

Sweet Syndrome: A rare mimicker of septic shock

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Introduction:

Sweet syndrome (SS), or acute febrile neutrophilic dermatosis, is a rare inflammatory condition with pathogenesis thought to be related to maladaptive elevations in G-CSF. SS is characterized by abrupt development of fever, neutrophilia and painful erythematous papules/ nodules, commonly found on the dorsal upper extremities. SS is often associated with underlying- or post-infectious states, pregnancy, IBD, drug reactions, or malignancy. Extracutaneous manifestations may occur, including pleural and pericardial effusions; renal, hepatic and pancreatic insufficiencies; and neuropsychiatric changes. Rarely SS can present as disseminated Sweet Syndrome (dSS) with multisystem inflammatory involvement and hemodynamic instability mimicking septic shock, although there will be no underlying infectious cause or improvement with antibiotics. Treatment of dSS with systemic steroids typically leads to clinical improvement.

Case Description:

A 74 year-old man with coronary artery disease and hypertension who was two weeks post admission for cholangitis with biliary tube placement and ATN, re-presented to our institution with fevers, weakness, subacute progressive abdominal distension and diffuse abdominal pain in the setting of serosanguineous biliary tube output. He was anemic and required 2U PRBC. He developed hypoxia with signs of possible transfusion associated circulatory overload and required MICU admission for respiratory support and diuresis. During this hospital stay, he developed scattered joint stiffness with elevated ESR and CRP, with an acute eruption of hemorrhagic blisters on his bilateral hands. A biopsy revealed neutrophilic dermatosis with massive dermal edema, consistent with SS. He was stabilized and discharged home.

One day following discharge, he re-presented with presumed septic shock from a biliary source, with subsequent hospital course further complicated by transfusion-dependent anemia, hypoxic respiratory failure, acute kidney injury, and persistent bilateral hand lesions. He was started on broad spectrum antibiotics, including vancomycin, cefepime and metronidazole, and underwent extensive workup. Imaging revealed bilateral pleural and pericardial effusions, though cultures and imaging revealed no clear source of infection. dSS was felt to be a unifying diagnosis, and antibiotics were discontinued in favor of methylprednisolone, which improved his systemic symptoms. He underwent a bone marrow biopsy revealing MDS versus developing AML. He chose to pursue hospice care and died 4 days after discharge.

Discussion:

dSS should be considered in patients who have acute onset of fever with painful erythematous papules/nodules, anemia, effusions and symptoms consistent with sepsis without a clear infectious source. Paramount is prompt exclusion of infectious causes before initiation of

steroids, as delays in corticosteroid treatment can lead to significant morbidity. Importantly, SS is often associated with an underlying malignancy, most commonly hematological, so further evaluation should be pursued. Treatment of the underlying malignancy is definitive therapy for SS.