Sweet syndrome presenting as a febrile rash in a returning traveller

Alexander Hart, MBBS*; Katharina Plenk, MD†§; David Carr, MD, CCFP-EM‡¶

ABSTRACT

Sweet syndrome was discovered in 1964 and is now well described in the dermatology literature. Knowledge of this unique febrile and painful dermatosis is important for the emergency physician because the syndrome can be readily identified and is extremely responsive to oral steroid therapy. Early diagnosis can greatly improve patient satisfaction and avoid days of ineffective treatment. An accurate and timely diagnosis of Sweet syndrome is also important to guide investigation into a number of associated diseases.

RÉSUMÉ

Le syndrome de Sweet a été découvert en 1964 et il est très bien décrit maintenant dans la documentation médicale en dermatologie. La connaissance de cette dermatose fébrile et douloureuse, unique en son genre a de l'importance pour le médecin d'urgence parce que le syndrome est facilement reconnaissable et qu'il réagit extrêmement bien à corticothérapie orale. La pose précoce du diagnostic peut accroître grandement la satisfaction du patient, et éviter d'avoir à subir des traitements inefficaces durant des jours. En outre, il importe de procéder à une pose rapide et exacte du diagnostic du syndrome de Sweet, car cela permet d'orienter l'exploration vers certaines maladies associées.

Keywords: Sweet's syndrome

CASE REPORT

We report a case of Sweet syndrome presenting as a febrile rash in a traveller returning from Cuba. The diagnosis was confirmed on biopsy, and the patient had an excellent response to steroid therapy.

A 46-year-old woman with no previous medical history or known allergies presented to an emergency department with a severely painful rash and fever while on vacation in Cuba. The rash appeared as raised maculopapular lesions on her limbs and trunk with sparing of her head and neck. She had no myalgia, arthralgia, chest pain, shortness of breath, or abdominal pain. She denied sick contacts, ocean exposure, animal bites, sexual contacts, new medications, or intravenous (IV) drug use. Initial treatment with dimenhydrinate provided little relief.

On examination, the patient was hemodynamically stable, febrile, alert, and oriented. She was in a significant amount of pain requiring parenteral opioid therapy. Her physical exam was highlighted by an extremely tender, violaceous, confluent maculopapular rash over her extremities (Figure 1). There were no blisters or vesicles, and Nikolsky sign was negative. There was no mucous membrane or conjunctival involvement. She had a normal oropharynx and no adenopathy. Her neck was supple, her chest clear, and her abdomen was soft.

Initial laboratory findings included an elevated white blood count of 21.9, an absolute neutrophil count of 19.1, an elevated erythrocyte sedimentation rate (ESR) of 104, and a C-reactive protein (CRP) of 80. The remainder of her routine blood work was non-contributory.

The patient was admitted for pain management and additional investigations. Further workup, including blood cultures, hepatitis panel, venereal disease research laboratory (VDRL), human immunodeficiency virus (HIV), rheumatoid arthritis (RA), antinuclear antibody (ANA), dengue, and chikungunya serology were all negative. A skin biopsy demonstrated a nodular infiltrative neutrophilic dermatosis.

The presentation of symptoms and biopsy results were consistent with Sweet syndrome. The patient was started on methylprednisone, 75 mg IV daily during her 24-hour stay and discharged home on 40 mg of pre-

From the *Division of Emergency Medicine; †Department of Medicine; ‡Department of Family and Community Medicine, Division of Emergency Medicine, University of Toronto, Toronto, ON; §Department of Medicine, Mackenzie Health Hospital, Richmond Hill, ON; and the ¶Department of Emergency Medicine, University Health Network (UHN), Toronto, ON.

