Sweet's Syndrome and Malignancy: A Case Associated with Multiple Oral Squamous-Cell Carcinoma

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Key Words

Sweet's syndrome, oral

Malignancy

Squamous-cell carcinoma

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Sweet's syndrome (SS) or acute febrile neutrophilic dermatosis [1] as marker of malignancy was suggested by Clemmensen et al. [2] in 1989. These patients show some of the severer cutaneous features of the disorder. The most relevant findings in this group of patients are: male predominance; vesicular, bullous and even ulcerative skin lesions; lower extremity, trunk and back location; oral mucous membrane involvement; anemia (83%); abnormal platelet counts (68%); absence of neutrophilia (53%); absence of pyrexia (12%); extracutaneous involvement, particularly of the musculoskeletal and renal systems, and frequent recurrence of skin symptoms. SS usually precedes the diagnosis of the tumor or its recurrence [3]. Corticosteroid therapy is normally very effective regardless of the presence of malignancy.

10-20% of patients with SS have an associated neoplasm [4]. More than 85% of individuals with malignancy-associated SS had a hematologic disorder, particularly acute myelogenous leukemia, which occurs in 42% of patients. Solid tumors were observed in 15% of malignancy-associated SS. Hematologic malignancy together with a solid tumor has been described in few patients with malignancy-associated SS.

The clinical update of SS requires the description of new peculiar cases. We report the first case of severe SS with multiple oral squamous-cell carcinoma and its clinical aspects, course and treatment.

On August 9, 1993, a 48-year-old white man with a history of alcohol abuse and tobacco habit suffered the sudden development of multiple, erythematous and tender plaques and nodules that appeared edematous and somewhat vesicular. Some had a circinate pattern with central clearing and a peripheral vesicular ring (fig. 1). Some lesions formed ulcers and crusts. There were no mucosal abnormalities. Within a short time, the lesions spread from the upper extremities to the trunk, neck and face. The patient had malaise and fever. No weight loss was noted. Laboratory

findings showed macrocytic hypochromic anemia, ANCA negative, and no neutrophilia. Hematologic findings did not suggest a hematologic neoplasm. Corticosteroid therapy and intravenous antibiotics were prescribed. Skin biopsy revealed marked neutrophilic infiltration with the presence of interstitial and perivascular lymphocytes in the papillary and superficial reticular dermis. The epidermis showed acanthosis and focal parakeratosis. No evi-

Fig. 1. Bluish erythematous, tender and edematous plaques and some-what vesicular nodules that evolved to form ulcers and crusts and spread from the upper extremities to the trunk, neck and face.

dence of leukocytoclastic vasculitis could be found. Because blood and skin cultures were negative for infection, antibiotic therapy was withdrawn.

The patient was discharged from the hospital 5 days later because of dramatic improvement of the cutaneous lesions with corticosteroid therapy. Two months after, the patient complained of subcutaneous nodules on the left side of the submandibular region. Physical examination revealed 2 tumors; one was located on the right side of the soft palate and the other on the left side in the mouth floor. Biopsies were performed and both showed squamous-cell carcinoma (T,N < > M., stage I for the soft palate tumor and T,N,M., stage III for the mouth floor tumor according to the AJCC classification). In November 1993,

radiotherapy was initiated and a complete remission of both lesions was obtained.

In June 1994, new cutaneous lesions suddenly appeared. Physical examination demonstrated a relapse of the mouth floor squamous-cell carcinoma and the growth of a new lesion in the retromolar region. In the follow-up, a second retromolar tumor was observed. Squamous-cell carcinoma was diagnosed. Radical treatment was refused by the patient.

