Dermatology Practical & Conceptual

Reflectance Confocal Microscopy of Sweet Syndrome

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Key words: Confocal Microscopy, Neutrophilic Dermatosis, Biopsy, Diagnosis

Citation: Skowron F, Carbonnelle-Puscian A, Ardigò M. Reflectance Confocal Microscopy of Sweet Syndrome. *Dermatol Pract Concept*. 2025;15(1):4929. DOI: https://doi.org/10.5826/dpc.1501a4929

Accepted: August 26, 2024; **Published:** January 2025

Funding: None.

Competing Interests: None.

Authorship: All authors have contributed significantly to this publication.

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Introduction

Sweet syndrome or acute febrile neutrophilic dermatosis is a rare disease, characterized by an acute onset of tender erythematous plaques with dense neutrophilic infiltrate in the dermis [1].

Case Presentation

A 57-year-old woman developed multiple sore papules initially on the shoulders that within 3 days extended to the trunk, upper limbs, and thighs along with a mild fever 38°C. Her medical history revealed penicillin hypersensitivity and functional colonic disease. Papules were erythematous, edematous, enlarged up to 3 cm, and had a central darker area. Blood count, ionogram, C-reactive protein and renal function were normal. A papular lesion on the wrist was analyzed (Figure 1A). Dermoscopy showed homogenous violaceous pattern with some pinpoint reinforcements and a central purpuric zone (Figure 1B). Reflectance confocal microscopy (RCM) showed dark areas and numerous, single or aggregates, bright cellular structures of different shapes at the dermo-epidermal junction (Figure 2A) and some isolated bright cells surrounded by a dark acellular area at upper epidermal layers (Figure 2B). Biopsy revealed an acanthotic epidermis, intense papillary edema, and an intense polymorphous inflammatory infiltrate in the dermis composed mainly of neutrophils. There was no leukocytoclasia or vasculitis (Figure 1C). An idiopathic Sweet syndrome was diagnosed. The eruption resolved over 3 weeks with a regimen of systemic corticosteroid with progressive tapering. A work-up revealed no sepsis, no cancer, no hemopathy, or inflammatory diseases.

Conclusions

Sweet syndrome (acute febrile neutrophilic dermatosis) is a rare disease affecting typically women in the third through

