Histiocytoid Bullous Sweet Syndrome: A Case Presentation and Discussion

Dr. Derek Hirschman, DO
Department of Dermatology ■ Beaumont Hospital, Farmington Hills, MI

INTRODUCTION

Histiocytoid Sweet Syndrome (HSS) is a rare inflammatory cutaneous disease classified as a histological variant of Sweet’s Syndrome (SS) also known as acute febrile neutrophilic dermatosis. Described in 1964 by Robert Douglas Sweet, SS often presents as a sudden onset of painful erythematous plaques and nodules associated with fever, leukocytosis, and neutrophilia. Clinically, lesions have dense diffuse infiltration of mature neutrophils with papillary dermal edema. In 2005, Requena, with colleagues, classified HSS as a variation of SS. Although the two are undistinguishable clinically, microscopically the diseases show infiltrate of different cell lineage.

The infiltrate of HSS consists primarily of immature myeloid cells that histologically resemble histiocytes.

CASE REPORT

Patient is a 62-year-old Jordanian female who presented with an acute onset of painful, pink-to-flesh colored, edematous annular plaques with associated hemorrhagic bullae affecting mainly the bilateral ventral forearms and bilateral hands. Clinical presentation is illustrated in Figures 1-3. Patient also had scattered erythematous, asymptomatic papules throughout much of her lower back. Patient was previously healthy with a past medical history of hypothyroidism for many years well controlled with levothyroxine. Patient admitted to relatively severe pain in bilateral hands, as well as associated fever and chills for 3 days prior to presentation. Patient had three punch biopsies done (two for H&E and one for DIF) with a differential diagnosis of bullous sweet syndrome, bullous erythema multiforme, bullous pemphigoid, and LABD. Patient was treated with an intramuscular injection of 60mg of triamcinolone hexacetonide and 4mg of dexamethasone sodium phosphate, along with topical triamcinolone 0.1% cream. Patient was seen 1 week later and had significant improvement in both pain and in lesion severity with only mild post-inflammatory erythema noted. Pathology showed a lichenoid infiltrate of histiocytoid mononuclear cells within the upper dermis in conjunction with a few collections of neutrophils and focal marked papillary edema and spongiotic vessels. IHC stains for myeloperoxidase stained positive the scattered neutrophils as well as the histiocytoid mononuclear cells. Direct immunofluorescence revealed no significant immune complex deposition. Based on the histology and clinical presentation, the diagnosis of histiocytoid bullous sweet syndrome was made.

No further workup was deemed necessary by the hematology/oncology department at this time. Patient was treated with a one month tapering dose of prednisone with the eventual addition of dapsone. At approximately 3 months post initial presentation, patient remains under good control with no lesions on 25mg of dapsone alone.

DISCUSSION

The pathogenesis of SS continues to be unknown, but SS often presents in three clinical settings: classical (or idiopathic), drug induced, or malignancy associated (especially myeloid leukemias and myelodysplastic syndrome). The classic form often most commonly occurs in healthy, middle-aged (30-60) females and may be associated with preceding infection (URI, strep., etc.), connective tissue disease, inflammatory bowel disease, or even pregnancy. Initial published data suggested that HSS typically behaved in a benign fashion and lacked an association with possible underlying malignancy. However, more recent case series suggest that between 36%-53% of HSS cases are in fact associated with some hematologic disorder, again, most commonly myelodysplastic syndrome and myelogenous leukemias. Both HSS and SS prognosis seem to be predominantly dictated by the medical associated conditions, instead of the skin disease itself. Sweet’s syndrome may recur following either spontaneous remission or secondary to treatment induced clinical resolution and the duration of remission is variable depending on the underlying cause. Typically classical cases show less risk of recurrence whereas, in cancer patients, Sweet’s syndrome recurrences are more common. Systemic corticosteroids are the first-line therapies for both SS and HSS and rapid clearance of skin lesions after steroid initiation is one of the minor criteria for diagnosing SS.

CONCLUSION

Presented is a case report of classic histiocytoid sweet syndrome (clinically bullous variant) with an unknown etiology that showed rapid clearance with systemic corticosteroids and sustained clearance with dapsone. Despite the negative workup in this patient, due to the possible association with hematologic malignancy, it is important to screen all patients diagnosed with SS or HSS.

REFERENCES