Correspondence to: Alexander Hart, Sunnybrook Health Sciences Centre, 2075 Bayview Ave, C7-53, Toronto, ON M4N 3M5; Email: alexhart86@gmail.com

CJEM 2018;20(3):476-478 DOI 10.1017/cem.2017.26

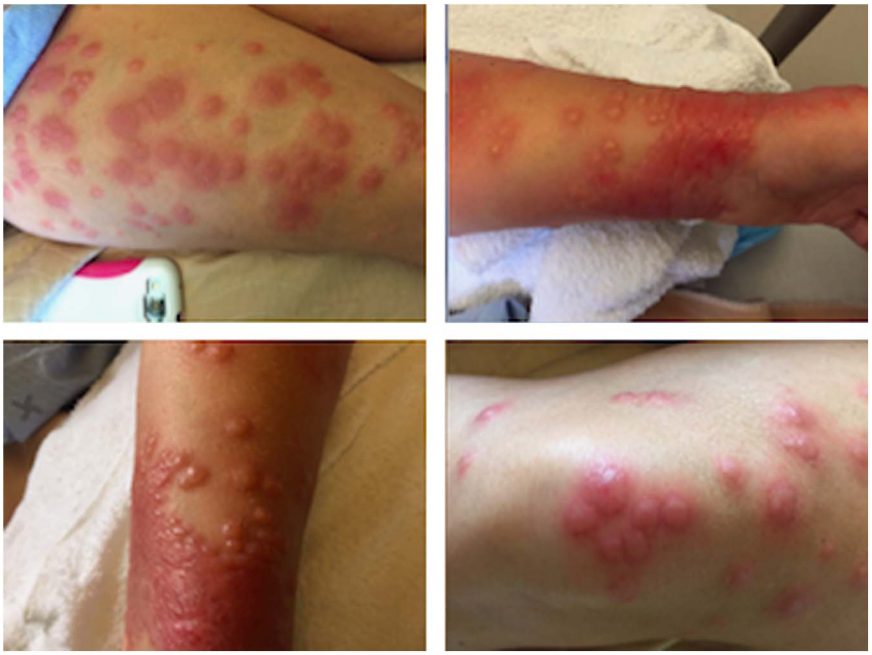

Figure 1. The rash as it appeared on presentation to the emergency department. It is raised and exquisitely tender to the touch. *Clockwise from top-left*: The patient's right thigh, left wrist, left knee, and left wrist again.

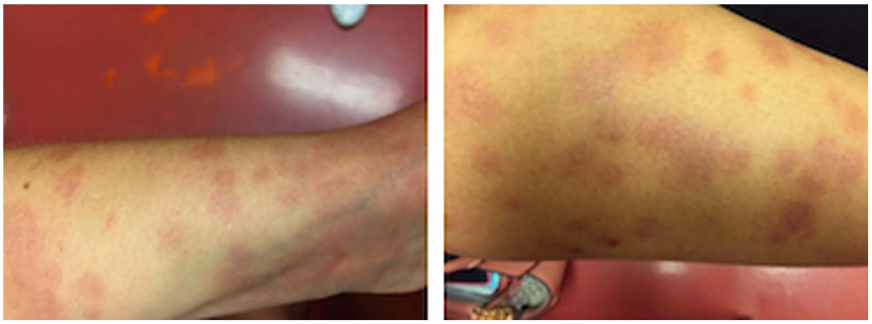

Figure 2. The same rash, as it appeared 1 month following initial steroid therapy. The lesions are flat and non-tender. *Left to right*: Patient's left leg, right thigh.

dnisone daily with specialist follow-up. The patient experienced rapid and complete resolution of her pain within 72 hours of starting therapy, and the rash faded over several weeks (Figure 2).

DISCUSSION

A patient presenting with an unusual rash can be diagnostically challenging to even seasoned emergency physicians. A rash arising in a febrile patient returning from the tropics can also be more concerning for zoonoses and other rare diseases that may cloud the clinical picture.

Avoiding such pitfalls is critical. Broadly speaking, most returning travellers with fever will not be suffering from a tropical illness. However, and more specific to our case, a rash presenting without pruritus but with frank pain requiring opiate analgesia is a rash not to be dismissed as benign.

Acute febrile neutrophilic dermatosis, better known as Sweet syndrome, falls under this spectrum. Although the confirmatory test is not available in the emergency setting, Sweet syndrome is a disease pattern that can be readily identified and effectively treated by the astute clinician.

