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A Rare Case of Classical Sweet Syndrome – Case Report

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ABSTRACT

This case report presents a 30 year-old female who developed an acute febrile illness with painful, erythematous plaques over the left lower limb and right upper limb. A histopathological study revealed extensive neutrophilic infiltrates in the dermis, which is consistent with Sweet Syndrome. The patient responded favorably to systemic corticosteroid treatment, with the skin lesions and systemic symptoms cleared. This example shows the importance of detecting Sweet Syndrome, as well as the necessity for early diagnosis and fast treatment to reduce morbidity and potential consequences

Keyword: *Neutrophilic dermatosis , Erythematous plaques, immune-mediated hypersensitivity, Systemic corticosteroids*

1. INTRODUCTION

Leukocytosis with neutrophilia, a sudden painful papulonodular skin eruption linked to a febrile prodrome, and histological evidence of neutrophilic infiltration of the upper dermis in the absence of leukocytoclastic vasculitis are the hallmarks of Sweet's syndrome, also known as acute febrile neutrophilic dermatosis, a rare inflammatory disease. Generally speaking, there are three forms of Sweet's syndrome: drug-induced, malignancy-associated, and classical (or idiopathic). It is believed to be the most common example of a class of illnesses called neutrophilic dermatoses.[1-3] Though the exact etiology remains unknown, studies conducted since Sweet's syndrome was identified have indicated that autoinflammatory pathways involving the innate as well as adaptive immune system ultimately result in their failure leading to immune-mediated hypersensitivity and a role for cytokines such as IL-1β, IL-17, and TNF-α. 5-[9]. In the absence of vasculitis, skin biopsy results of diffuse neutrophilic dermal aggregations play a crucial role in the diagnosis, which is established worldwide utilizing a diagnostic strategy that uses major and minor criteria. Systemic corticosteroids continue to be the mainstay of therapeutic approaches.

2. CASE PRESENTATION

A 30 years old female who is a known case of Auto inflammatory Syndrome since 2021 and managed with steroids came to our OPD with reports of a high-grade fever, Dry cough and Sore throat for past 6 days. History of Painful Left lower limb swelling till thighs along with some rashes was present. On Examination Patient was Febrile with tender swelling of Left Lower Limb. Painful erythematous rashes was over Left lower limb and Right upper limb. All routine investigations were done. CBC showed Neutrophilic leucocytosis. CRP and ESR was elevated(391 and 87 respectively). Rheumatology opinion was obtained and advised to do C- ANCA , P- ANCA , ANA- IFA, Sr Fibrinogen and Sr LDH levels and orders were followed. Dermatology opinion was obtained and advised for Skin Biopsy. C- ANCA and P- ANCA was negative. Sr Fibrinogen levels were mildly elevated(846). Sr LDH was within Normal limits. ANA -IFA showed weakly positive for speckled pattern. Skin Biopsy showed LOBULAR NEUTROPHILIC PANNICULITIS. SUBTLE SUPERFICIAL DERMAL OEDEMA. DERMAL PERIVASCULAR NEUTROPHILIC INFILTRATES with possibility of subcutaneous SWEET'S SYNDROME may be considered after clinical correlation.

Image 1; Day 1 Erythematous lesions in Left lower limb and Right Upper limb

Rheumatology review was obtained and advised for Steroid pulse therapy i.e. Inj. Methyl Prednisolone 1 gram for 3 days and NSAIDs. Patient improved clinically after every dose of steroid. Rashes and pain were in decreasing trend as shown in pictures.

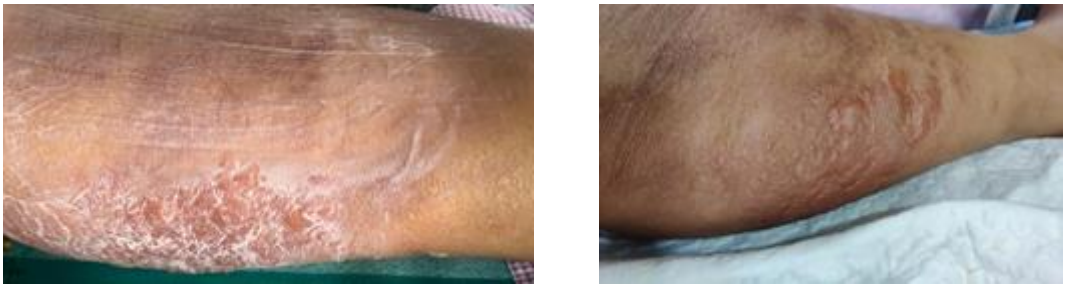

Image 2; Resolving Erythematous lesions in Left lower limb after starting Steroids

Rheumatology review was obtained before discharge and advised for low dose oral steroids and NSAIDs for 10 days. Patient was symptomatically improved and hence discharged with medications as prescribed by Rheumatologist. On Follow-up after 10 days , Skin lesions was completely resolved and remained symptoms free and steroids were tapered down over a period of 6 weeks.

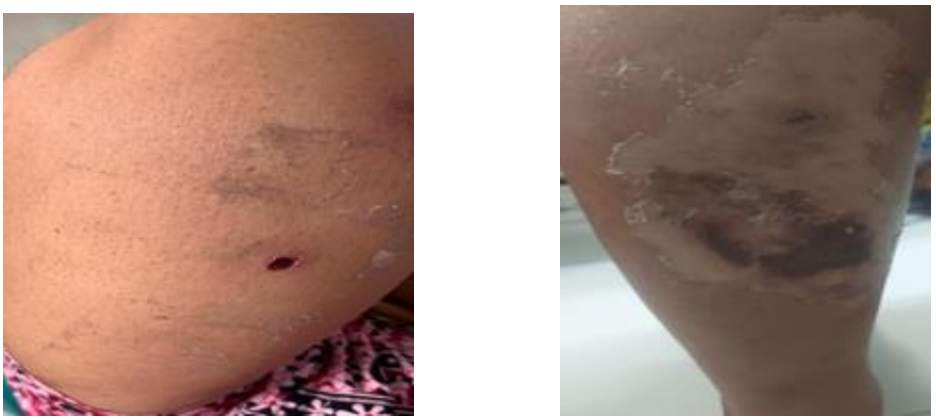

Image 3: Lesions getting resolved on Follow-up

3. CONCLUSION

Sweets syndrome is characterized by sudden development of painful erythematous or violaceous plaques or nodules, as well as cutaneous neutrophilic infiltration, without leucocytic vasculitis. A few minor criteria that were clarified were elevated Lymphocytes , arthralgia, conjunctivitis, Upper respiratory tract infection or Gastrointestinal infection prior to disease , a favorable response to steroids, and a potential underlying malignancy. Using these criteria, our patient may be diagnosed with Sweet syndrome without having any connection to cancer. Due to its rarity, this case has been documented so that the consulting physician might consider it as a differential diagnosis among a number of related illnesses

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