ORIGINAL RESEARCH PAPER

Volume-7 | Issue-7 | July-2018 | PRINT ISSN No 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

SWEET'S SYNDROME IN CHILD : A CASE REPORT

Pediatrics

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ABSTRACT

Sweet's syndrome is characterized by a group of clinical features and physical findings that comprise of pyrexia, hematologic and tissue neutrophilia associated with painful reddish plaques, histopathologically characterized by the presence of abundant mature polymorphs. Here we present a case of 6 years old girl who presented with fever, corhyza, pain abdomen and numerous dermatologic lesions of 2 weeks duration. On examination , she had plaques and papules with elevated margins all over body. Labs revealed neutrophilia with raised CRP. Skin biopsy revealed dense neutrophilic infiltration of dermis. This case is being reported because this disease should be considered as differential in children with pyrexia of unknown origin with tender plaques and hematologic and histopathologic neutrophilic leukocytosis.

KEYWORDS

Introduction:

Sweet's syndrome SS ,also known as acute febrile neutrophilic dermatosis, first described by Sir Robert Douglas Sweet in 1964 (1). It is clinically characterized by fever, blood and tissue neutrophilia, leading to development of tender, erythematous inflammatory skin lesions (papules, nodules, plaques) and histopathologically by the presence of abundant mature neutrophils (2). It presents in three clinical settings : classical (para infectious) SS, depicting a hypersensitivity reaction preceding infection; malignancy associated (para neoplastic) SS (in children usually associated with acute myeloid leukemia); and as an adverse drug reaction sometimes in connection with certain underlying diseases (drug induced SS) (3).

Diagnoostic criteria for SS were proposed by Su and Liu in 1986 and modified in 1994 by von den Driesch (4). (table 1).

Table 1: Diagnostic criteria for sweet syndrome[4]
Diagnostic criteria : Both major criteria and 2 of the 4 minor criteria

Major:

1. Abrupt onset of nodules or painful erythematous plaques
2. Histopathologic evidence of a dense neutrophilic infiltrates in the dermis without leukocytoclastic vasculitis

Minor:

1. Fever above 38°C Infection of the respiratory tract or gastrointestinal tract preceding the medical condition or association with vaccination, inflammatory disease, neoplasia, or pregnancy
2. Presence of the following laboratory findings (3 of 4): ESR above 20 mm; positive C-reactive protein; leukocyte count above 8000, neutrophil blood count over 70% Excellent response to treatment with corticosteroids or potassium iodide.

Histologically , the disease is characterized by the presence of a dense dermal neutrophilic inflammatory infiltrate associated with subepidermal edema of variable intensity and nuclear dust which was seen in histopathologic examination too.

Apart from dermatologic involvement, various other organ systems are involved too. Mucosal involvement is most common causing airway obstruction due to edematous and aphthous ulcerations of aerodigestive tract. Sometimes headache, myalgias and arthralgias may accompany. The disease usually resolves by itself. Recurrence is considered as the most common complication.

Treatment usually includes corticosteroids both systemic, topical or intralesional. Pediatric population is usually resistant to steroids. Oral application of potassium iodide results in rapid resolution of symptoms. In patients who are either resistant to steroids or in whom steroids are contraindicated, drugs like clofazimine, cyclosporine, dapsone and indomethacin can be tried. (5,6)

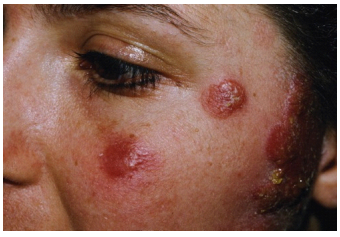
Case report:

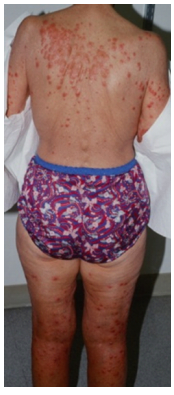
A 6 year old girl presented in outdoor department with history of sudden onset of skin lesions of 12 days duration. The lesions initially appeared over face and later involved whole body. There was history of upper respiratory tract infection 1 week prior to onset of lesions. There was no history of any drug intake neither there was any systemic involvement.

On examination, she was sick with fever of 103 degree farhenheit with bilateral conjunctival suffusion with upper cervical lymphadenopathy. There were multiple tender papules and plaques with central hyperpigmentation and elevated borders. Some lesions were resolving. The lesions were typically “liquid to look, solid to touch”. All systems were grossly normal.

Lab investigations revealed hemoglobin of 10.4 gm/dl, microcytic hypochromic type of RBCs, TLC of 32,000 per cu mm with 84% polymorphs. No immature cells or band forms were present. ESR was 54 mm in first hour and CRP was positive (84 mg/ L). The biopsy of skin lesions showed a dense polymorphic infiltrate in upper dermis with vascular sparing.

The girl was managed with oral prednisolone (1 mg/kg/day), topical potassium iodide over skin lesions and oral clarithromycin. Patient improved with above mentioned treatment and there was clearance of skin lesions. Patient was discharged in stable condition after 1 week.





Discussion:

Sweet's syndrome is a rare disease in pediatric age group and accounts for 8 percent of cases with no sexual predominance.

There are 2 forms of the disease : idiopathic and malignancy asscociated. There are previous case reports in pediatric population associated with hematologic disorders. One fifth of cases are associated with hematoproliferative or solid tumors. Relapses are usually associated with malignancy . secondary causes include infections, inflammatory bowel diseases, rheumatologic disorders, pregnancy, primary immunodeficiency and drug or vaccine induced conditions.(7)

In present case the patient had conjunctival involvement with history of upper respiratory infection. The diagnosis is based on clinical and histopathologic findings consistent with literature. There was significant rapid response to steroids and potassium iodide.

Hence sweet syndrome should be kept in mind when patient has a history of high grade fever, skin lesions and elevated inflammatory markers. In pediatric age group, if there is recurrence, malignancy should be ruled out.

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