First described in 1964,[1] reports of Sweet syndrome have been numerous in the literature worldwide and thus should not be relegated as diagnostic esoterica.[2-4] The disease arises with a heavy prevalence in women but does not appear to discriminate based on race or geography.[5] Patients can appear severely ill and present, as in our case, with significant pain and distress.

https://doi.org/10.1017/cem.2017.26 Published online by Cambridge University Press

Fever typically precedes a focal or disseminated erythematous to violaceous papular or vesicular rash. The syndrome is often associated with arthralgias, myalgias, and a sense of malaise.[6,7] It is not uncommon for this illness to be initially confused with infectious etiologies or, as with our patient, other more benign skin eruptions.[8] Failure to identify the true etiology leads to a delay in treatment and prolonged suffering.

Unfortunately, the symptoms of Sweet syndrome are as non-specific as its laboratory findings that include neutrophilia and a raised ESR and CRP. Sweet syndrome can be diagnosed only by a skin biopsy, and therefore it is imperative for the emergency department physician to facilitate an urgent biopsy.[5,9]

Sweet syndrome is a disease not to be missed, because it may be associated with undiagnosed underlying malignancies, other inflammatory conditions, or medications.[10,11] The patient in our report went on to receive a full age specific malignancy workup, including endoscopy and computed tomography scanning as a result of the strong paraneoplastic associations in patients with Sweet syndrome. The most common inciting pharmaceuticals include granulocyte colony stimulating factor (GCSF), retinoic acids, as well as certain antibiotics and anti-epileptics.[10] A thorough medication history should thus be taken because removing the offending agent is imperative.

As soon as the diagnosis is established on biopsy or recognized clinically, treatment is simple and effective. Beyond removing any offending agent and treating underlying causes, pharmacological therapy for Sweet syndrome consists of a course or oral prednisone 0.5 – 1.5 mg/kg/day with a 2- to 6-week taper. Although the rash may persist several days to weeks, the pain will begin to diminish rapidly as it did in 24 hours, in our case. Refractory cases can be treated with 3 days of pulsed high dose methylprednisolone at a dose of 1000 mg/day.[7,12] This case highlights several salient points for the emergency physician to consider. First, not all fevers in returning travellers are from tropical sources. Second, the constellation of signs and symptoms in Sweet syndrome, if recognized early and appropriately, can offer rapid and satisfactory relief to an otherwise suffering patient. Finally, any rash suspicious for Sweet syndrome merits close follow-up and should not be discharged without arranging proper malignancy-related investigations.

Competing interests: None declared.

REFERENCES

1. Sweet RD. An acute febrile neutrophilic dermatosis. *Br J Dermal* 1964;76:349-56.
2. Tasaduq F, Duncan D, Wani L. Sweet's syndrome. *Am J Med* 2010;123(8):694-6.
3. Dabade TS, Davis MDP. Diagnosis and treatment of the neutrophilic dermatoses (pyoderma gangrenosum, Sweet's syndrome). *Dermatol Ther* 2011;24:273-84.
4. Cohen PR, Kurzrock R. Sweet's syndrome: a neutrophilic dermatosis classically associated with acute onset and fever. *Clin Dermatol* 2000;18:265-82.
5. Abbas O, Libbi AG, Rubeiz N. Sweet's syndrome: retrospective study of clinical and histologic features of 44 cases from a tertiary care center. *Int J Dermatol* 2010; 49:1244-9.
6. Cohen PR, Kurzrock R. Sweet's syndrome revisited: a review of disease concepts. *Int J Dermatol* 2003;42:761-78.
7. Cohen PR, Kurzrock R. Sweet's syndrome: a review of current treatment options. *Am J Clin Dermatol* 2002; 3(2):117-31.
8. Withrow RA, Shenenberger D, Kalish VB, et al. Cellulitis unresponsive to antibiotics. *J Fam Practice* 2011;60(3): 149-51.
9. Anzalone CL, Cohen PR. Acute febrile neutrophilic dermatosis (Sweet's syndrome). *Curr Opin Hematol* 2013; 20:26-35.
10. Maalouf D, Battistella M, Bouaziz JD. Neutrophilic dermatosis: disease mechanism and treatment. *Curr Opin Hematol* 2015;22:23-9.
11. Paydas S. Sweet's syndrome: a revisit for hematologist and oncologists. *Brit Rev Oncol Hematol* 2013;86:85-95.
12. Schadt CR, Callen JP. Management of neutrophilic dermatoses. *Dermatol Ther* 2012;25:158-72.

https://doi.org/10.1017/cem.2017.26 Published online by Cambridge University Press