Giant cellulitis-like Sweet syndrome: An underrecognized clinical variant mimicking skin and soft tissue infection

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A B S T R A C T

A new clinical variant of Sweet syndrome, called giant cellulitis-like Sweet syndrome, can masquerade as cellulitis because the patients present with an acute onset of large erythematous plaques, fever, and leukocytosis with neutrophilic predominance. This case describes a 90-year-old female with a history of invasive ductal carcinoma of the breast who presented with 3 days of erythema of the right chest and right leg. Physical examination was notable for well-demarcated, blanching erythematous rashes involving the right chest and right lower extremity. Laboratory data was notable for neutrophilic leukocytosis. A clinical diagnosis of cellulitis was made initially, and intravenous cefazolin was initiated. The rash had only partially improved with antibiotics. Skin biopsy revealed a dense neutrophilic infiltrate, which was consistent with Sweet syndrome. Based on the widespread plaques, this case was considered a “giant cellulitis-like” variant of Sweet syndrome. Clinicians should have a high index of suspicion for Sweet syndrome when assessing a patient with fever, neutrophilia and erythematous skin plaques atypical of cellulitis because this condition does not respond to antimicrobial therapy and requires systemic glucocorticoid therapy.

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Introduction

There are many clinical mimics of cellulitis. A new clinical variant of Sweet syndrome, called giant cellulitis-like Sweet syndrome, has recently been reported [1]. This clinical variant can masquerade as cellulitis because the patients present with an acute onset of large erythematous plaques, fever, and leukocytosis with neutrophilic predominance.

Case presentation

A 90-year-old female with a history of invasive ductal carcinoma of the right breast previously treated with lumpectomy and whole breast radiotherapy, currently in remission, presented with 3 days of confusion, generalized weakness, and erythema of the right chest and right lower extremity. The patient did not have a history of collagen vascular disease or other immunocompromising condition, and denied recent changes in skin products, detergents, or medications. A review of systems was negative for fever, chills, headache, nausea, vomiting, diarrhea, dysuria, cough, or dyspnea. Physical examination was notable for pruritic, well-demarcated, erythematous blanching patches involving the right breast, axillary and flank area, and right lower extremity but sparing the right arm and the left side of the body (Fig. 1). The right thigh and leg were warm and tender. There was no lymphadenopathy and no lymphangitic streak. The leukocyte count was 29,100 per microliter with 78 % neutrophils. A clinical diagnosis of cellulitis was made initially, and intravenous cefazolin was initiated. However, her rash had only partially improved after the initiation of antibiotics, and leukocytosis with neutrophilic predominance had persisted. Because of the limited response to antimicrobial therapy and the atypical distribution of the rash, a skin biopsy of the right chest wall was performed. Skin biopsy revealed a dense dermal neutrophilic infiltrate with no evidence of leukocytoclastic vasculitis, and the histopathologic diagnosis of Sweet syndrome was made. Based on the widespread plaques, this case was considered a “giant cellulitis-like” variant of Sweet syndrome. Oral prednisone at 80 mg per day was initiated on hospital day 6, and the course of antibiotics was completed on day 10. There was no evidence of hematologic malignancy based on peripheral blood smear and flow cytometry. The patient was discharged with outpatient follow-up with dermatology. The rash was resolved by the time of a 2-week follow-up visit.

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Discussion

Sweet syndrome, also known as acute febrile neutrophilic dermatosis, is a rare inflammatory disorder characterized by the sudden onset of painful, edematous, and erythematous plaques. Skin findings are typically asymmetrical in distribution, with the upper extremities, trunk, head, and neck being the typical sites of involvement [2]. Lesions are usually several millimeters to centimeters in diameter, but large cellulitis-like plaques have been reported as a rare clinical presentation of Sweet syndrome, as seen in our patient [1]. Sweet syndrome can mimic cellulitis and erysipelas with fever and leukocytosis with neutrophilia being the main clinical manifestations in addition to the skin lesions. Furthermore, this relatively newly described clinical variant of Sweet syndrome may be more likely to be misdiagnosed as cellulitis; even more potentially confusing is that Sweet syndrome triggered by cellulitis has been reported [3]. However, the simultaneous appearance of multiple asymmetric lesions with a predominance in the upper extremities and trunk and the lack of regional lymphadenopathy or lymphangitis can alert clinicians to the possibility of Sweet syndrome. Skin biopsy is required to confirm the diagnosis by detecting histopathologic evidence of a dense neutrophilic infiltrate without leukocytoclastic vasculitis [4]. Systemic glucocorticoids are the mainstay of treatment. Prednisone is recommended at 0.5–1 mg/kg per day initially, followed by a tapering dosage to prevent a recurrence. Clinical improvement is expected after a few doses and complete resolution at 1–2 weeks of treatment [5]. When a secondary bacterial infection occurs, skin lesions can partially improve if treated with antibiotics, as in this case. In conclusion, clinicians should have a high index of suspicion for Sweet syndrome when assessing a patient with fever, neutrophilic leukocytosis and erythematous skin plaques with features not typical of cellulitis such as distant lesions, asymmetrical distribution, truncal involvement and lack of response to standard antimicrobial therapy for skin and soft tissue infection, because this condition does not respond to antimicrobial treatment and requires systemic glucocorticoid therapy.

Author contribution

All the authors have participated in the preparation of the manuscript and have approved this submission. Hayato Mitaka wrote the initial manuscript draft, performed the literature review, and prepared the final manuscript. Rita Jammal and Joseph Saabiy wrote the initial manuscript draft and performed the literature review. Stanley Yancovitz and David C. Perlman reviewed and edited the manuscript.

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Patient consent

The patient consent for publication has been obtained from the patient.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Declaration of Competing Interest

None declared.

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Fig. 1. Large erythematous plaque over (A) the right chest through flank, and (B) the right lower extremity.