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Case Report

A case of classical sweet syndrome with episcleritis

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ABSTRACT

Sweet's syndrome (acute febrile neutrophilic dermatosis) is characterized by a constellation of clinical symptoms, physical features, and pathologic findings which include fever, neutrophilia, tender erythematous skin lesions (papules, nodules, and plaques), and a diffuse infiltrate consisting predominantly of mature neutrophils that is typically located in the upper dermis. Sweet's syndrome presents in three clinical settings: classical (or idiopathic), malignancy-associated, and drug-induced. Classical Sweet's syndrome (CSS) usually presents in women between the age of 30 to 50 years, is often preceded by an upper respiratory tract infection and may be associated with inflammatory bowel disease and pregnancy. We report here a case of classical sweets syndrome with the typical histopathological findings who reported well to treatment.

Keywords: Classical, Episcleritis, Neutrophilic dermal infiltrate, Sweets syndrome

INTRODUCTION

Sweet's syndrome (acute febrile neutrophilic dermatosis) is characterized by a constellation of clinical symptoms, physical features, and pathologic findings which include fever, neutrophilia, tender erythematous skin lesions (papules, nodules, and plaques), and a diffuse infiltrate consisting predominantly of mature neutrophils that is typically located in the upper dermis.[1] Sweet's syndrome presents in three clinical settings: classical (or idiopathic), malignancy-associated, and drug-induced. Classical Sweet's syndrome (CSS) usually presents in women between the age of 30 to 50 years, is often preceded by an upper respiratory tract infection and may be associated with inflammatory bowel disease and pregnancy. We report here a case of classical sweets syndrome with the typical histopathological findings who reported well to treatment.

CASE REPORT

This is the case of a 50-year-old female housewife from Kozhikode who presented with complaints of fever of 1 week duration, fatigue/body ache/arthralgia of 1 week, erythematous rash over body of 5 days and redness of both eyes of 2 days duration. Her fever was low grade associated with severe fatigue, malaise, body ache and arthralgia, 2 days into the fever developed mildly painful, non-pruritic erythematous skin lesions which appeared initially on her back and later on both upper extremities. Three days later she also had redness of both her eyes. There was history of a preceding upper respiratory infection 2-3 wks back and history of drug intake for the same details of which were not available. There was no history of arthritis, no history of dysuria, no discoloration of urine, no history of abdominal pain or loose stools, no recurrent oral ulceration, photosensitivity or hair loss, no history of significant weight loss. She was a post-menopausal female, mother of two children with past history of atopy. There was no past of drug allergy. There was no history of any significant medical illnesses like diabetes mellitus, systemic hypertension, tuberculosis in the past.

So, the clinical possibilities considered from this history were of any cutaneous infections, drug induced rash, an underlying collagen vascular disease/vasculitis or sweet syndrome.

Clinical examination of the patient showed that she was conscious, oriented. There was an erythematous mildly tender non-pruritic plaque like rash predominantly on the back and upper extremities.

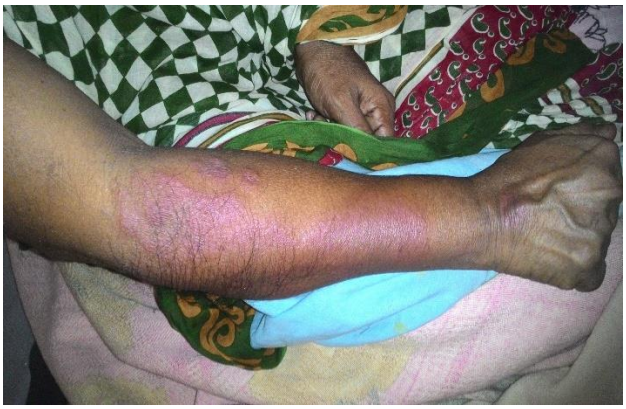

Figure 1: Skin rashes noted over extensor aspect of right forearm.

