CASE REPORT

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Pediatric Sweet Syndrome: A rare Cause of Fever of Unknown Origin

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ABSTRACT

Sweet syndrome or acute febrile neutrophilic dermatosis, is characterized by fever, elevated neutrophil count, painful erythematous cutaneous lesions that have an infiltrate of mature neutrophils typically located in the upper dermis. In general, it is associated with infections, malignancy and drugs. Sweet syndrome is rarely seen in pediatric patients. In this report, we present a child who was diagnosed with sweet syndrome because of a rare disease in children with unknown etiology of fever.

Key Word: Fever, Children, Sweet syndrome

Introduction

Fever of unknown origin is defined as week long fever which can not be diagnosed inspite of careful physical inspection, detailed history and laboratory investigations generally include abdominal tomography. In pediatric patients, fever of unknown origin may develop due to various infectious and non-infectious causes. The underlying cause of fever of unknown origin is generally investigated in 5 categories as infectious diseases, malignant conditions, collagen vascular diseases, miscellaneous diseases and undiagnosed etiology (1). Sweet syndrome also known as acute febrile neutrophilic dermatosis is defined by Robert Douglas Sweet in 1964 (2). The revised diagnostic criteria, having sudden onset painful erythematous plaque or nodules, rarely vesicle, pustule or bullous lesions on the skin and in histopathological evaluation not having leukocytoclastic vasculitis are accepted as major criteria. Minor criteria are preceeding infection/vaccination with associated diseases (inflammatory, malignant) or pregnancy, fever, and general malaise, elevated erythrocyte sedimentation rate (ESR), c reactive protein (CRP), segmented neutrophils and bands >70 %, leukocytosis > 8000/µl, and response to steroids. For diagnosis, two major and at least one minor criteria are required[3].
We presented a child who was diagnosed as sweet syndrome with fever and rash.

Case Report

We report an eighteen-month male patient with a normal prenatal history and having fever and maculopapular rash starting at two months. The patient was investigated for fever and rash, his skin biopsy was carried out but any diagnosis could not be reached and he was taken into follow-up in which his fever and rash continued. He was admitted in another hospital with the diagnosis of lower respiratory tract infection and was transferred to our hospital with the diagnosis of fever of unknown origin. His body temperature was 38.5 ℃ and the other vital symptoms were stable. Physical examination revealed painful erythematous or violaceous papules and plaques on her entire body and the patient seemed restless. In the physical examination, approximately 5 cm liver midclavicular was palpable.
There were no symptoms and signs related to the patient's eyes. White blood cell (WBC) was measured 11600/mm3, 54% of it being (leukocyte with polymorphic nuclei) PMNL. CRP was 69mg/lt and erythrocyte sedimentation rate (ESR) was 47mm/h. Hepatosplenomegaly was discovered in abdominal ultrasonography. Chest radiography, bone marrow examination and immunoglobulin levels and lymphocyte sub-groups were normal. Histopathological evaluation result of liver biopsy was reported normal. In the skin biopsy, superficial and deep perivascular, periadnexal, and intestial dermatitis with neutrophile dominant infiltration was reported. (Figure1).

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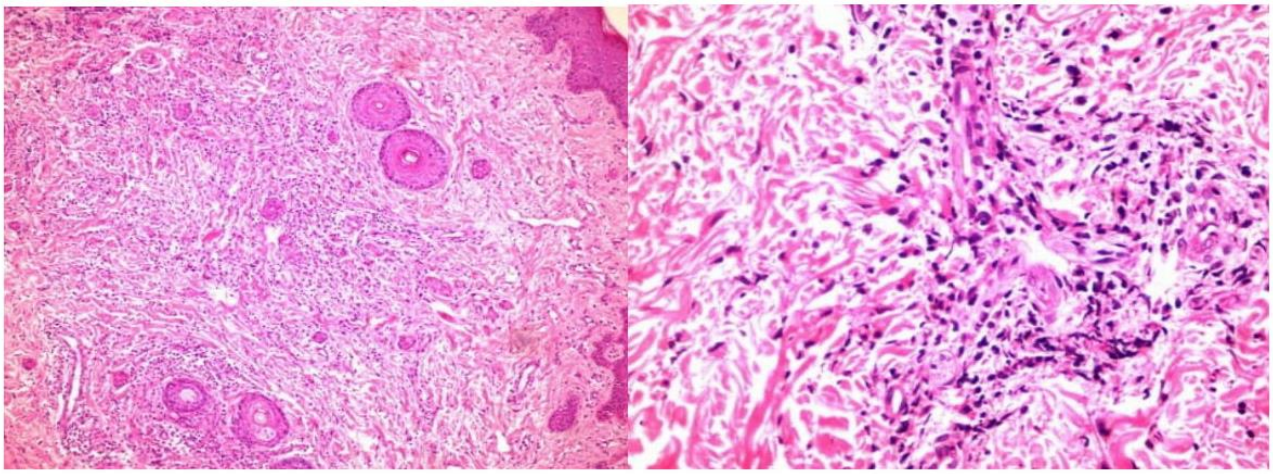

