TWO CASES WITH SWEET'S SYNDROME

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SUMMARY : *Acute febrile neutrophilic dermatosis (Sweet's syndrome) is a rare disorder. Here we present two cases with Sweet's syndrome successfully treated with corticosteroids.*

Key Words: *Sweet's Syndrome, Acute Febrile Neutrophilic Dermatosis, Treatment.*

INTRODUCTION

Acute febrile neutrophilic dermatosis was first described in 1964 by Sweet (1). The disorder was initially characterized by an acute eruption of painful erythematous plaques and nodules associated with fever, leukocytosis and dense papillary dermal neutrophilic infiltrates on biopsy specimen.

Etiological factors such as malignities, infections, genetic predisposition and inflammatory bowel diseases can be found in only 50% of the patients. In this paper we present two cases of Sweet's syndrome.

CASE REPORTS

CASE I: A 55-years-old female was admitted to our clinic with fever, malaise and an abrupt onset of pinkish violaceous papules. These papules were located on the arms, legs and neck, then became vesicular, infiltrated and tender (Fig. 1). Pathergy test, which was performed to exclude Behçet's disease, was negative. She had nephrectomy because of hypernephroma 3 years before and had an upper respiratory tract infection two weeks prior to the consultation. A fever up to 38.5°C was detected. On laboratory investigations, hemoglobin level was 10.6 g/dl, white blood cell count was 9900/mm^3 and platelet count was 235000/mm^3. Erythrocyte

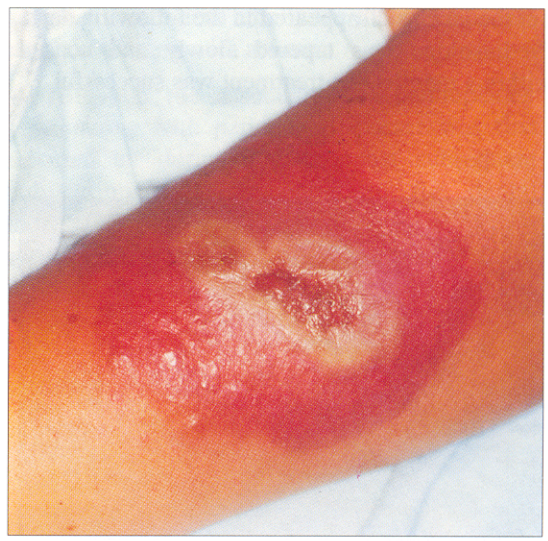

Fig. 1: Sweet's syndrome lesions located on the forearm of the first patient.

sedimentation rate was 31mm/hour. C- Reactive Protein (CRP) was positive. The biopsy specimen showed perivascular infiltrate of inflammatory cells, composed mainly of neutrophils arranged as a band in the dermis. There was no vascular damage.

Topical corticosteroids twice daily improved the lesions and no systemic therapy was needed.

CASE II: A 43-years-old female was admitted to our clinic with fever, malaise and erythematous tender lesions on the neck and legs of one week duration. These lesions started as small pinkish papules and vesicules (Fig. 2). She had had a similar attack 10 years ago and an upper respiratory tract infection one month ago. When a 38.5°C fever was detected, white blood cell count was 12600/mm^3 and platelet count was 182000/mm^3. Erythrocyte sedimentation rate was 12 mm/hour. CRP was positive. The biopsy specimen taken from a single lesion showed band like infiltration of neutrophils in the dermis without any evidence of vasculitis.

40 mg/day of oral prednisolone was commenced as treatment. Initially an exacerbation was observed in the first few days but the lesions disappeared in the following days. Prednisolone was tapered slowly and ceased within a month. The treatment was successful.

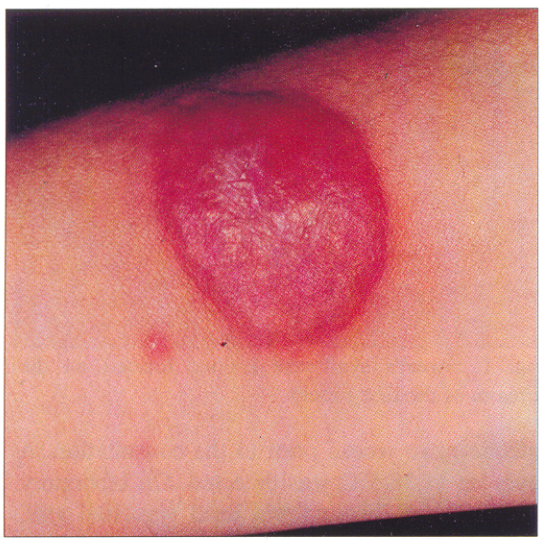

Fig. 2: Clinical presentation of the second patient.

DISCUSSION

Sweet's syndrome is characterized by a rapid onset and fast growing, tender, erythematous, 2-10 cm diameter plaques. The lesions tend to be on the face, neck and extremities. These tender plaques may have a vesicular or pustular appearance peripherally. 86% of patients have a preceding upper respiratory infection. Sweet syndrome can be infectious or reactive. Diagnostic criteria and several accompanying diseases with Sweet's Syndrome can be seen in Table I and Table II. In both of our patients, upper respiratory tract infection was evident.

