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# Sweets Syndrome: A Case Report

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# Add Sweet's Syndrome to your dermatologic differential

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## INTRODUCTION

Sweet syndrome is a rare inflammatory disease that is marked by abrupt onset of painful erythematous plaques or nodules and often accompanied by fever and leukocytosis. While there is considered to be three types of Sweet's syndrome; classical, malignancy associated, and drug-induced, the most common type remains classical[4]. Classical Sweet's syndrome is associated most commonly with URI or GI infections, inflammatory bowel disease, or pregnancy though less frequent associations exist with several other medical conditions. Malignancy associated Sweet's syndrome is most commonly associated with hematologic malignancies however it has been detected in solid tumor cancer as well. Drug-induced Sweet Syndrome has been associated with several medications however Granulocyte-colony stimulating factor is the most commonly recognized[6]. Sweet syndrome can develop in any age patient, although age 30-60 is most common for classical. In Classical Sweet syndrome, 80% of cases have been in women.

## CASE REVIEW

**History**

Patient is a 44 year old male who presented with an area of painful erythema on the dorsum of right ankle, left ankle and foot, back of neck, and dorsal bilateral middle fingers extending to wrists.
All the above joints painful to move, and difficult to ambulate. The area on the back of his neck and occipital scalp were last to erupt and are not as painful.
+Fatigue, malaise, subjective chills
No new medications
No fever (+subjective chills), no abdominal pain/nausea/vomit/diarrhea/constipation
PMH: HIV (last viral load 84copies/ml 2018), syphilis (distantly treated),Type 2 Diabetes Mellitus, social anxiety, low testosterone, hyperlipidemia
Medications: abacavir-dolutegravir-lamivudine, atorvastatin, bupropion, fluoxetine, semaglutide, testosterone cypionate IM

**Physical Exam**

128/72, 96, 16, 98.3F, O2 98%RA. 226lb 12.8oz, 5ft 11.5in
Gen: well nourished, appears fatigued but no acute distress
HEENT: oral mucosa moist/pink without lesions. Neck supple, no lymphadenopathy except bilateral <1cm moveable occipital nodes
Extremities: Right wrist with dorsal erythema, mild edema, bilateral middle digits with mild edema and violaceous skin discoloration. MCP, wrists and ankles with bilateral erythema, stiff and painful on active and passive rom
Skin: see above for skin changes overlying joints. Neck and occipital scalp with erythematous plaques with raised well-defined borders. No fluctuence or tenderness in this area. Skin changes overlying joints painful to palpation (see photos)

**Assessment/Plan**

44 year old HIV positive male distantly treated for syphilis, with new onset rash, chills and polyarthritis/polyarthralgias
Differential diagnosis included vasculitis, syphilis, reactive arthritis, sweet's syndrome
Biopsy obtained of right ankle and neck lesions
RPR titer, cbc with differential, cmp, blood cultures, CRP, HIV RNA

**Outcome/Follow up**

Biopsy: neutrophilic dermatosis consistent with Sweet's Syndrome. Pertinent lab data: RPR 1:2, CRP 32.9 mg/dl, WBC 14.5.
Treatment: Oral prednisone 1 mg/kg/day. Within three days, the patient had clinical improvement. Prednisone was tapered over six weeks and the patient's rash and symptoms did not recur.

## MANAGEMENT

Once a diagnosis of Sweet's syndrome is established, management should focus on identifying underlying infection or disease processes, eliminating any precipitating drug exposure, and evaluation for malignancy including age-appropriate screening. Corticosteroid therapy is the first-line treatment for Sweet's syndrome and often induces rapid clinical response. Oral prednisone started at 0.5 to 1mg/kg per day often produces clinical improvement within 48 hours and lesions often resolve within 2 weeks. After the disease is controlled, the prednisone dose can be tapered over 4-6 weeks (1). Although most patients will need systemic corticosteroids, a minority with small localized cutaneous lesions may respond to high dose topical steroids. When avoidance of systemic corticosteroids is needed, alternative first line therapies include colchicine, dapsone and potassium iodide have efficacy based on case reports and retrospective studies (2).

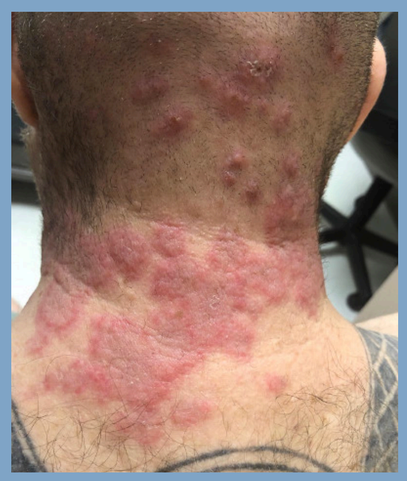
Diagnostic criteria for classical Sweet's syndrome versus drug-induced Sweet's syndrome

