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Case Report

Acute Febrile Neutrophilic Dermatosis (Sweet's Syndrome): A Rare Presentation with Diagnostic Challenge: Case Report and Literature Review

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Abstract:

Background:

Sweet's syndrome, or febrile neutrophilic dermatosis, is a rare inflammatory condition characterized by the abrupt onset of painful skin lesions and systemic symptoms such as fever. It is strongly associated with malignancies, infections, and drug-induced triggers, with characteristic histopathological findings of dermal neutrophilic infiltration.

Case Presentation:

We report the case of a middle-aged woman who was initially hospitalized for planned lumbar surgery. She developed acute follicular tonsillitis, treated with intravenous ceftriaxone. Despite initial improvement, she subsequently developed painful papular and papulo-nodular skin lesions over her face and upper extremities. A skin biopsy revealed aggressive neutrophilic dermal infiltration, consistent with Sweet's syndrome. The condition was unresponsive to doxycycline but responded dramatically to oral prednisone, resulting in complete lesion resolution within 10 days.

Discussion:

This case underscores the importance of recognizing infections, such as Group A beta-hemolytic streptococcal tonsillitis, as potential triggers for Sweet's syndrome. It also highlights the diagnostic value of skin biopsy and the effectiveness of corticosteroids as first-line therapy.

Conclusion:

Clinicians should maintain a high index of suspicion for Sweet's syndrome in patients presenting with papular or papulo-nodular skin lesions, especially if preceded by fever, and particularly in the presence of potential triggers like infections or certain drug exposures. Prompt recognition and appropriate treatment with corticosteroids can lead to rapid clinical improvement.

Key words: sweet's syndrome; neutrophilic dermatoses; immunology; corticosteroids; nodular skin lesions

Introduction

Sweet's syndrome, also known as febrile neutrophilic dermatosis, is a rare condition characterized by painful, inflammatory skin lesions and a distinctive histological finding of neutrophilic infiltration in the dermis. [1] This syndrome can affect individuals of any gender or ethnic background and has no

specific geographic predilection. [2] While the exact cause remains unclear, Sweet's syndrome is often associated with malignancies, as well as drug reactions, particularly to fluoroquinolones. Clinically, it typically presents with systemic symptoms such as fever, followed by the development of painful papular or papulo-nodular lesions, often on the upper limbs, face, or neck. The condition is commonly seen in middle-aged women and can be triggered by infections, particularly upper respiratory or gastrointestinal infections, or by inflammatory bowel disease. In this case, the patient presented with fever, tonsillitis, and subsequent painful skin lesions, which were histologically confirmed as

neutrophilic dermal infiltration, further highlighting Sweet's syndrome's potential association with infectious triggers.

Case presentation

A 48-year-old female presented to our hospital with a six-month history of lower back pain radiating to both lower limbs at a nonspecific dermatome, associated with numbness, paresthesia and neurogenic claudication, which had not improved with conservative management. Her past medical history included type 2 diabetes mellitus, hypertension and hypothyroidism, and her past surgical history included anterior cervical discectomy and fusion at the C3-7 levels. She reported no known allergies.

Routine blood tests, including a thyroid profile, were within normal limits, and the lumbosacral spine MRI showed L3-4/L4-5 lumbar canal stenosis. She was prepared for lumbar canal decompression within 48 hours.

On the morning of surgery, the patient complained of a sore throat, headache, and myalgia, with high-grade fever. The patient was evaluated, and laboratory tests showed an increase in C-reactive protein (CRP) from 7 to 62 mg/L. A chest X-ray was unremarkable. Based on these findings, the primary treating team diagnosed her with acute follicular tonsillitis and started intravenous ceftriaxone (2g once daily), though a throat culture was not obtained prior to starting antibiotics, resulting in significant clinical improvement.

However, two days later, she developed painful papular and papulo-nodular skin lesions over her right forearm and hand (Figure 1). Laboratory investigations revealed elevated inflammatory markers, including a significant increase in CRP to 110 mg/L, an erythrocyte sedimentation rate (ESR) of 30 mm/h, and a white blood cell (WBC) count of 13 × 10^9/L, predominantly neutrophils at 10.4 × 10^9/L (81.2%).

