

Sweet's Syndrome in Childhood: A Case Treated with High Dose Methyl Prednisolon

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Purpose: Sweet's syndrome, which is also defined as acute febrile neutrophilic dermatosis, is a rare disease characterized by recurrent painful erythematous plaques, fever, peripheral leukocytosis and dense dermal neutrophilic infiltration. A case with Sweet's syndrome was presented in this paper because the disease is very rarely described in childhood.

Patient and Method: The case was admitted with a five-day history of fever and painful skin eruption on the both of the legs. Painful, sharply demarcated, erythematous plaques were found on both tibias. In laboratory investigation; leukocytosis, elevated ESR and CRP were obtained.

Result: The cutaneous biopsy revealed band-like infiltration of dermis with neutrophils. Prednisolone at dose of 0.5-2mg/kg/day is generally used for the treatment of Sweet's syndrome.

Conclusion: Our case was interesting also because he was successfully treated with high dose methyl prednisolone. We showed that high dose-methyl prednisolone was highly effective. Ann Med Sci 2002;11:55-58

Key words: Sweet's syndrome, dermatosis, childhood, treatment

Sweet's syndrome, which is also defined as acute febrile neutrophilic dermatosis, is a rare disease characterized by recurrent painful erythematous plaques, fever, peripheral leukocytosis and dense dermal neutrophilic infiltration^{1,2}. This entity was first described by Robert Sweet in 1964³. Classically Sweet's syndrome occurs in middle age woman. The disease rarely occurs in childhood^{1,2,3}.

It is thought that the cause of the disease is an either hypersensitive reaction to a bacterial or viral infection or an immunologic response to inflammatory and neoplastic disease. Massive peripheral and tissue leukocytosis suggests a possible pathogenic role of leukotactic mechanism^{1,2,4}. It was suggested that interferon γ , interleukin-3,6,8 and G-CSF have been involved in the pathogenesis of the disease and activation of cytokine cascade might be responsible for the local and systemic activation of neutrophils, and

clinical and pathologic findings^{5,6}. Development of the Sweet's syndrome during the G-CSF therapy supported the possible role of the hematopoietic mediators in pathogenesis⁷.

A case with Sweet syndrome that was treated with high dose methyl prednisolone has been presented in this paper because of the rarity of disease in childhood.

Case Report

A four year old boy was admitted with a five-day history of fever, pain and swelling of the ankles and painful skin eruption on the both of legs. He had been admitted to the hospital with complaints of fever and arthralgia six months ago. He had been diagnosed as having septic arthritis and received antibiotics, but no microorganism had been able to be isolated from the puncture of the joint. Erythematous plaques had been obtained on the upper extremities during the treatment and considered as drug eruption.

His body temperature was 39 °C on admission. In the physical examination, previous lesion on the arms caused mild atrophy and had a wrin-

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kled of the skin. His ankles were swollen and painful with movement. Painful, sharply demarcated, erythematous plaques with pseudovesicular and pustular appearance were found on both tibias. The sizes of the largest lesion were 4x6cm in diameter on the left, 8x6 cm in diameter on the right. In laboratory investigation; haemoglobin level was 10.4 g/dl, white blood cell count was 31.800/mm³ with 85% polymorphonuclear leukocytes, erythrocyte sedimentation rate (ESR) was 65 mm/hour, C-reactive protein (CRP) was 6.9 mg/dl (normal<0.5 mg/dl), ANA and ADNA was negative.

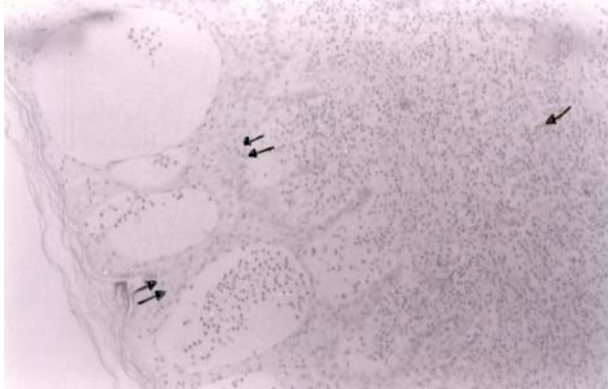


Figure 1. Band like neutrophilic infiltration (↑) and exocytosis of cells through epidermis (↑↑) (HEx20)



Figure 2. Healing lesion with erythematous plaque on leg after steroid therapy, healed lesion with mild hyperpigmentation and local skin atrophy on the arm.

Antibiotic treatment (Teikoplanin) was initiated for suspected erysipelas, but no response to the antibiotic therapy was obtained within the first five days. Microbiological examination of punctuated pustules and blood remained sterile. During this period the cutaneous lesions shows arcuate annular patterns with a faint central white-discoloration. His clinical, dermatological features and previous history suggested Sweet's syndrome and cutaneous biopsy was performed. Band-like infiltration of dermis with neutrophils and exocytosis of cells through epidermis were the main characteristics of the biopsy (Figure 1).

