

Sweet Syndrome in childhood*

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Abstract: Sweet syndrome or acute febrile neutrophilic dermatosis is a recurrent and rare skin disease caused by the release of cytokines, with diverse possible etiologic causes. It presents clinically with polymorphic skin lesions, fever, arthralgia, and peripheral leukocytosis. In general, it is associated with infections, malignancy and drugs. It usually regresses spontaneously and treatment is primarily to control the basic disease. The authors report the case of a child of 1 year and 11 months who developed Sweet syndrome.

Keywords: Case reports; Child; Sweet Syndrome

INTRODUCTION

Sweet Syndrome is a febrile neutrophilic dermatosis, characterized by the onset of painful erythematous-violaceous lesions that may be located mainly on limbs, trunk, face and neck.^{1,2}

The disease is universally distributed among the races. In adults, women are more commonly affected, mainly with the idiopathic and drug-induced forms. The first episode generally occurs between the ages of 30 and 50 years.^{1,3} The cases associated with neoplasms, on the other hand, are equally distributed between the genders.³ The cases described in children are rare (8% of the total), equally distributed between the genders and are preceded by three to one week by infectious diseases of the upper respiratory tract.^{1,3}

Remission of cutaneous symptoms is variable; without treatment the lesions may persist for several months or even experience spontaneous remission. In case of relapse, there is strong association with malignant diseases such as neoplasm recurrence.^{1,3}

CASE REPORT

Patient, female, one year and eleven months old, with onset of erythematous-papular lesions on the forehead for around 3 weeks that progressed to erythematous-violaceous lesions in plaques involving all the face (Figures 1 and 2). There was also onset of lesions on the lower limbs bilaterally, mainly on the posterior surface of limbs, characterized by erythematous plaques

with raised borders and central necrosis (Figure 3). There was no itching and discreet pain in lower limb lesions. She did not present lesions in the oral cavity, trunk, palms and soles of feet. She had been using ferrous sulfate and montelukast sodium for 3 months.

During the last week the patient had fever varying between 37.5°C and 38.5°C. She was eating well, without vomit and/or diarrhea. The following tests were requested:

- Hemogram: red blood cells 4,000,000; hemoglobin 9.91; hematocrit 31; platelets 442,000
- Leukogram: 12,300 leukocytes with 51% segmented, 2% band neutrophils, 34% lymphocytes and 8% eosinophils
- EAS: volume 20ml, density 1010, slightly cloudy, citrine in color, 20-30 pyocytes, 5-8 red blood cells, ketone bodies (1+).
- Biochemical tests, abdomen ultrasound and thorax X-ray without alterations.

A cutaneous incisional biopsy was done on a lesion on a left lower limb. Treatment for urinary infection was started with cephalexin (100mg/kg/day) and prednisolone (1mg/kg/day) for one week. The histopathologic result was febrile neutrophilic dermatosis (Figures 4 and 5). After 5 weeks of treatment with oral corticotherapy there was clear improvement of lesions, mainly edema regression, infiltration and erythema, in addition to absence of febrile symptoms.

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FIGURE 1:
Erythematous-violaceous plaques on right hemiface



FIGURE 2:
Erythematous-violaceous plaques on left hemiface

DISCUSSION

The Sweet Syndrome (SS) is an acute, febrile neutrophilic dermatosis, first described by Robert Douglas Sweet, in 1964¹ It is characterized by the acute onset of fever, erythematous-violaceous cutaneous lesions, usually painful, located mainly on limbs, trunk, face or neck.² The inflammatory edema may impart the aspect of lesions with vesiculations, but palpation shows that actually the lesions are solid, somewhat softened; the name given to this is pseudovesiculation. As the lesion regresses, there may be a central lightening of the lesion color, giving it a



FIGURE 3:
Erythematous plaques on posterior surface of lower limbs with elevated borders and central ulceration (target aspect)

target aspect similar to erythema multiforme.^{1,2}

Women are more frequently affected and seem to be particularly involved by the idiopathic or drug-induced forms.^{1,4} The onset is habitually described as occurring between the ages of 30 and 60 years, being rare in childhood.^{2,4}

It is frequently related to infectious diseases, inflammatory diseases, drugs and neoplasms.² Neoplasms are associated with malignant disease at a rate of 21%, usually as hematologic disorder.^{1,2} The neoplasm more commonly linked to SS is Acute Myeloid Leukemia (AML).^{1,2,4}

In pediatric patients, the dermatosis is usually preceded by signs and symptoms of infection in the upper respiratory tract, from one to three weeks before the onset of cutaneous lesions.^{4,5} Vacaro et al⁴ suggested that infectious agents might be triggers of reactive skin diseases, either through direct damage or molecular mimicry.⁶

The pathogenesis of the disease suggests vasculitis by altered immune complex and neutrophil function. The authors presume that the child developed the classical or idiopathic form of the disease. Montelukast sodium acts as inhibitor of leukotrienes.⁷ Leukotrienes stimulate leukocyte function, including lysosomal enzyme release, polymorphonuclear leukocyte adhesion and aggregation. Therefore, their inhibition would go against the SS theory. As the child presented the reaction for the first time and all tests were within normal standards, the authors also believed, at first, that a relationship with neoplasms was not very likely.