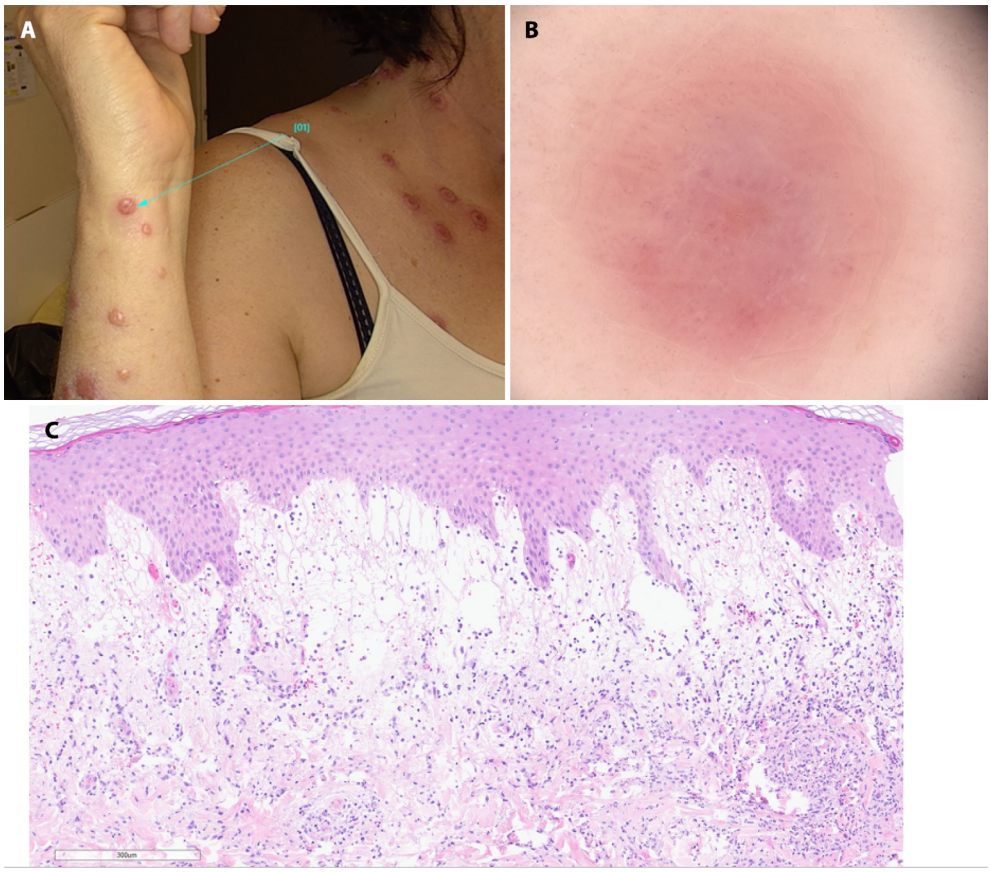

Figure 1. Clinical findings: (A) Multiple erythematous papules with central darker area on the upper trunk and arms (green marker 01: recent papular lesion on the wrist analyzed). (B) Dermoscopy: homogenous violaceous pattern with some pinpoint reinforcements and a central purpuric zone. (C) Histological findings (H&E, original magnification ×10): intense papillary edema, and an intense polymorphous inflammatory infiltrate in the dermis composed mainly of neutrophils.

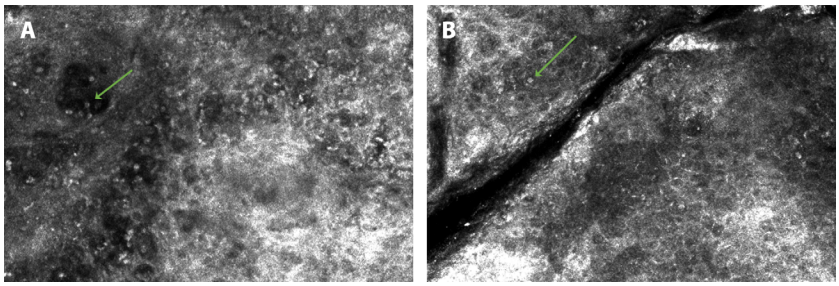

Figure 2. RCM findings: (A) Dark areas and numerous, single or aggregates, bright cells of different shape resembling polymorpho-nucleated cells (green arrow) at dermo-epidermal junction and (B) some isolated bright cells surrounded by a dark acellular area at upper epidermal layers.

sixth decades [1]. The patient presents with a rapid onset of tender, sharply demarcated inflammatory papules on the face, upper chest and back, which spread over the trunk and limbs in a few days. Most cases are idiopathic, and sometimes they are associated with drug reaction, infection, cancer, or inflammatory diseases. Dense neutrophilic infiltrate in the dermis without leukocytoclastic vasculitis is a major criterion for definite diagnosis of Sweet syndrome [2]. Intense papillary edema is another frequent histologic feature. Reflectance confocal microscopy (RCM) is a

noninvasive diagnostic tool that evaluates the epidermis and papillary dermis at a cellular level. It has been shown that RCM is efficient in psoriasis in monitoring neutrophils which appear as bright round to oval cells surrounded by a dark acellular area [3]. Edema appears as dark homogenous area just below the epidermis [4]. In our case report, RCM allowed us to observe these 2 important histologic criteria: intense neutrophilic infiltrate and papillary edema. One limit of RCM is the inability to determine presence or absence of vasculitis. However, it seems that RCM findings with neutrophilic infiltrate and dermal edema, in a context of abrupt tender erythematous lesions on the upper trunk and limbs, is helpful in diagnosing Sweet syndrome and swiftly initiating corticosteroid systemic treatment.

Conclusion: RCM is useful for managing Sweet syndrome in real time.

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