

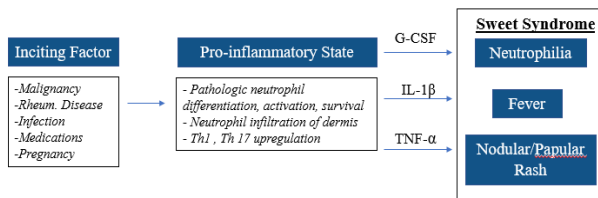
# Sweet Syndrome: A Rare Mimicker of Septic Shock

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## Introduction

- Sweet syndrome (SS) is a rare inflammatory condition characterized by abrupt onset fever, neutrophilia and painful erythematous papules / nodules thought to be related to maladaptive elevations in G-CSF, IL-1 $\beta$  and TNF- $\alpha$ .
- SS can be idiopathic, or can be associated underlying inflammatory condition, infection, malignancy or medication reaction. Underlying causes must be investigated.
- Extracutaneous manifestations can occur, including pleural and pericardial effusions; renal, hepatic and pancreatic insufficiencies; and neuropsychiatric changes.
- Rarely, SS can cause disseminated inflammation (dSS) with multisystem inflammatory involvement, and hemodynamic instability, mimicking septic shock.
- Treatment of dSS with high-dose systemic steroids typically leads to rapid clinical improvement.



1A

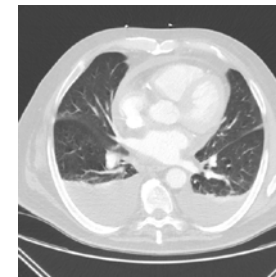


1B

**Figure 1:** Upper extremity bullae over the course of hospitalization. A) Lesions at time of first eruption. B) Lesions at day 7, during his subsequent hospitalization. Images document skin findings prior to treatment with corticosteroids.

## Case Presentation

- 74 year-old man with CAD, HTN and admission two weeks prior for cholangitis with biliary tube placement re-presented with fevers, generalized weakness, and subacute progressive abdominal distension with pain in setting of serosanguinous biliary tube output.
- On his first admission, patient was anemic requiring 2 U PPRBC. He developed TACO requiring ICU admission. In the ICU, developed scattered joint stiffness with elevated ESR (>130) and CRP (233), with an acute eruption of hemorrhagic blisters on his bilateral hands (**Fig. 1A**).
- Biopsy revealed intraepidermal neutrophilic dermatosis with massive dermal edema, consistent with possible SS. He was discharged home without antibiotics.
- One day after discharge, he presented to an outside hospital with presumed septic shock, prompting transfer to SMH. Sepsis secondary to a biliary source was suspected and he was treated with vancomycin, cefepime and metronidazole.
- Course was complicated by:
  - Acute hypoxic respiratory failure with persistent bilateral pleural effusions requiring BiPAP.
  - Transfusion-dependent anemia with smear revealing dysplastic myeloid cells - poikilocytic erythrocytes, hypogranular neutrophils and large platelets
  - New onset anxiety, possible hallucinations, acute kidney injury
- Imaging of chest, abdomen, pelvis, oral cavity did not reveal source of infection. Blood, urine, biliary tube cultures sterile.
- Hand bullae persisted. (**Fig. 1B**). Without identified infectious source, presentation was thought to be consistent with dSS. Dermatology recommended methylprednisolone.
- Significant improvement in skin lesions and respiratory status following steroid initiation.
- Bone marrow biopsy revealed MDS vs developing AML, likely driver of his anemia and dSS. With news of diagnosis, patient and family elected comfort care. He died 4 days after discharge.



**Figure 2:** CT angiogram obtained at time of admission revealed a small pericardial effusion and bilateral pleural effusions - known associations with extracutaneous SS.

## Conclusions

- Consider dSS in patients with fever, painful erythematous papules/nodules, anemia, effusions and hemodynamic instability meeting SIRS criteria with unclear source of infection. dSS may present similarly to septic shock.
- Early treatment with high dose corticosteroids is critical to clinical improvement, which should be evident within the first three days.
- Many cases of SS are due to underlying inflammation, thus must evaluate for underlying infection, rheumatological disease or malignancy.
- Definitive management requires treatment of the underlying inflammatory condition, if an etiology can be identified.

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