SS has been reported in association with a variety of systemic conditions such as cancer (hematologic malignancies, myelodysplastic syndromes and solid tumors), infections (bacterial, fungal and protozoan), inflammatory bowel disease (Crohn's disease and ulcerative colitis), rheumatologic diseases (Behçefs disease, drug-induced lupus erythematosus and rheumatoid arthritis), miscellaneous disorders (Dressler's syndrome, immunizing agents, subacute thyroiditis, sarcoidosis and POEMS syndrome), drugs (G-CSF and minocycline) and pregnancy. The main malignancy associations with SS are hematologic disorders, especially leukemias, although many others have also been described.

A few cases of SS are associated with solid tumors [4-6]. Cohen et al. [4] showed that malignancy-associated SS has been observed in 41 individuals with solid tumors. The initial appearance of SS lesions preceded or occurred concurrently (within 1 month) with the detection of an unsuspected solid tumor in at least 42% of patients. In our case, SS was the first sign of a neoplasm and a marker for recurrence.

A very complete list of solid tumors associated with SS is well documented in the literature: breast, thyroid, cervix, larynx (1 case) or vagina carcinomas and superficial spreading malignant melanoma. Very few patients with SS and solid tumors had also an associated hematologic malignancy.

Like our patient, most individuals with solid tumors have cutaneous SS lesions on the upper extremities. Cutaneous lesions are also present, on the head and neck, lower extremities, trunk and back. Similar to patients with idiopathic SS, but in contrast to those with hematologic-malignancy-associated SS, oral lesions are rare in SS associated with a solid tumor. Like those with solid tumors, our patient also presented fever during the episodes of SS. No extracutaneous manifestations of SS were present during the episodes in contrast to those patients with hematologic malignancy. The patient suffered from recurrent episodes of SS with the disease progression. The absence of neutrophilia does not exclude a diagnosis of SS in neoplastic patients.

In patients with SS, evaluation for malignancy, hematologic neoplasm and solid tumor is required. SS can be considered as a marker of progression of neoplastic diseases.

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Dermatology 1996;193:155

Measurement of Actinic Erythema in Healthy Subjects and in Subjects with Polymorphous Light Eruption Using a Tristimulus Colorimeter

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Key Words

Actinic erythema · Polymorphous light eruption ·

Tristimulus colorimeter

Garigue et al. [1] recently administered routine minimal erythema doses to 72 patients. They then used a tristimulus colorimeter to measure the erythema in 3 different modes: L*, a* and b*. Mode L* corresponds to the sample luminance envisioned as a point along a black-white axis, whereas a* and b* are chromaticity coordinates for the red-green and yellow-blue axes, respectively. Not surprisingly, measures with mode L* were able to differentiate skin color categorized as light, intermediate or dark. Surprisingly, however, noninvasive measures with mode a* distinguished between healthy subjects and those with a photodermatosis, namely either polymorphous light eruption or benign summer light eruption. While the minimal erythema doses appeared normal clinically, they were abnormal compared to those of nonphotosensitive volunteers on chromometric measurements. These observations may indeed reflect interesting pathophysiological mechanisms.

It is important to remember that so far the authors have not uncovered a way to distinguish between a photosensitive and nonphotosensitive individual based on a single erythema reading. To me, these results merely say that if one looks at a group of photosensitive patients, their values in the a* mode tend to differ from those of nonsensitive individuals. They have yet to determine the threshold value for disease. Would the authors care to venture a number value which tends to discriminate between sensitive and nonsensitive individuals? Furthermore, it remains to be seen if these results can be duplicated in other series and whether chromometric differences in actinic erythema will be uncovered in patients suffering from additional types of photosensitivity.

Finally, the fact that there was no difference between polymorphous and benign summer light eruptions in these measurements would seem to lend further support to the belief that the latter is merely a minor clinical variant of the former. I wondered how the authors felt about this interpretation.

Reference 1 Garigue J, Marguery MC, Malmay MF, El Sayed F, Bazex J: Measurement of actinic erythema in healthy subjects and in subjects with polymorphous light eruption using a tristimulus colorimeter. *Dermatology* 1995; 190: 31-34.

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