Sweet syndrome: Clinical presentation, associations, and response to treatment in 77 patients

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Background: Sweet syndrome is a neutrophilic dermatosis with cutaneous tender lesions that can be associated with malignancies, infections, systemic inflammatory disorders, and medications. Although numerous studies have described Sweet syndrome, few studies have systematically investigated Sweet syndrome.

Objective: We sought to describe characteristics and treatments of patients with Sweet syndrome and evaluate clinical differences depending on the underlying cause.

Methods: A retrospective study was conducted to identify patients with Sweet syndrome evaluated at Mayo Clinic from 1992 to 2010.

Results: Of 77 patients with Sweet syndrome (mean age of onset 57 years), 43 (56%) were male. Eighteen patients (23%) reported a preceding infection. A total of 41 (53%) patients were classified as having classic Sweet syndrome, 27 (35%) patients had malignancy-associated Sweet syndrome, and in 9 (12%) patients drug-induced Sweet syndrome was considered. In all, 21 patients had a hematologic malignancy or myeloproliferative/myelodysplastic disorder, whereas 6 patients had solid tumors. The mean hemoglobin level, in both male and female patients ($P < .0443$ and $P < .0035$, respectively), was significantly lower in malignancy-associated versus classic and drug-induced Sweet syndrome. Systemic corticosteroids were the most frequently used treatment (70%).

Limitations: This is a retrospective study and represents patients from a single academic center.

Conclusions: Sweet syndrome is a distinctive disorder with certain clinical and histologic characteristics, which usually has a complete response to systemic corticosteroids. It is important to evaluate Sweet syndrome patients who have laboratory evidence of anemia for an underlying malignancy. (J Am Acad Dermatol 2013;69:557-64.)

Key words: drug-induced Sweet syndrome; malignancy-associated Sweet syndrome; Sweet syndrome; treatment.

Acute febrile neutrophilic dermatosis, also known as Sweet syndrome, was originally described in 1964 by Robert D. Sweet.[1] Years later, Su and Liu[2] proposed several criteria to be implemented as guidelines for the diagnosis of Sweet syndrome. After having been further modified in 1989[3] with the description of additional cases (Table I), these criteria continue to be used in clinical practice. Sweet syndrome is a skin condition characterized by the following features: tender

Abbreviations used:

MDS: myelodysplastic syndrome
MPD: myeloproliferative disorder

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erythematous plaques, nodules, or papules; prodromal symptoms such as fever, malaise, or arthralgia; and diffuse infiltrate of neutrophils in the papillary dermis.

Sweet syndrome can be further classified into classic, malignancy-associated, and drug-induced Sweet syndrome, depending on the clinical setting in which the disease develops. Classic or idiopathic Sweet syndrome is characterized by the abrupt onset of tender erythematous skin lesions and histopathological findings of neutrophilic infiltrate in the dermis, without evidence of vasculitis. Patients may present with prodromal symptoms of fever or known infection before the appearance of skin lesions. The cutaneous manifestations can also present concurrently with fever, arthralgia, generalized malaise, or conjunctivitis. The predominant laboratory finding that can be encounter in these patients is leukocytosis, an increased erythrocyte sedimentation rate, or both.[3]

CAPSULE SUMMARY

- This article identifies and describes a large cohort of patients with Sweet syndrome who sought treatment.
- Typically, patients had a complete response to treatment, with a low recurrence rate that did not vary regardless of the cause.
- This study demonstrates the importance of doing an extensive workup for malignancy in patients who present with Sweet syndrome and anemia.

Numerous retrospective reviews and case reports have supported a strong association between Sweet syndrome and malignancies. Acute myelogenous leukemia is considered the most common hematologic malignancy associated with this condition, and genitourinary tumors are the most frequent solid tumor cancers in patients with cancer-associated Sweet syndrome.[4,5] Another clinical setting that can be encountered when evaluating patients with Sweet syndrome is the onset of the disease caused by the use of certain medications. A temporal relationship between drug ingestion and clinical presentations of Sweet syndrome should be established to classify a patient as having drug-induced Sweet syndrome.[6]

Our objective was to report the experience of a single referral institution with a large series of patients with Sweet syndrome, and review the demographics, clinical presentation, associated diseases, treatments, and outcomes of these patients.

METHODS

Identification of patients and data collection

After institutional review board approval, we used the institutional patient database to identify the records of patients with a diagnosis of Sweet syndrome seen at Mayo Clinic, Rochester, MN, between 1992 and 2010. Patients who denied research authorization for the reviewed of their medical record were excluded from the study. We initially identified 113 patients with a possible diagnosis of Sweet syndrome; 2 reviewers examined the medical records of these patients to confirm the cases met the criteria for diagnosis. Of the 113 cases identified, 36 were excluded: 5 cases had no lesions present at the time of initial evaluation at the Mayo Clinic; 21 cases had features that were not consistent with Sweet syndrome, either through histopathological or clinical findings; and 10 cases had a revised diagnosis upon evaluation of the medical charts (7 cases of vasculitis, 1 case of erythema nodosum, and 2 cases of pyoderma gangrenosum). We therefore included in our series 77 patients whose final assessment was consistent with the diagnosis of Sweet syndrome based on Su and Liu's proposed criteria.[2,3]

We abstracted the following data from the medical records: patient demographic information, location of lesions, description of lesions, clinical characteristics of Sweet syndrome, histopathologic findings, preceding infection, associated medications, malignancies or any other concurrent medical condition previously identified to be associated with Sweet syndrome, laboratory data, treatment used for Sweet syndrome, and response to treatment. All cases found to be associated with a malignancy had to meet postulates of Curth,[7] which define the criteria for establishing a relation between an internal malignancy and a cutaneous disorder.

Response to therapy

Patient response to therapy was classified as complete response, partial response, or no response.

Statistical analysis

Continuous variables were summarized with means, medians, and ranges. Categorical variables were reported using proportions and percentages. Comparison among variables of interest and the classification of Sweet syndrome were evaluated by using Wilcoxon rank sum and χ^2 tests. All tests were 2-sided, and P values less than .05 were considered statistically significant.