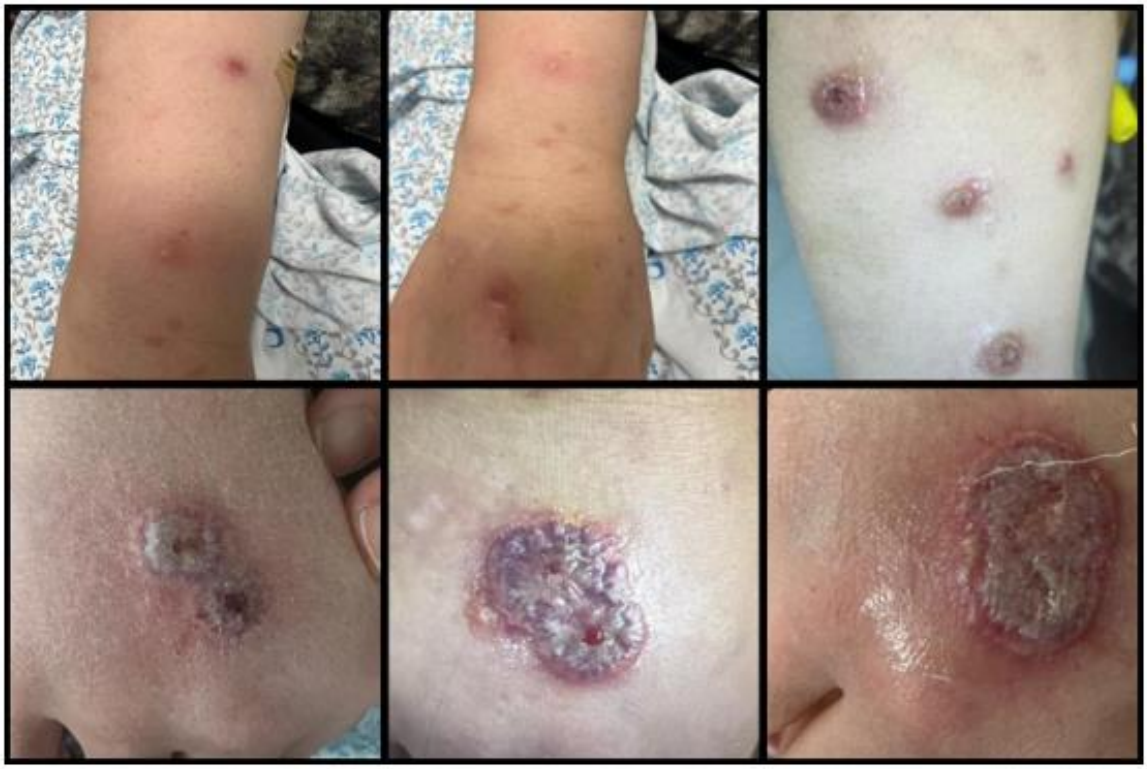

Figure 1: The above images illustrate the progression of the papulo-nodular skin lesions over the right forearm and hand. Chronologically from top left to right.

In consultation with our infectious diseases specialist, Sweet's syndrome was highly suspected. To confirm the diagnosis, a skin biopsy and an Antistreptolysin O (ASO) titer were ordered. The ASO titer result was positive (200 ToddU/ml), pointing towards infection. Additionally, a swab culture from one of the skin lesions was performed, but it was negative for bacterial growth.

Following the biopsy, doxycycline (100mg twice daily) was initiated; however, over the next few days, the patient showed no improvement, and new lesions developed on her left forearm. Upon receiving the skin biopsy results, aggressive neutrophilic infiltration of the dermis was observed (Figure 2), confirming the diagnosis of Sweet's syndrome. Given the elevated ASO titer, we strongly suspect streptococcal pharyngitis as the underlying trigger.

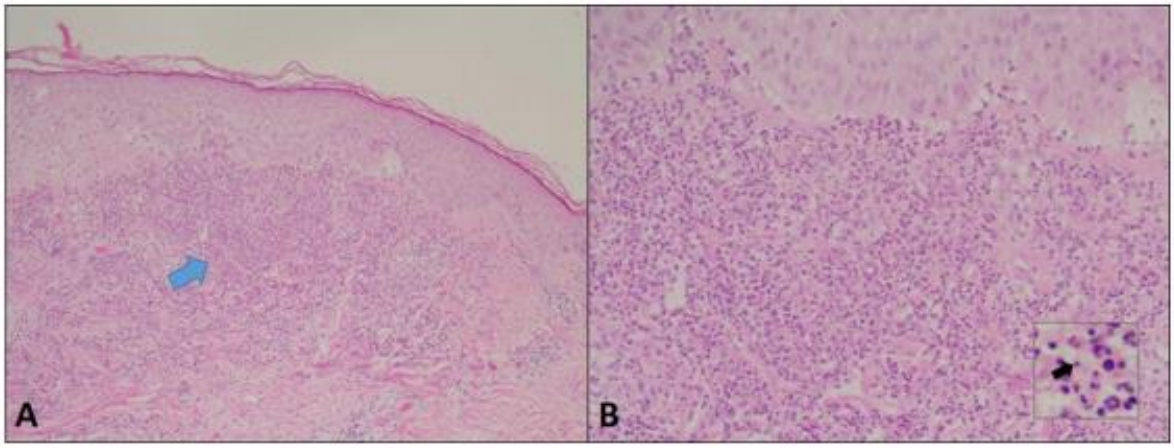

Figure 2: Sweet syndrome, skin biopsy. A. Dense dermal neutrophilic infiltrate with intact overlying epidermis (arrow) (H&E, 4X) B. Higher magnification shows the abundant neutrophils (H&E, 10X). The insert shows presence of fragmented neutrophils; leukocytoclasis (arrow).