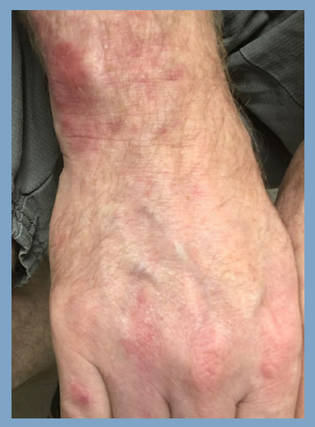
| Classical[a] | Drug-induced[b] |
|---|---|
| (1) Abrupt onset of painful erythematous plaques or nodules | (A) Abrupt onset of painful erythematous plaques or nodules |
| (2) Histopathologic evidence of a dense neutrophilic infiltrate without evidence of leukocytoclastic vasculitis | (B) Histopathologic evidence of a dense neutrophilic infiltrate without evidence of leukocytoclastic vasculitis |
| (3) Pyrexia >38°C | (C) Pyrexia >38°C |
| (4) Association with an underlying hematologic or visceral malignancy, inflammatory disease, or pregnancy, OR preceded by an upper respiratory or gastrointestinal infection or vaccination | (D) Temporal relationship between drug ingestion and clinical presentation, OR temporally-related recurrence after oral challenge |
| (5) Excellent response to treatment with systemic corticosteroids or potassium iodide | (E) Temporally-related resolution of lesions after drug withdrawal or treatment with systemic corticosteroids |
| (6) Abnormal laboratory values at presentation (three of four): erythrocyte sedimentation rate >20 mm/hr; positive C-reactive protein; >8,000 leukocytes; >70% neutrophils | |

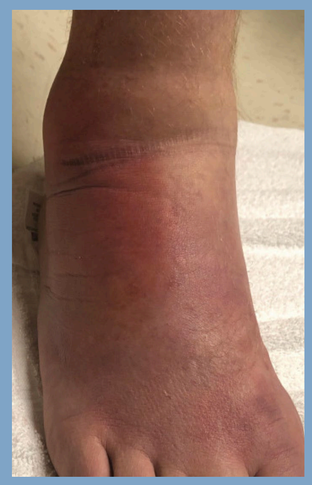
[a]The presence of both major criteria (1 and 2), and two of the four minor criteria (3, 4, 5, and 6) is required in order to establish the diagnosis of classical Sweet's syndrome; the patients with malignancy-associated Sweet's syndrome are included with the patients with classical Sweet's syndrome in this list of diagnostic criteria.

[b]All five criteria (A, B, C, D, and E) are required for the diagnosis of drug-induced Sweet's syndrome.

Ref: Cohen, Philip. Sweet's Syndrome-a comprehensive review of an acute febrile neutrophilic dermatosis. Orphanet J Rare Dis. 2007; 2: 34.







Disclosure Statement

*Authors of this presentation have the following to disclose concerning possible financial or personal relationships with commercial entities that may have a direct or indirect interest in the subject matter of this presentation: none*

## DISCUSSION

This case provides an excellent example of Sweet's Syndrome and its clinical considerations. A 44 year old male HIV positive patient presented with arthralgias, chills, malaise and a plaque-like rash on the back of his neck, head and wrist. The biopsy confirmed neutrophilic dermatosis consistent with Sweet's Syndrome. It is extremely important to consider the possible causes of Sweet's Syndrome, while the diagnosis is being established and treatment is initiated. Our patient was up to date on preventative cancer screening for his age, and thorough physical exam did not reveal any suspicion for occult malignancy. He had no antecedent or concurrent acute infection to precipitate Sweet's Syndrome. With respect to medications, Abacavir is known to cause neutrophilic dermatosis, but the patient had been on this medication for greater than one year, and there was not a strong temporal relationship between drug initiation and onset of symptoms. Additional evidence against Abacavir as a causative agent included no recurrence when prednisone was tapered even though patient was continued on Abacavir. There are few case reports of Sweet's Syndrome associated with HIV infection, but a causal relationship has not been established (3). Consideration was given to syphilis infection, but patient was previously treated and had no increase in his titers. The clinical opinion of our team and the consulting dermatologist was that this case was idiopathic.

## KEY LEARNING POINTS

Consider Sweet's Syndrome in the differential diagnosis of any patient who presents with the abrupt onset of painful erythematous papules, plaques or nodules.

**Major diagnostic criteria:**

1) Abrupt onset of painful erythematous plaques or nodules.
2) Histopathologic evidence of dense neutrophilic infiltrate without evidence of leukocytoclastic vasculitis.

**Minor diagnostic criteria (need two):**

1) Pyrexia >38C.
2) Association with underlying hematologic or visceral malignancy, inflammatory disease for pregnancy, or preceded by URI or vaccination.
3) Excellent response to treatment with systemic glucocorticoids or potassium iodide.
4) Abnormal lab studies

The mainstay of treatment is corticosteroids and improvement is often seen within 48 hours. Steroid dose is tapered over 4 to 6 weeks.

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