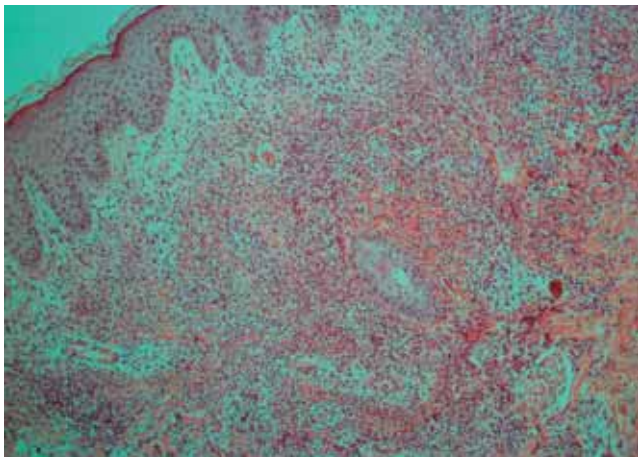


FIGURE 4: 100x - Perivascular neutrophilic nodular infiltrates. Important edema of the papillary dermis

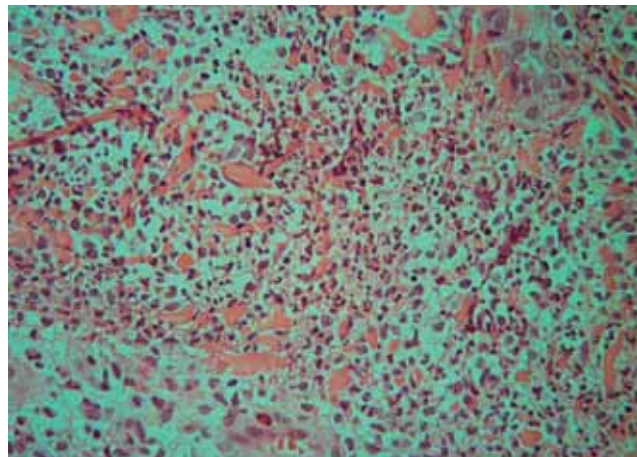


FIGURE 5: 400x- Perivascular neutrophilic nodular infiltrates in middle and upper dermis

The diagnostic criteria for SS were proposed by Su and Liu in 1986 and modified by Von den Driesch in 1994.⁶ In order to receive this diagnosis the patient must have 2 major criteria and 2 minor criteria. We can cite as major criteria: 1) the sudden onset of painful erythematous or violaceous plaques or nodules; 2) a predominantly neutrophilic dermal infiltrate, without leukocytoclastic vasculitis. The minor ones would be: 1) fever or prior infection; 2) arthralgia, conjunctivitis or malignancy; 3) leukocytosis; 4) favorable response to use of corticosteroid therapy.

Histopathology shows visible perivascular neutrophilic nodular infiltrates, with necrophiliac cariorrexis. Even though there is no primary vasculitis, blood vessels may be secondarily involved in immunologic response, an infrequent finding.^{6,8}

Laboratory alterations include peripheral leukocytosis with neutrophilia and elevated speed of erythrocyte sedimentation or C-reactive

protein, particularly in cases where SS associated with malignancy, leukopenia, anemia and thrombocytopenia have been reported.^{3,9,10}

We may cite as differential diagnoses erythema multiforme, erythema nodosum, erythema elevatum diutinum, Hansen's disease, pyoderma gangrenosum, bromoderma, leukocytoclastic vasculitis and facial granuloma with eosinophilia.⁸

The treatment is based on corticosteroid therapy, either oral (30 to 60mg/day for 4 to 6 weeks) or intralesional¹⁻⁶. In the studied case oral corticosteroid therapy was chosen due to dissemination of lesions and because that was the recommended way for the patient's age group.

The lesions usually leave no scars and even without treatment tend to achieve spontaneous cure in 2 to 4 weeks. There frequently are recurrences in between 30 and 50% of cases⁶⁻¹⁰, depending on the etiology. □

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