Subsequently, oral prednisone (20mg twice daily) was added, and doxycycline was discontinued, leading to complete resolution of the lesions within one week. This dramatic clinical response supports the diagnosis of Sweet's syndrome. On follow-up, the patient remained afebrile, stable and symptoms-free.

Discussion

Sweet's syndrome, also referred to as febrile neutrophilic dermatosis, is a rare condition characterized by a constellation of clinical signs and symptoms, alongside distinctive histopathological findings. This syndrome can occur regardless of gender or ethnic background and has been reported globally, with no specific geographic predilection [3-5]. Although the exact pathogenesis remains unclear, various associated conditions and triggers have been identified. Importantly, Sweet's syndrome is strongly linked with malignancies, both hematologic and solid tumors [5, 8, 9]. It can also arise as an adverse reaction to certain drugs, such as fluoroquinolones [6]. When evaluating patients with Sweet's syndrome, it is essential to consider malignancies and drug-induced triggers as underlying causes, as these significantly influence management and prognosis.

Despite differences in etiology, the clinical and histological presentation of Sweet's syndrome remains consistent across cases. Clinically, the condition is characterized by the abrupt onset of painful papular or papulo-nodular skin lesions, which predominantly affect the upper limbs and face [3-5].

Histologically, Sweet's syndrome is marked by intense neutrophilic infiltration in the dermis without evidence of infection [3-5].

The most common variant of Sweet's syndrome is the classical form, which predominantly affects middle-aged women [5, 6, 8]. This variant often follows an infectious episode involving the upper respiratory or gastrointestinal tract, or it may occur in the context of inflammatory bowel disease [7, 8]. Regardless of the triggering factor, the characteristic clinical and pathological features remain consistent, underscoring the unifying nature of the disorder. In this context, our patient's presentation of fever, tonsillitis, and subsequent painful skin lesions, confirmed histologically as dermal neutrophilic infiltration, highlights Sweet's syndrome's potential association with infection as a trigger.

Clinically, patients with Sweet's syndrome often present with systemic symptoms, such as fever, followed by the appearance of painful skin

lesions a few days later [5, 6]. These lesions, typically nodular or papular, are most commonly distributed over the face, neck, and upper extremities. The combination of systemic symptoms and characteristic dermatological findings forms the basis for clinical suspicion and guides further diagnostic evaluation.

Diagnosing Sweet's syndrome generally requires a combination of clinical assessment and histopathological confirmation via a skin biopsy, which typically demonstrates dense neutrophilic infiltrates in the absence of infection. Accurate diagnosis is critical, as it directly impacts treatment strategies, which can vary depending on the underlying etiology. Corticosteroids remain the cornerstone of therapy and are often associated with dramatic clinical response [3, 4, 5, 6, 8, 9].

It is imperative to emphasize that, when evaluating patients with Sweet's syndrome, clinicians should always consider underlying malignancies and drug-induced triggers, as these associations significantly influence both management and prognosis.

Patient consent

Written informed consent was obtained from the patient.

Conflict of interest statement

The authors declare that they have no conflict of interest to disclose.

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References

1. Khan U, Rizvi H, Ali F, Lebovic D.(2016). Sweet syndrome: a painful reality. *BMJ Case Rep*. Dec 23:bcr2016217606.
2. Mollaeian A, Roudsari H, Talebi E.(2019). Sweet's Syndrome: *A Classical Presentation of a Rare Disease. J Investig Med High Impact Case Rep*.
3. SWEET RD. (1964).AN ACUTE FEBRILE NEUTROPHILIC DERMATOSIS. *Br J Dermatol. 1964 Aug-Sep;*76:349-56.
4. von den Driesch P.(1994). Sweet's syndrome (acute febrile neutrophilic dermatosis). *J Am Acad Dermatol*.Oct;31(4):535-56; quiz 557-560.
5. Cohen PR.(2007). Sweet's syndrome--a comprehensive review of an acute febrile *neutrophilic dermatosis. Orphanet J Rare Dis*. Jul 26;2:34.
6. Chaudhry AR, Iftikhar I, Choudhry SA, Islam R, Islam H.(2023). *Fluoroquinolone-Induced Sweet Syndrome: A Case Report.* Cureus.Mar 31;15(3):e36952.

7. Anzalone CL, Cohen PR.(2013). Acute febrile neutrophilic dermatosis (Sweet's syndrome). *Curr Opin Hematol. Jan;*20(1):26-35.
8. Villarreal-Villarreal CD, Ocampo-Candiani J, Villarreal-Martínez A.(2016). Sweet Syndrome: A Review and Update. *Actas Dermosifiliogr.* Jun;107(5):369-378. English, Spanish.
9. Marzano AV, Cugno M, Trevisan V, Fanoni D, Venegoni L, Berti E.et.al.(2010). Role of inflammatory cells, cytokines and matrix metalloproteinases in neutrophil-mediated skin diseases. *Clin Exp Immunol.* Oct;162(1):100-107.

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