

Providence St. Joseph Health

## Providence St. Joseph Health Digital Commons

---

Providence St. Vincent Internal Medicine

Oregon Academic Achievement

---

4-29-2020

### Unpainfully Sweet

Khoi Nguyen

*Providence St. Vincent, Internal Medicine Residency, Portland, Oregon, [Khoi.Nguyen2@providence.org](mailto:Khoi.Nguyen2@providence.org)*

Alex Schafir

*Providence St. Vincent, Internal Medicine Residency, Portland, Oregon, [alexander.schafir@providence.org](mailto:alexander.schafir@providence.org)*

Follow this and additional works at: [https://digitalcommons.psjhealth.org/psv\\_internal](https://digitalcommons.psjhealth.org/psv_internal)



Part of the [Internal Medicine Commons](#)

---

#### Recommended Citation

Nguyen, Khoi and Schafir, Alex, "Unpainfully Sweet" (2020). *Providence St. Vincent Internal Medicine*. 1. [https://digitalcommons.psjhealth.org/psv\\_internal/1](https://digitalcommons.psjhealth.org/psv_internal/1)

This Poster is brought to you for free and open access by the Oregon Academic Achievement at Providence St. Joseph Health Digital Commons. It has been accepted for inclusion in Providence St. Vincent Internal Medicine by an authorized administrator of Providence St. Joseph Health Digital Commons. For more information, please contact [digitalcommons@providence.org](mailto:digitalcommons@providence.org).



# Unpainfully Sweet

Khoi Nguyen, DO and Alex Schafir, MD FACP

Providence St. Vincent, Internal Medicine Residency, Portland, Oregon

## INTRODUCTION

**Sweet's syndrome (ss), or acute febrile neutrophilic dermatosis,** is characterized by sudden onset of fever, leukocytosis and erythematous plaques or nodules infiltrated by neutrophils. There are three main clinical settings in which Sweet's syndrome has been described:

Classical SS	Drug Induced SS	Malignancy Associated SS
<ul style="list-style-type: none"> <li>➤ Infection (URI and GI)</li> <li>➤ Inflammatory (UC and Crohns)</li> <li>➤ Pregnancy</li> </ul>	<ul style="list-style-type: none"> <li>➤ Granulocyte-colony stimulating factor</li> <li>➤ Furosemide, Nitrofurantoin, Bactrim</li> </ul>	<ul style="list-style-type: none"> <li>➤ Hematologic (AML)</li> <li>➤ Solid (Breast, GU, GI)</li> </ul>

The diagnosis of Sweet's syndrome often has a temporal association with the discovery or relapse of cancer as reported in this case.

## CASE REPORT

A 64-year-old woman with recurrent breast cancer on targeted and hormonal therapy presented to the ED with acute onset of fever and diffuse non-tender body rash.

Vitals suggested sepsis and she was treated with broad spectrum antibiotics. A chest xray, viral panel, blood cultures, echocardiogram and urinalysis were unrevealing.

Erythrocyte sedimentation rate and C-reactive protein were elevated. There were 90% neutrophils.

When her erythematous plaques became vesicular, she was treated with IV Acyclovir for possible disseminated zoster. This was discontinued when biopsy for HSV/HZV returned negative.

Skin biopsy, however, showed dense neutrophilic infiltration consistent with Sweet Syndrome. Treatment with prednisone resulted in rapid clinical improvement.

Major Criteria:
<ul style="list-style-type: none"> <li>➤ Abrupt onset of painful erythematous plaques or nodules</li> <li>➤ Histopathologic evidence of a dense neutrophilic infiltrate without evidence of leukocytoclastic vasculitis</li> </ul>
Minor Criteria:
<ul style="list-style-type: none"> <li>➤ Pyrexia &gt;38° C</li> <li>➤ Association with underlying hematologic or visceral malignancy, inflammatory disease or pregnancy, OR preceded by upper respiratory infection, gastrointestinal infection, or vaccination</li> <li>➤ Excellent response to treatment with systemic glucocorticoids or potassium iodide</li> <li>➤ Abnormal laboratory values at presentation (three of four of the following: erythrocyte sedimentation rate &gt;20 mm/hour, positive C-reactive protein, &gt;8000 leukocytes, &gt;70 percent neutrophils)</li> </ul>

