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PHOTODISTRIBUTED ACUTE FEBRILE NEUTROPHILIC DERMATOSIS: A CASE REPORT

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Abstract
Photoexposed area involvement in acute febrile neutrophilic dermatosis is uncommon and has rarely been described in literature. We report a case of photodistributed Sweets syndrome in a middle aged lady who responded to symptomatic treatment with NSAID and systemic antibiotic. Sweets syndrome is an uncommon inflammatory disorder characterized by the abrupt appearance of painful, oedematous and erythematous papules and plaques or nodules on the skin. Fever and leukocytosis frequently accompany cutaneous lesions. Our patient presented with fever and abrupt onset of painful erythematous plaques over exposed parts of the body.

Key words: Acute febrile Neutrophilic dermatosis; photodistributed; colchicine

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Introduction

Acute febrile neutrophilic dermatosis or Sweet's Syndrome (SS) is characterized by a constellation of clinical symptoms, physical features, and pathologic findings which include fever, neutrophilia, tender erythematous skin lesions and a diffuse infiltrate consisting predominantly of mature neutrophils that are typically located in the upper dermis. Sweet's syndrome presents in three clinical settings: classical (or idiopathic), malignancy-associated, and drug-induced [1]. Approximately 70% of cases are idiopathic and the paraneoplastic form is present in 10-20 of the cases [2]. We present an unusual presentation of a case of Idiopathic or Classical Acute neutrophilic dermatosis in middle aged lady involving photoexposed regions and review the literature.

Case Report

A 45 year old woman presented to the outpatient department with history of painful lesions over the face and extremities accompanied by fever. She gave history of photosensitivity with burning sensation over the forearms. There was no history of sore throat, cough, arthralgia, drug intake or any systemic complaints prior to the present illness. Clinical examination revealed erythematous well defined tender plaques with raised margins over the V area of neck (Fig. 1), malar region (Fig. 2), bilateral palms, and legs, with pseudovesiculation over extensors of forearms and dorsum of hands (Fig. 3). The photo-protected areas were spared. A clinical differential diagnosis of Sweet's syndrome, Hansen's disease with type 2 reaction and erythema multiforme was considered. General examination revealed that she was febrile with a temperature of 102° and mild anemia. s

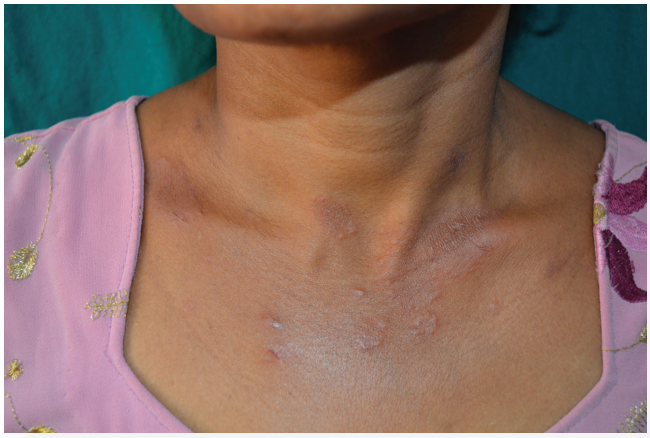

Figure 1. Multiple erythematous plaques and papules on V area of neck.

Figure 2. Erythematous plaque on left malar area.

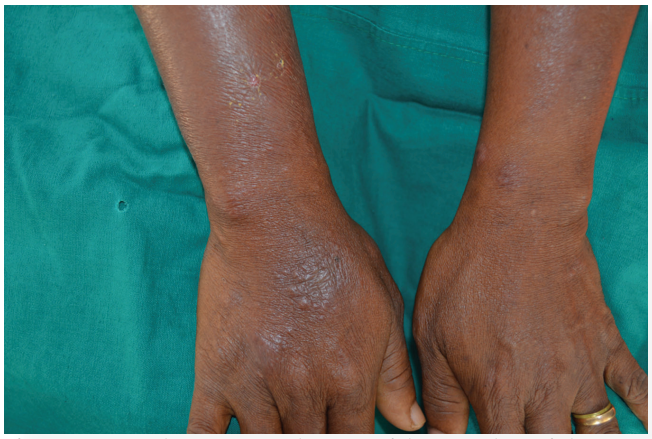

Figure 3. Erythematous plaque with pseudovesicle over dorsum of hand.