Clinical and laboratory features of patient did not confirm the association of any malignant or connective tissue disease. Following a skin biopsy, antibiotic was stopped and methyl prednisolone therapy was started with 30 mg/kg/day, tapered to 20 mg/kg/day at day three, to 10 mg/kg/day at day six and 5 mg/kg/day at day ten. Fever and leucocytosis resolved within two days and skin lesion started healing within a few days (Figure 2). Then prednisolone at a dose of 2 mg/kg/day continued for one month. No relapse was observed in the 24-month follow-up period.

Discussion

Sweet's syndrome is a rare inflammatory cutaneous disease that is most commonly seen in adults¹. It may be difficult to recognize when it is presented in childhood. Approximately 30 children with Sweet's syndrome have been reported in English literature⁸⁻³³. The youngest child was seven weeks old at diagnosis²⁵. Although the disease is mostly seen in women in adulthood, there is no sexual predominance in childhood. A clinical hallmark of the syndrome is the occurrence of multiple painful, raised erythematous plaque on the skin. Lesions are frequently seen on the face in adults^{1,2}. In our case, lesions were located on both legs. Generally cutaneous plaques heals without any scar. In our patients previous lesions had been healed with atrophic scar. Secondary cutis laxa has been described within the areas of the involvement in some children^{12,15,16,30}. Although mucosal involvement may be present in Sweet's syndrome we did not obtain it in our patient. A high fever up to 40 °C has been described in 85-90% of patients. Arthralgia and arthritis occur in 25-50% of patients^{1,2}. Besides arthritis, sterile osteomyelitis has been observed in pediatric cases^{17,19,23}. Findings of arthritis had been noted in the previous history and on admission. Although lung, kidney, liver, eye involvement has been reported, there was no evidence of those in our patients^{1,2}. Leucocytosis with 80% to 90% percent polymorphonuclear leucocyte, elevated ESR and CRP has been described in most patients^{8,30}. Autoantibodies to neutrophil cytoplasm antigen (ANCA) could be found in some patient³⁴.

Sweet's syndrome is classified as neutrophilic dermatosis. The main histologic characteristics of the disease are intense dermal infiltrates consisting of neutrophils with little evidence of primary vasculitis, prominent edema of dermal papilla^{1,2,3}. Histopathologic features of the skin biopsy of our patient were in agreement those described in literatures.

Sweet's syndrome is accepted as idiopathic and cause of disease remains unclear. Although

it is not an infectious disease most of the patients have a history of respiratory or gastrointestinal system infections preceding the signs of the disease by one to three weeks^{1,2}. Sweet's syndrome has been associated with specific systemic disorders including Behçet's disease, inflammatory bowel disease, connective tissue diseases, systemic infectious disease and hematopoietic malignancy^{20,21,31,32,35-39}. Although there was no underlying disease, the preceding upper respiratory system infection was present in the history.

Two major and four minor criteria for diagnosis have been proposed. Typical cutaneous lesion and histopathologic findings are accepted as major criteria. Association with inflammatory or neoplastic disease, fever >38 °C, abnormal laboratory response (leucocytosis, elevated ESR, positive CRP reaction) and an excellent response to treatment with systemic corticosteroids are accepted as minor criteria. The presence of both major and two minor criteria is required for diagnosis^{1,2}. Our patient had all major and major-ity of minor criteria.

The standard therapy for Sweet's syndrome is glucocorticoids^{1,2}. Other agents include potassium iodide⁴⁰, colchicine⁴¹, doxycycline⁴² have also been shown to induce regression of Sweet's syndrome. Different pharmaceutical formulations of glucocorticoids used for treatment of the disease have been reported in literature. Excellent results with prednisolone therapy at dose of 0.5-2mg/kg/day have been reported^{1,2,8,24,29,31}. Topical and intra-lesional glucocorticoids were also used for localized lesion^{39,43}. We could not find any case that has been administered high dose methyl prednisolone except one case treated successfully with pulse methylprednisolone and chlorambucil⁴⁴. We showed that high dose methyl prednisolone was highly effective. Short-term relapses after the end of steroid therapy were reported in 25% of patients. Our patient treated with high dose methyl-prednisolone has been relaps-free for 24 months.

In summary, Sweet's syndrome is a distinct clinical entity that is most commonly seen in adults. It may be difficult to recognize it when it is present in a child, thus the patient could be diagnosed as infection or connective tissue disorder. and treated wrongly when a child with a complaint of fever and painful erythematous plaque on skin is unresponsive to the antibiotics the diagnosis of Sweet's syndrome must be kept in mind. High dose methyl prednisolone may be an alternative for the treatment of these patients.

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