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Case report

Necrotising sweet syndrome mimicking necrotising fasciitis: a diagnostic challenge

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Abstract

Sweet syndrome is a neutrophilic dermatosis characterised by fever and multifocal tender, erythematous plaques or papules, which may enlarge or coalesce to form plaques with an uneven mamillated surface. Necrotizing sweet syndrome (NSS) is a severe and locally aggressive variant that may resemble necrotizing soft tissue infection clinically. In contrast to necrotizing fasciitis where prompt surgical debridement and broad-spectrum antibiotics are the mainstay of treatment, necrotizing sweet syndrome responds to systemic corticosteroids and immunosuppressants. Surgical interventions may in fact result in propagation of the disease due to the pathergy phenomenon.

Herein we report a case of necrotizing sweet syndrome involving lower limbs mimicking necrotizing fasciitis where initial surgical interventions resulted in a threatened limb and therapeutic challenges. The patient responded to high dose systemic corticosteroids and immunosuppressants. This case highlights the importance of communication between the surgeon, dermatologist and pathologist for early diagnosis of necrotizing sweet syndrome to reduce morbidity.

Introduction

Sweet syndrome (SS), or acute febrile neutrophilic dermatosis, is characterised by hyperpyrexia, neutrophilia, painful skin lesions including erythematous to violaceous nodules and plaques with uneven mamillated surface, and dense neutrophilic infiltrates in the dermis and subcutaneous tissues on histopathological examination. The skin lesions may have a pseudo vesicular appearance due to the associated pronounced oedema[1]. However, some plaques may develop vesiculation, bullae or pustules within them. These lesions favour upper extremities, head and neck but may occur anywhere. Pathergy is an important clinical feature of the disease where new lesions are induced by cutaneous trauma such as after biopsies, peripheral intravenous cannulation, venipuncture, vaccination, insect bites, and even after sunburns[1].

Necrotizing sweet syndrome (NSS) was first described in 2012 as a new variant of SS, with only a few cases reported in the literature to date[2]. This is characterised by locally aggressive disease with deep-seated necrosis, including myonecrosis which responds to high dose systemic corticosteroids[3].

Herein, we report a case of a necrotizing sweet syndrome that mimicked necrotizing fasciitis on presentation, thereby posing a diagnostic challenge to the clinician.

Case report

A 33-year-old previously healthy female was referred to the dermatology unit from the surgical unit with a large, deep necrotic ulcer with exposed tendon and bone over left shin area and generalized eruption of urticated erythematous papules and plaques (Figure 1). She has presented to the emergency department with painful purpuric macules over bilateral lower limbs for 10 days. She developed a sore throat and was prescribed oral co amoxiclav 1 day before the onset of the rash. Some of macules over the left lower limb progressed to large painful bullae which ruptured giving rise to necrotic painful ulcers at the time of admission. At the surgical unit, she was clinically diagnosed with necrotizing fasciitis (NF) and was managed with repeated wound debridement including fasciotomy and intravenous broad-spectrum antibiotics. However, the deep tissue necrosis worsened despite extensive wound debridement and later she developed generalised erythematous indurated plaques and was referred to the dermatology unit.

Neutrophilic dermatosis with pathergy phenomenon evidenced by worsening of the wound with each

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surgical debridement was suspected at the dermatology unit. This was supported by the skin changes evident as purpura and erythematous plaques surrounding the ulcer as well. Skin biopsy revealed papillary dermal oedema, inflammatory infiltrate composed of lymphocytes and neutrophils with necrosis of vessel walls and RBC extravasation with no organisms isolated from the tissue cultures.

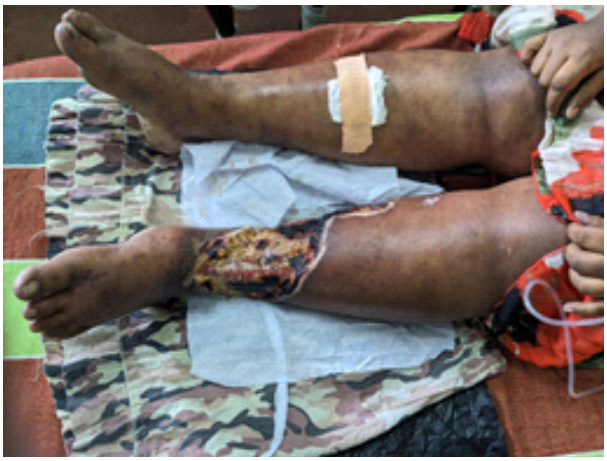

Figure 1. *Deep necrotic ulcer with exposed tendon and bone over left shin with urticated erythematous papules and plaques on the rest of the skin*

Necrotizing sweet syndrome was confirmed and patient was started on oral prednisolone 1mg/kg combined with intravenous methylprednisolone pulse therapy which resulted i2n resolution of the purpura and fever. Necrotic wound was managed by a multidisciplinary team consisting of dermatology, plastic surgery and vascular surgery specialists. Initially negative pressure wound therapy was done to control the oedema and exudation followed by split thickness skin grafting. The donor site also developed a necrotic ulcer denoting pathergy phenomenon. Further immunosuppressive therapy with IV rituximab, IVIG and IV infliximab were required to achieve disease remission.

She was tapered off immunosuppressant medications over the next 1 year and is currently stable without any maintenance treatment. She was screened for associated underlying systemic diseases associated with SS including haematological malignancies and the results were negative.

Discussion

Necrotizing sweet syndrome may clinically resemble necrotizing fasciitis (NF), a rapidly progressing, mono- or polymicrobial soft tissue infection that tracks along fascial planes[2]. NF is a surgical emergency with high mortality rate, requiring serial debridement and broad-spectrum antibiotics. Aggressive surgical debridement of NF has been shown to improve the historically high mortality rate[4] whereas serial debridement of NSS will exacerbate the condition because of pathergy, leading to exacerbation of systemic symptoms and more substantial soft tissue loss[2,4].

Discerning the difference between life-threatening necrotizing fasciitis and NSS early in the disease process prior to serial surgical debridement may be difficult. However, early diagnosis and treatment will alter the disease course and potentially the patient's outcome. In patients with clinical features of necrotizing infection but with negative tissue cultures and who appear to be progressing despite surgical debridement and antibiotics, a diagnosis of NSS should be considered. A multidisciplinary approach including dermatology, surgery, microbiology, and pathology specialists will increase the likelihood that the correct diagnosis being made expeditiously.

This case highlights the need for high clinical suspicion to distinguish a noninfectious NSS from NF and suggests evaluating a response to initial limited debridement as well as initial microbiologic studies before more extensive surgery. Early collaboration between the surgical and dermatology specialties will serve to save the limb of the patient.

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