found a number of cases with these characteristics,^{3,4} and a series of 7 cases has been published recently.⁵ The majority presented erythematous papules that coalesced into plaques. In some cases there was fever and general malaise. Skin biopsies revealed an intense neutrophilic infiltrate in the dermis associated with edema of the papillae and occasional leukocytoclasia, but without vasculitis. The lesions always resolved without leaving a scar, and no recurrence of the primary tumor or metastases were detected that could explain the origin of the skin lesions.

In our case, the principal conditions included in the initial differential diagnosis were infections such as erysipelas, cellulitis, folliculitis, or herpes zoster, erysipeloid cutaneous metastases, thrombophlebitis, contact eczema, and a recall phenomenon. It was possible to exclude these conditions based on the histology and additional tests, as well as on the clinical course of the syndrome.

The pathogenesis of Sweet syndrome restricted to an area of lymphedema is unknown, though a number of theories have been proposed based on the fact that altered lymph drainage promotes the action of certain cytotoxic agents, with the consequent appearance of inflammatory responses.⁶

In conclusion, we believe that Sweet syndrome restricted to an area of postmastectomy lymphedema is a relatively often unrecognized complication of a neoplasm as common as breast cancer, and its prevalence is probably underestimated as its atypical appearance can lead to confusion with other disorders.

Conflicts of interest

The authors declare no conflicts of interest.

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