Fever is almost always present. Usually there is neutrophilic leukocytosis. However in some instances, especially in patients with hematological malignancies such as French, American, British Acute Myeloid Leukemia subtypes 4 and 5 (AML M4 and AML M5) neutrophilic dermatosis can occur during granulocytopenia. In such cases cells in the dermal infiltrate derive from monocytoid AML M4 and AML M5 leukemic cells (2). There is no neutrophilia in 50% of patients with malignancy (3).

Arthralgia or arthritis occurs in 33% to 62% of patients. Myalgia is a common feature. Ocular involvement is reported in 4% to 72% of patients and may manifest as conjunctivitis, episcleritis, subconjunctival bleeding, inflammatory glaucoma, iritis or limbal nodules (4-6). Renal involvement with proteinuria and hematuria can be seen. Pulmonary manifestations of Sweet's syndrome, consisting of bilateral infiltrates, bronchiolitis obliterans- organizing pneumonia (BOOP) and pleural effusions have been described (7).Osteomyelitis and nervous system involvement can also be seen. Pancolitis with diffuse transparietal infiltration with mature neutrophils have been described (8).

Approximately 20% of cases are associated with malignancy. Attacks of Sweet's syndrome may precede the diagnosis of neoplasia by 3 months to 6 years. Patients with associated malignancy are often males with mucosal symptoms, anemia and frequent recurrence of skin symptoms (9, 10).

A typical histopathological finding is a dense perivascular infiltrate composed largely of neutrophils. Some of the neutrophils may show

Table - 1: Diagnostic criterias for Sweet's syndrome.

MAJOR CRITERIA: 1.Abrupt onset of tender erythematous plaques or nodules occasionally with vesicles, pustules or bullae 2. Predominantly neutrophilic infiltration in the dermis without leucocytoclastic vasculitis.
MINOR CRITERIA: 1. Preceded by a nonspecific respiratory or gastrointestinal tract infection or vaccination or associated with: Inflammatory diseases such as chronic autoimmune disorders and infections Hemoproliferative disorders or solid malignant tumors Pregnancy 2. Accompanied by periods of general malaise and fever (>38°C) 3. Laboratory values during onset: ESR>20; C-reactive protein positive; segmented nuclear neutrophils stabs >70% in peripheral blood smear 4. Leukocytosis >8000 (three of the four of these values are necessary) 5. Excellent response to treatment with systemic corticosteroids or potassium iodide
Both major and two minor criteria are needed for diagnosis.

Table - 2: Accompanying diseases with Sweet's syndrome.

1. Streptococcal infections 2. Inflammatory bowel diseases 3. Myelodysplastic syndrome 4. Nonlymphocytic leukemia and other hematological malignancies 5. Solid tumors 6. Pregnancy 7. Vaccines

nuclear fragmentation. There may be some mononuclear cells such as lymphocytes and histiocytes and occasional eosinophils. The inflammatory cells typically assume a band like distribution throughout the papillary dermis. There is usually vasodilatation and swelling of endothelium with moderate erythrocyte extravasation and prominent edema of the upper dermis, which in some instances may result in subepidermal blister formation. Extensive vascular damage is not a feature of Sweet's syndrome (11). However it must be kept in mind that histological features of vasculitis such as erythrocyte extravasation, intramural inflammatory cells and fibrinoid necrosis can be found in lesions of acute febrile neutrophilic dermatosis, and their presence does not rule out this diagnosis (12). Samples taken from older lesions may show prominency of lymphocytes and histiocytes. In fact Delabie et al revealed that the dermal infiltrate in the majority of cases contain numerous histiocytes that mimic neutrophils (13). These mononuclear cells secrete cytokines such interleukin (IL)-1 as IL-1 and IL-8, which are responsible for the clinical manifestations and neutrophilic infiltration (2). IL-1 induce fever and arthritis and in association with IL-8 and Granulocyte-Macrophage Colony Stimulating Factor is responsible for the recruitment, stimulation, and proliferation of neutrophils (14).

In the differential diagnosis erythema nodosum should be considered. Erythema multiforme, drug eruptions and urticaria are the other conditions which need to be excluded. Although erythema nodosum is the most confusing disease with Sweet's syndrome there are reported cases of histopathologically proven simultaneous occurrence of Sweet's syndrome and erythema nodosum (15).

The usual therapy for Sweet's syndrome is prednisone or prednisolone. However up to 30% of patients with Sweet's syndrome treated with corticosteroids had a relapse after the reduction of the steroid dosage. Alternatives to

corticosteroid treatment include potassium iodide, colchicine, dapsone, clofazimine and nonsteroidal anti-inflammatory drugs (NSAID). Action mechanisms of NSAID are inhibition of chemotaxis, migration and oxidative metabolism of neutrophils. Studies are continuing particularly on indomethacin (14).

We consider these two cases of Sweet's syndrome worth reporting, as upper respiratory tract infection is accused being of the triggering factor in both cases.

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