The photo-protected areas were spared. A clinical differential diagnosis of Sweet's syndrome, Hansen's disease with type 2 reaction and erythema multiforme was considered. General examination revealed that she was febrile with a temperature of 102° and mild anemia. Investigations revealed a Haemoglobin of 10.6 gm/dl, neutrophilia of 72% and raised ESR of 106mm/1 hour. ASO and ANA tests were negative, but C - reactive protein was positive. A slit skin smear examination was negative for acid fast bacillus. Systemic and radiological examination for underlying cause revealed no abnormalities. A skin biopsy was performed. Microscopic examination of skin biopsy showed marked edema in papillary dermis (Fig. 4) with diffuse interstitial and perivascular infiltration by predominantly neutrophils and blood vessels lined by plump endothelial cells with no evidence of vasculitis (Fig. 5). Patient was treated symptomatically with NSAID,s topical steroids and systemic antibiotics. Parenteral amoxicillin and clavulinic acid in the dose of 1.2 gm three times a day was used intravenously for the initial 3 days and then switched over to oral for a total duration of 7 days. Classical Sweets syndrome is usually associated with underlying infections and our patient had fever, abrupt onset of painful lesions and her ESR was raised with positive C reactive protein hence a course of systemic antibiotic was used. Clobetsol propionate 0.05% cream 30 gm diluted with 70 gms of cream base was used topically twice daily for 7 days and then once daily for 7 days. She improved and the lesions healed with scaling (Fig. 6). As she had dramatic improvement with systemic antibiotic and NSAID's, systemic steroids were not considered. She had a recurrence after 2 weeks and was treated with colchicine 0.5 mg three times a day. She has been on colchicine for about 2 months without any recurrences.

Discussion

Acute neutrophilic dermatosis was first described in 1964 by Robert Douglas Sweet, and has been termed Sweet's syndrome [3]. Classic Sweet's syndrome occurs in middle-aged women after a nonspecific infection of the respiratory or gastrointestinal tract. Raised erythematous plaques with pseudoblistering and occasionally pustules occur on the face, neck, chest, and extremities, accompanied by fever and general malaise. Involvement of the eyes, joints, and oral mucosa as well as internal manifestations of Sweet's syndrome in the lung, liver, kidneys, and central nervous system has been described. The disease is thought to be a hypersensitivity reaction. Approximately one-third of patients with Classical Sweets syndrome experience a recurrence of the dermatosis.

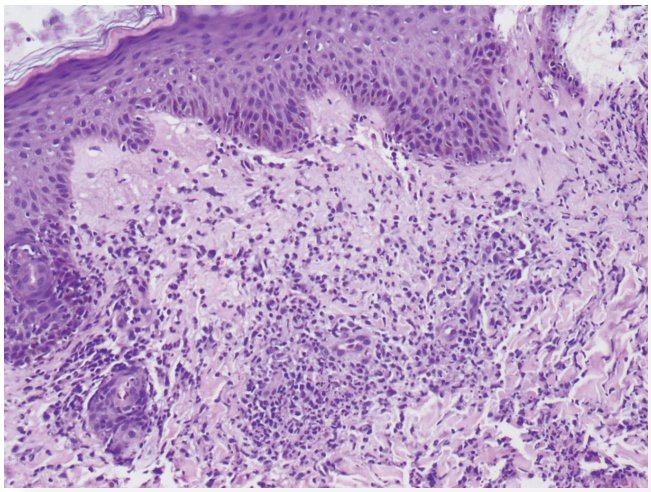

Figure 4. Photomicrograph showing marked edema in papillary dermis.

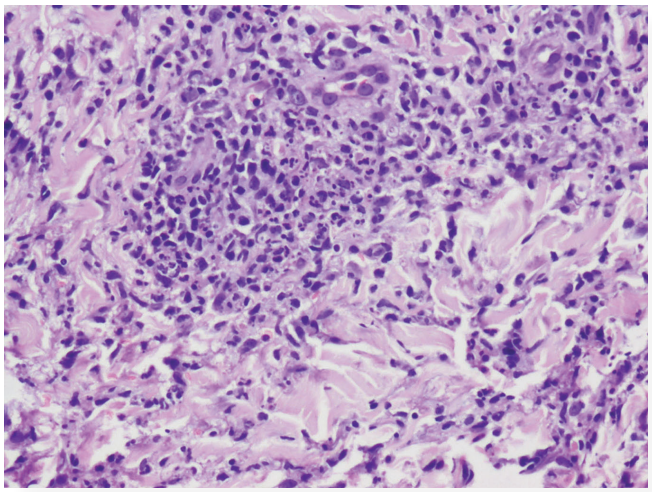

Figure 5. Diffuse infiltration of the dermis by neutrophils with no evidence of vasculitis H & E.

