Sweet's Syndrome - An Interesting Skin lesion

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Abstract :

Introduction : Sweet's Syndrome(SS) also known as "Acute Febrile Neutrophilic Dermatosis" is classified as a neutrophilic dermatosis based on its chief clinico-pathological findings - sudden onset of painful, erythematous, edematous skin lesions (papules, plaques and nodules), with infiltration of mature neutrophils in dermis. We present a case of 41 years old female patient, who came to Sheth V.S General Hospital, Ahmedabad with chief complaints of multiple erythematous papular skin lesions over elbows, knees and back since 8 days. Laboratory investigations were done and punch biopsy was performed from the skin lesion and sent for histopathological examination; which revealed basal cell vacuolization of the epidermis and dense nodular perivascular neutrophilic infiltration, with prominent edema of upper dermis.

Key words : Acute febrile neutrophilic dermatosis, Neutrophilic infiltration, Sweet's syndrome

Introduction :

The Sweet's Syndrome (SS) was originally described by Dr.Robert Douglas Sweet in 1964, as an "Acute Febrile Neutrophilic Dermatosis".[1] Sweet's Syndrome is an uncommon inflammatory cutaneous disorder. The skin eruptions which frequently present on the face, neck, trunk and extremities are generally accompanied by signs of systemic inflammation including pyrexia, malaise and arthralgia. These lesions are often accompanied by leukocytosis and fever, with more than 75% of patients having systemic symptoms. [2, 3]

The etiology of Sweet's Syndrome is unknown, but several features suggest that the dermatosis results from a hypersensitivity reaction to an eliciting antigen; the source of antigen may be diverse such as bacterial, viral or tumor. It may be associated with antecedent infections, malignancies, autoimmune diseases, drugs and vaccines, granulocyte-colony stimulating factor as well as chemotherapy or idiopathic (associated with upper respiratory or gastro-intestinal infection ,viral infections, pregnancy or inflammatory bowel diseases).[1,4] Most commonly, it affects women between the ages of 30 to 70 years (female: male-7:1).

However, Sweet syndrome has been reported in children and younger adults also. Clinically, it is characterized by acute onset of fever, cutaneous manifestations such as raised painful, erythematous, well-demarcated papules and plaques, typically few centimeters in size on the face, neck, trunk and extremities. Systemic symptoms include malaise, fever, headache, conjunctivitis, and arthralgia. In some patients with malignancy, fever may be absent.

Histologically, a dense perivascular neutrophilic infiltration with leukocytosis is the hallmark of Sweet's Syndrome. [1, 5]

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Case report :

A 41 years female patient came to Sheth V.S General Hospital, Ahmedabad with chief complaint of multiple erythematous papular skin lesions over elbows, knees and back and with fever, since 8 days. She was otherwise asymptomatic without history of chills, joint pain or gastrointestinal symptoms. On examination, there was no genital or oral ulceration. Physical examination revealed multiple annular plaques showing central hyperpigmentation; and peripheral erythematous plaques over bilateral elbows, knees and back. No abnormality detected over palms, soles, scalp, nails, hairs, mucous membranes and in nerves. There were no anesthetic patches, hypopigmented patches or peripheral nerve thickening. Laboratory results showed elevated white blood cell count (16,000/cmm) with neutrophilic leukocytosis (78% neutrophils), erythrocyte sedimentation rate (ESR) was raised (48mm after 1 hour). C-reactive protein and Anti Streptolysin O(ASO) titers were within normal limits. Punch biopsy

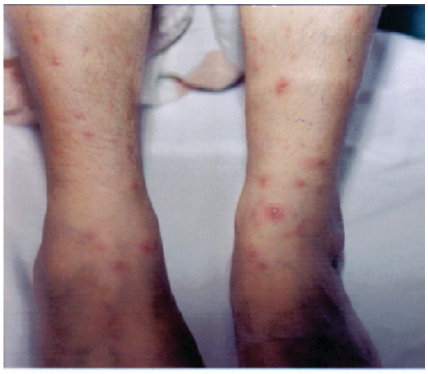
was performed from the skin lesion and was sent for histopathological examination.

Histopathological examination:

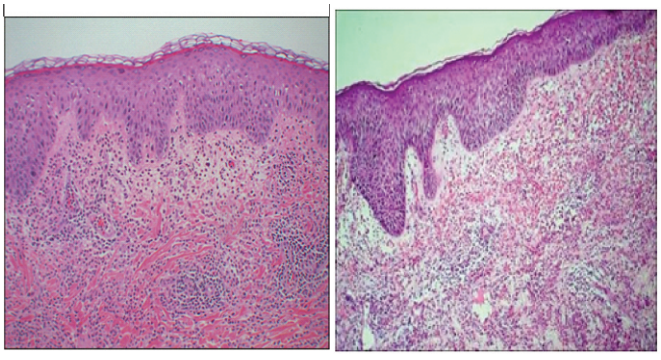
Macroscopic examination showed single skin covered soft tissue portion which was brownish in color and measured 0.4 cm. On cut section, it was whitish.

