Sweet syndrome: a hint to underlying malignancy
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Background

- Sweet syndrome, also known as acute febrile neutrophilic dermatosis.
- It is a rare inflammatory condition characterized by sudden erythematous skin lesions, fever, leukocytosis with neutrophilia, and neutrophilic dermal infiltrates into the papillary dermis on biopsy.
- It accounts for 10–15 percent of autoimmune disorders in Myelodysplastic Syndrome (MDS) patients and we present one such interesting clinical case.

Case Presentation

- A 56-year-old gentleman with history of pulmonary embolism, and anemia presented with worsening skin and oral ulcers for one week and fever and chills for three days.
- He developed a maculopapular rash three months ago that was initially responsive to steroids. However, since stopping the steroids, he has developed skin and oral ulcers that have progressed and gotten worse.
- Physical examination revealed multiple nonbleeding oral ulcers and multiple foul-smelling necrotic skin ulcers primarily on the abdomen, chest, upper extremities, neck, and groin with pustular base and discharge.
- Evaluation of larynx revealed epiglottis, vocal fold edema, and a white lesion on the left vocal fold.
- Debridement of wounds revealed necrotic tissue with focal acute severe inflammation and focal fat necrosis with Pseudomonas aeruginosa and Staphylococcus epidermidis.

Work Up

- Initial labs: Macrocytic anemia with reticulocyte count of 11.1%, leukocytosis with neutrophilia and bands.
- Dermatopathology: Subcorneal fistulous and focal epidermal ulceration, as well as a dense diffuse dermal neutrophilic crypt infiltrate with negative immunofluorescence, consistent with Sweet Syndrome.
- Bone biopsy: Hyper cellular bone marrow with myeloid predominance and significantly decreased erythroid and megakaryocyte counts.
- Cytogenetics/FISH analysis: Chromosome 12 deletion and chromosome 3 translocation, consistent with MDS.

Discussion

- A diagnosis of sweet syndrome secondary to MDS was made, however hospital course was complicated with septic shock and acute hypoxic respiratory failure from recurrent aspirations caused by oral ulcers.
- Given rapid clinic deterioration, patient was made comfort measures only per patient and family wishes and he died shortly thereafter.

Hospital Course

- Sweet syndrome is difficult to diagnose as it requires meticulous integration of clinical and laboratory data.
- Early detection and treatment are critical in malignancy-associated sweet syndrome.
- The primary management is systemic corticosteroid therapy with pulsed intravenous glucocorticoids and immunomodulatory therapies in refractory cases, in addition to treating the underlying malignancy.