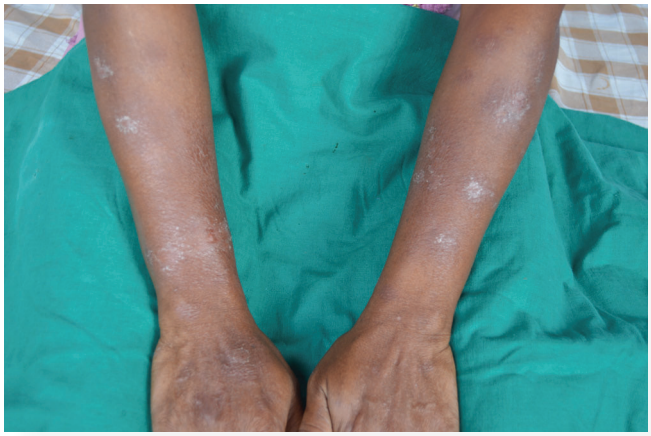

Figure 6. Healing of the lesions with scaling.

The malignancy-associated Sweet's syndrome can occur as a paraneoplastic syndrome in patients with an established cancer or individuals whose Sweet's syndrome-related hematologic dyscrasia or solid tumor was previously undiscovered; the dermatosis can precede, follow, or appear concurrent with the diagnosis of the patient's cancer. Hence, can be the cutaneous harbinger of either an undiagnosed visceral malignancy in a previously cancer-free individual or an unsuspected cancer recurrence in an oncology patient. Drug-induced Sweet's syndrome most commonly occurs in patients who have been treated with granulocyte-colony stimulating factor; however, other medications may also be associated with this form of Sweet's syndrome. The pathogenesis of Sweet's syndrome may be multifactorial and still remains to be definitively established. Clinical and laboratory evidence suggests that cytokines have an etiologic role.

Histologically the hallmark is a diffuse infiltrate consisting predominantly of mature neutrophils typically located in the upper dermis and no evidence of leukocytoclastic vasculitis.

Extracutaneous manifestations in the form of alveolitis, sterile osteomyelitis, renal, hepatic, and central nervous system involvement have also been reported .Although it is not one of the common life-threatening dermatoses, Sweet's syndrome can potentially cause significant pulmonary involvement and respiratory compromise and one needs to be aware of this condition.

Drug induced SS in photo-exposed regions have been described previously [4]. There are occasional case reports describing the worsening of idiopathic Sweet syndrome after sun exposure or photo distribution of lesions [5]. Lesions have also been described at site of previous phototoxic reaction [6]. The pathomechanism could involve either an isomorphic Koebner reaction, classically described in neutrophilic dermatoses, or the direct action of UV-B on neutrophil activation and recruitment in skin through the production of cytokines, such as interleukin 8 or tumor necrosis factor α [7]. Our patient presented with abrupt onset of fever and painful erythematous plaques over extensors of forearms, V area of neck and both malar area. She had a raised ESR, neutrophilia, positive C reactive protein and histopathology showed perivascular neutrophilic infiltration without evidence of vasculitis satisfying 2 major and 2 minor criteria's to arrive at a diagnosis of Sweets syndrome.

Systemic corticosteroids are the therapeutic gold standard for Sweet's syndrome. After initiation of treatment with systemic corticosteroids, there is a prompt response consisting of dramatic improvement of both the dermatosis-related symptoms and skin lesions. Topical application of high potency corticosteroids or intralesional corticosteroids may be efficacious for treating localized lesions. Other first-line oral systemic agents are potassium iodide and colchicine. Second-line oral systemic agents include indomethacin, clofazimine, cyclosporine, and dapsone. The symptoms and lesions of Sweet's syndrome may resolve spontaneously, without any therapeutic intervention; however, recurrence may occur.

Systemic antibacterials against Staphylococcus aureus frequently result in partial improvement of Sweet's syndrome lesions when they are impetiginized or secondarily infected. In some patients with dermatosis-associated bacterial infections, organism-sensitive specific systemic antibacterials have been helpful in the management of their Sweet's syndrome. Although patients with hematologic malignancy-associated Sweet's syndrome often receive cytotoxic chemotherapy agents and antimetabolic drugs for the treatment of their underlying disorder, these agents are seldom used solely for the management of the symptoms and lesions of Sweet's syndrome. Spontaneous resolution is not uncommon in classical SS. Recurrence may occur in one third patients despite appropriate treatment. Our patient responded to a course of systemic antibiotic, topical steroid and non-steroidal anti-inflammatory drug and did not need systemic steroids. The relapse which occurred after 2 weeks was well controlled with colchicine. Knowledge about this entity and its early recognition and prompt treatment is important to prevent devastating outcomes.

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