Haemotoxilin-eosin ×40 Haemotoxilin-eosin ×200

Fig. 1. Neutrophile dominant infiltration in dermis and subcutis

In our patient, 2 major and 3 minor criteria were present according to the diagnostic criteria of sweet syndrome[3].With the diagnosis of Sweet syndrome, in the first three days 30mg/kg/day intravenous methylprednisolone was given and after 3 days, treatment was progressed with 2mg/kg/day oral prednisolone. After the second day of steroid treatment, the patient did not have fever. In the first twenty-four hours, his rash decreased and color fading was observed. Activeness of the patient increased in the first week of follow-up and his painful facial expression got better. At the next follow-up, his rash significantly regressed and acute phase reactants returned to normal levels. Size of the liver regressed from 5 cm to 2 cm. Dose of the steroid was planned to be cut off incrementally; however, when the dose was 0.1 mg/kg/day, the rash and fever started again. Thus, steroid treatment was continued, 1 mg colchicine was applied.

Discussion

Infection and several other diseases can be categorized in the etiology of fever of unknown origin. Although it is rarely seen, sweet syndrome should be considered in the case of children who has fever of unknown origin and rash[1].Sweet syndrome is diagnosed less in children under 5 than in adults. There is a case with signs of sweet syndrome as early as three days (4,5,6). Sweet syndrome has recently been grouped into three: classic, paraneoplastic, and related to drug. Classic sweet syndrome can be defined as idiopathic sweet syndrome which can not be diagnosed and also occurs following upper respiratory tract infection and gastrointestinal infection (7,8). Classic-idiopathic sweet syndrome is frequently seen in children. 42% of the pediatric patients diagnosed with sweet syndrome is in the group of classic-idiopathic. Cases with parainflamatory (33%) and paraneoplastic (25%) sweet syndrome come respectively. Cases of drug-induced sweet syndrome is reported relatively less (6,9). Our patient's rash and fever increased from time to time but they never came to an end. Erythrocyte sedimentation rate and CRP value were high level. The number of white blood cells was 15000 /mm^3 and PMNL domination was in existence. Patient's skin biopsy was compatible with notrophilic dermatosis. With these symptoms, the patient with normal values for the diseases related to sweet syndrome and also had no history of drug usage was diagnosed with classic-idiopathic sweet syndrome.

For the treatment of sweet syndrome, systemic steroid which equals to 1 mg/kg/day prednisone is generally used. With the corticosteroid treatment, symptoms get better quickly, but symptoms re-occurred in 20-30% of the cases (3-8). Intravenous pulse methylprednisolone was used as topical or into the lesion and steroid treatment was applied as single or combined. Potassium iodide and colchicine are first generation drugs used in the treatment for sweet syndrome (7,8). In order to provide a concrete solution and prevent recurrence, we applied pulse-methylprednisolone and oral prednisolone treatment; however, when the steroid was cut off, the complaints started again. Instead of long-term steroid treatment, colchicine treatment with the dose of 0.5 mg was applied.

Idiopathic sweet syndrome may be the first symptom of systemic inflammatory diseases and malignancy; therefore, clinical follow-up is vital.

During the active period of this disease, weekly follow-ups are recommended and in these follow-ups, along with ESR, complete blood count should be monitored (9).

Differential diagnosis should be made with other neutrophilic dermatosis diseases such as aseptic pustules, Neutrophilic urticarial dermatoses, Pyoderma gangrenosum and Marshall's syndrome (10).

Consequently, even if sweet syndrome is defined as a concomitant hypersensitivity reaction to infections, immunodeficiencies, drug usage, malignancy and systemic inflammatory diseases, causes of this syndrome are not known completely. Sweet syndrome should be considered if there is rash in the children with diagnosis of fever of unknown origin.

References

1. Chusid MJ. Fever of unknown origin in Childhood. Pediatr Clin N Am 2017; 64: 205-230.
2. Sweet RD. An acute neutrophilic dermatosis. Br JDermatol 1964; 76: 349-356.
3. Von den Driesch P. Sweet's syndrome (acute febrile neutrophilic dermatosis).J Am Acad Dermatol 1994; 31: 535-560.
4. Garcıa-Romero MT, Ho N. Pediatric sweet syndrome. A retrospective study. International Journal of Dermatology 2015; 54: 518-522.
5. Halpern J, Salim A. Pediatric sweet syndrome: case reportand literatüre review. Pediatr Dermatol 2009; 26: 452-457.
6. Uihlein LC, Brandling-Bennet HA, Lio PA, Liang MG. Sweet syndrome in children. Pediatr Dermatol 2012; 29: 38-44.
7. Anzalone CL, Cohen PR. Acute febrile neutrophilic dermatosis (Sweets syndrome). CurrOpinHematol 2013; 20: 26-35.
8. Cohen PR. Neutrophilic dermatoses: a review of current treatment options. Am J Clin Dermatol 2009; 10: 301-308.
9. Hospach T, von den Driesch P, Dannecker GE. Acute febrile neutrophilic dermatosis (Sweets syndrome) in childhood and adolescence: two new patients and review of the literature on associated diseases. Eur J Pediatr 2009; 168: 1-9.
10. Webb K, Hlela C, Jordaan HF, at al. A Review and Proposed Approach to the Neutrophilic Dermatoses of Childhood. Pediatr Dermatol 2015; 32: 437-446.