

## A severe case of steroid recalcitrant Sweet syndrome

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**Introduction:** High-dose systemic steroids are the first line and most common treatment for Sweet Syndrome. Management strategies are difficult and not well established when systemic steroids alone are ineffective.

**Case Description:** A 58-year-old man with no significant past medical history was admitted to the hospital with fevers, and diffuse erythematous nodules and large necrotic plaques of the bilateral arms. He underwent two skin biopsies both of which showed dense dermal neutrophilic infiltrates consistent with Sweet syndrome. He subsequently underwent an extensive workup for underlying etiologies and was found to have myelodysplastic syndrome on bone marrow biopsy, for which he was initiated on decitabine. For his cutaneous findings, he was started on high-dose steroids with 1 mg/kg of PO prednisone equivalent which was up-titrated to a maximum dose of IV solumedrol 120 mg daily. After several days of high-dose steroids, his necrotic plaques continued to progress, and he developed oral lesions and new oxygen requirements with concern for pulmonary Sweet syndrome. He was then started on super saturated potassium iodide (max dose of 500 mg TID) with continued progression of skin lesions. One dose of infliximab was given at 10 mg/kg with continued development of worsening and new lesions. At this time, after multidisciplinary discussions, repeat bone marrow and skin biopsies were performed which re-confirmed the myelodysplastic and Sweet syndrome diagnoses. He was then started on pulse steroids at 1 gram of solumedrol for three days as well as anakinra at 100 mg SQ daily. In the subsequent days to weeks, he began to have slow stabilization and improvement of his skin lesions and systemic symptoms (Figure 1).

**Summary:** Here we present a rare and severe case of steroid recalcitrant Sweet syndrome which slowly stabilized after addition of pulse steroids, super saturated potassium iodide, infliximab, and anakinra. This case highlights the complex management strategies for steroid recalcitrant Sweet syndrome.

**Figure 1: Timeline showing progression of skin lesions with treatment modalities.**



### Legend

**A:** Necrotic plaques on the left arm at the start of Sweet syndrome treatment. **B:** Worsening necrotic plaques now involving the right arm. **C:** Development of new violaceous papules and plaques on the face. **D:** Flattening of violaceous papules and plaques on the face. **E:** Start of healing with formation of granulation tissue on the bilateral arms.

*Abbreviations:* D: "Day" and the following number indicates number of days since the start of treatment for Sweet syndrome. SSKI: Super saturated potassium iodide. SQ: Subcutaneous.