Microscopic examination revealed epidermis showing basal cell vacuolization. Upper dermis showed dense nodular perivascular neutrophilic infiltration accompanied by lymphocytes, histiocytes and occasional eosinophils. Papillary dermis showed edemawith inflammatory cells.

Figure 1: Multiple erythematous papular skin lesions over extremities



Figures 2 & 3: Dense nodular perivascular neutrophilic infiltrate with prominent edema of dermis.



Patient was given Prednisolone; the dose of which was tapered and stopped after gradual healing of the lesions. On regular follow up of the patient, there were no recurrences till date.

Discussion:

Sweet's syndrome (SS) was first described by Dr.Robert Douglas Sweet in 1964, as an "Acute Febrile Neutrophilic Dermatosis. [1] It is characterized by fever, neutrophilia, cutaneous eruptions consisting of erythematous papules and plaques and a dermal nonvasculitic neutrophilic infiltration on skin biopsy.[2, 3] These plaques are painful but nonpruritic. [6] Other skin manifestations such as pustules, vesicles, purpura, ulcers and hemorrhagic lesions have also been described. [1] Seventy five percent of patients have some prodromal illness, most commonly an upper respiratory tract infection. [7] Sweet's syndrome should be regarded as a cutaneous marker of systemic diseases. It has been associated with malignancies in about 20 to 25% of patients. [8] Common complications of Sweet's syndrome (SS) include arthralgia, arthritis, conjunctivitis, iridocyclitis and rarely involvement of central nervous system. Sweet's syndrome (SS) is more common in females (female: male -7:1) with the mean age of 52 years. [1] The pathogenesis of Sweet's syndrome (SS) is poorly understood. Cytokines, such as Granulocyte colony stimulating factor (G-CSF), interleukins (IL-1, IL-6 OR IL-8) deposited in the dermis, may be responsible for the immunopathogenic and clinical manifestations of Sweet's syndrome. [1]

For definitive diagnosis of Sweet's Syndrome, following major & minor criteria have been defined.

Two major criteria are: (1) Abrupt onset of painful erythematous plaques or nodules (2) Histopathologic evidence of a dense neutrophilic infiltrate without evidence of primary leukocytoclastic vasculitis.

The minor criteria are: (1) Pyrexia greater than 38°C. (2) Association with an underlying hematological or visceral malignancy, inflammatory disease, or pregnancy or preceded by an upper respiratory or gastrointestinal infection or vaccination. (3) Excellent response to treatment with systemic corticosteroids, potassium iodide, or colchicines. (4) Abnormal laboratory values at presentation (3 of the following 4): erythrocyte sedimentation rate greater than 20 mm/hr, positive C-reactive protein, greater than 8000 leukocytes, and greater than 70% neutrophils. [1,7]

Table-1 : Differential Diagnosis of Sweet's syndrome:

Sr. No.	Disease	Histological Findings
1.	Abscess/ cellulitis	Positive culture for infectious agent
2.	Bowel(intestinal) bypass syndrome	H/O jejuna-ileal bypass surgery for morbid obesity.
3.	Erythema elevatum diutinum	Erythematous asymptomatic plaques often located on the dorsum of hands and elbows; younger lesions have microscopic features of leucocytoclastic vasculitis whereas older lesions have dermal fibrosis & mucin.
4.	Granuloma faciale	Yellow to red to brown asymptomatic facial plaques; there is a grenz zone of normal papillary dermis beneath which there is a dense diffuse inflammatory infiltrate of predominantly neutrophils (with microscopic features of leukocytoclastic vasculitis) & frequently numerous eosinophils.
5.	Leukemia cutis	Dermal infiltrate consists of immature neutrophils
6.	Pyoderma gangrenosum	Painful ulcer with overhanging undermined violaceous edges.
7.	Rheumatoid neutrophillic dermatitis	H/O rheumatoid arthritis, nodules & plaques.
8.	Leucocytoclastic vasculitis	Vessel wall destruction – extravasated erythrocytes, fibrinoid necrosis of vessel walls, karyorrhexis & neutrophils in the vessel wall.
9.	Neutrophillic eccrine hidradenitis	Neutrophils around eccrine glands, often in patients with acute myelogenous leukemia receiving induction chemotherapy.

Both of the major criteria and any two minor criteria should be met for the diagnosis.

Table-1 Shows the Differential Diagnosis of Sweet's syndrome.

Conclusion :

Sweet's Syndrome has a broad spectrum of clinical and pathologic findings in various areas of the world. Further investigations are necessary to determine the etiology and effect of environmental factors on the disease. We appreciate the difficulty in the initial diagnosis of Sweet Syndrome. Patient education should include the information about the variable course of this condition, as well as advice on self monitoring for signs and symptoms of other diseases.

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