

Sweet's Syndrome: A Short Review

Thanish Ahamed S¹, Karthikeyan Murthykumar², Saravana Kumar, Sampath Kumar³

First year BDS¹, CRRP², Assistant professor³

Department of Anatomy,

Saveetha Dental College and Hospitals.

Abstract:

Sweet's syndrome or acute febrile neutrophilic dermatosis is a rare disease characterized by painful violaceous erythematous skin lesions, fever and neutrophilic leukocytosis and dense dermal neutrophilic inflammatory infiltrate. It shows excellent response to corticosteroids. This review article briefly explains about Sweet's syndrome and its management.

Keywords: Epidemiology, neutrophils, sweet syndrome

INTRODUCTION:

Sweet's syndrome (SS) is an acute febrile neutrophilic dermatosis first described by Robert Douglas Sweet in 1964(1). Sweet reported cases of eight women treated at Plymouth General Hospital from 1949 to 1964, thus describing the four cardinal signs of the disease: fever, leukocytosis with polymorphonuclear predominance, painful elevated plaques on face, neck and extremities, and histologically, dense dermal infiltration with mature neutrophils. Later, Whittle et al. and Crow et al. were the first to use the term "Sweet's syndrome" as titles of their articles (2).

CLASSIFICATION:

Depending on its association with other diseases, SS can be divided into the following (3,4)

1. Classic or idiopathic
2. Malignancy associated
3. Drug induced.

Idiopathic :

Classically it is the most frequent subset of SS and it represents up to 70% of the cases in old series(5). It predominates in women, especially in patients aged under 45 years. Most recently, this subset has become less prevalent possibly due to better study of the patients (6).

Drug-induced:

More than 25 drugs have been related to the flare of S, but the most frequently implicated is the granulocyte-colony stimulating factor. Other drugs that are commonly associated with development of SS are trimethoprim-sulphamethoxazole, oral contraceptive pills, retinoids, minocycline, hydralazine, carbamazepine, bortezomib, and imatinib. As it is usual in other skin eruptions induced drugs, the disease fades with the withdrawal of the drug and flares up if its re-administrated.

Paraneoplastic SS:

Up to 20% of SS are paraneoplastic (7). The majority of paraneoplastic SS are associated with hematological malignancies, especially with acute myelogenous leukemia and myelodysplastic syndromes (8). About 15% of paraneoplastic SS are related with solid cancers, predominating breast, gastrointestinal, and genitourinary origin.

HISTOLOGY:

Histologically, the disease is characterized by the presence of a dense dermal predominantly neutrophilic inflammatory infiltrate associated with subepidermal edema of variable intensity and nuclear dust. Although criteria for defining the disease include absence of leukocytoclastic vasculitis, there are reports of SS associated with vasculitis (9).

SWEET SYNDROME IN INFANCY:

About 16 % of SS appears in children (10). Pediatric SS is similar to that in adult population, with only three differences: a) it is associated with immunodeficiency, b) it is less associated with malignancy and c) it is probably susceptible to recurrence (11).

TREATMENT :

The first line therapies for SS are systemic corticosteroids, potassium iodide, and colchicines (12). Systemic corticosteroids are the most widely used: the clinical response is fast and brilliant that it is considered a diagnostic criterion (13). Brilliant response to prednisone is darkened by the frequent recurrence: 20-30 % of the patients will suffer recurrences after treatment withdrawal and up to 10% of the cases will have a chronic and recurrent evolution for more than 1 year (14,15,2,16).

Potassium iodide is a therapy as fast and effective as systemic corticosteroids. In fact, the response to this agent was included in the diagnostic criteria by von den Driesch (2). Systemic symptoms disappear within 24 to 48 hours and cutaneous plaques in as much as 1 week. The dosage of potassium iodide is 300mg administered orally three times daily. The main adverse effects are gastrointestinal intolerance, hypothyroidism, and vasculitis (17).

The other first line therapy for SS is colchicines. This drug is administered at a dosage of 0.5mg, two or three times per day. It can be maintained from 2 to 4 weeks. About 90 % of the patients respond favorably within a few days and its main limitation are the gastrointestinal side effects (18).

CONCLUSION:

It is important that physicians in general and especially dermatologists know how to diagnose SS. Due to high association of the syndrome with malignant diseases, it is essential that malignancies be discarded and the patient follow up be maintained.

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