## Cutaneous Lesions and Histology

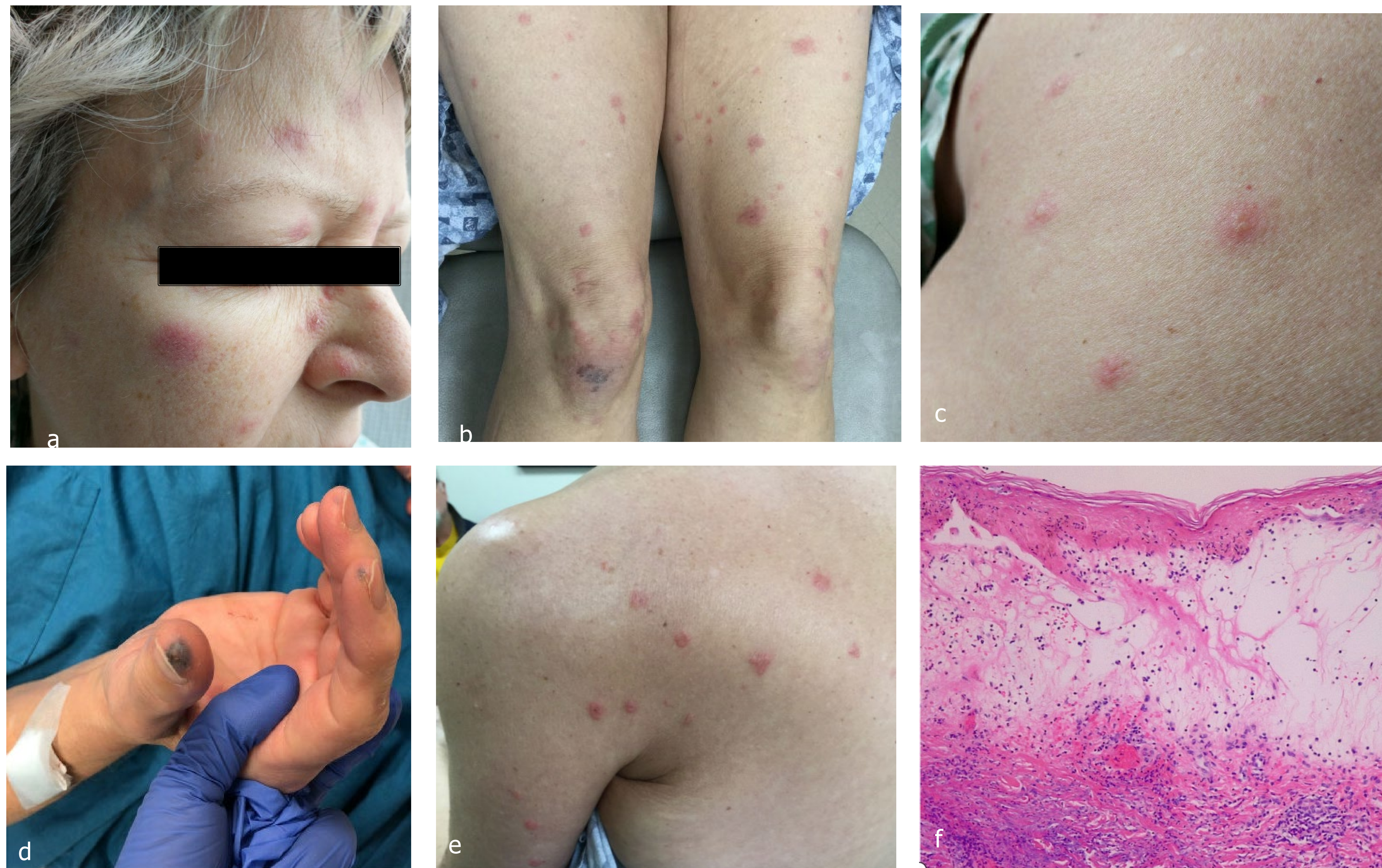


Figure a-e. Presenting lesions

Figure f. Biopsy showing neutrophilic infiltrate.

## DISCUSSION

Historically, the diagnosis of SS requires the presence of painful erythematous lesions (both Major Criteria and two of four Minor Criteria). The absence of pain and tenderness in this patient made the diagnosis challenging. After extensive evaluation to rule out infectious etiology, it was revealed through skin biopsy that her syndrome was more consistent with Sweet Syndrome despite having non-tender lesions. Given her recent recurrence of breast cancer, it was thought that her SS was more consistent with Malignancy-Associated SS (MASS) subtype.

MASS is most commonly associated with hematological malignancies such as AML, but also occurs coincident with solid tumors, such as carcinomas of GU organs, breast, and GI tract. In patients with a previous history of cancer, the diagnosis of SS usually heralds the onset of recurrence. Sweet syndrome may precede, follow, or appear concurrently with a malignancy. This patient was found to have a recurrence of breast cancer in the form of bone metastases two months prior to admission.

In summary, this case demonstrates an atypical painless presentation of SS and how evaluation for malignancy is indicated for patients with SS, particularly when there is an absence of other explanations such as recent infection, inflammatory disease or drug exposure.

Treatment : Prednisone 0.5mg to 1mg/kg per day. Symptoms usually improve within 48 hours and skin lesions resolves within 1 to 2 weeks. Steroid is then taper over course of 4 to 6 weeks.

## REFERENCES

1. Wite JM, Mufti GJ, Salisbury JR, du Vivier AW. Cutaneous manifestations of granulocyte colony-stimulating factor. Clin Exp Dermatol 2006; 31:206
2. Von den Driesch P. Sweet's syndrome (acute febrile neutrophilic dermatosis). J Am Acad Dermatol 1994; 31:535
3. Cohen PR. Sweet's syndrome – a comprehensive review of an acute febrile neutrophilic dermatosis. Orphanet J Rare Dis 2007; 2:34
4. Cohen PR, Kurzrock R. Sweet's syndrome and cancer. Clin Dermatol 1993; 11:149
5. Raza S, Kirkland RS, Patel AA, et al. Insight into Sweet's syndrome and associated malignancy: a review of current literature. Int J Oncol 2013; 42:1516