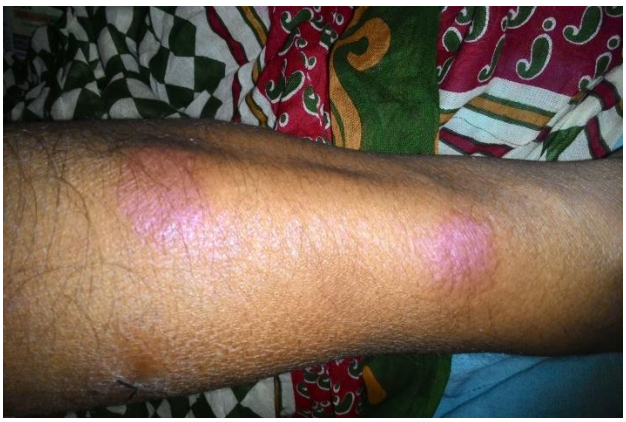

Figure 2: Erythematous non pruritic plaque skin lesions.

There was no evidence of pallor, icterus, clubbing, lymphadenopathy. She had redness of both eyes which on further ophthalmological evaluation showed dilated episcleral vessels suggestive of episcleritis.

Her vital signs were stable with a pulse rate was 88/min, regular blood pressure of 120/80 mmHg, respiratory rate was 18/min and temperature- 101 degree Fahrenheit. System examination of the patient was within normal limits, no evidence of any oral ulceration or hepatosplenomegaly.

Her Investigations were as follows total count-14800, differential count- P73L21M6, Hb-11.5 g/dl, Hemocrit-32.2%, mean corpuscular volume- 88fl, platelet count-4.21 lakhs/cumm. ESR was elevated initially being 100 mm/hr and repeat value was 67 mm/hr, urine routine-alb-nil, 1-2pc, Urine culture and sensitivity was sterile. Random blood sugar-100 mg/dl, blood urea/serum creatinine- 13/1.0, total bilirubin/conjugated bilirubin-0.6/0.2, total protein/albumin- 6.9/3.4, alanine aminotransferase/alkaline phosphatase- 24/106. Her C-reactive protein was >48, A-894 cells/cumm, Anti-nuclear antibody by immunofluorescence- negative, HIV, HBsAg, HCV screening was done which was negative, chest x-ray- normal, electrocardiogram was normal, ultrasound abdomen was normal, peripheral smear showed neutrophilia, no blasts seen. Skin biopsy was performed which showed:

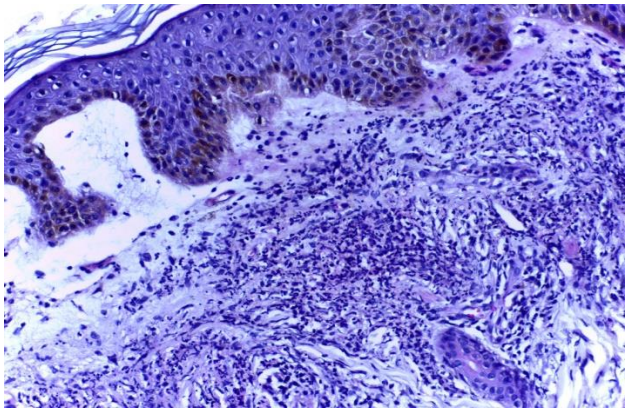

Figure 3: Histopathology showing dense neutrophilic dermal infiltrate suggestive of sweet syndrome.

High power view of the biopsy specimen showed dense neutrophilic infiltrate into the dermis particularly around the vessels which was consistent with the diagnosis of sweet syndrome.

DISCUSSION

Sweet syndrome/acute febrile neutrophilic dermatosis are of 3 types: classical/idiopathic, malignancy associated and drug-induced variants. Diagnostic criteria for classical/idiopathic sweet syndrome has 2 major and 4 minor criteria. Major criteria include: abrupt onset of painful erythematous plaques or nodules and histopathologic evidence of a dense neutrophilic infiltrate without evidence of leukocytoclastic vasculitis. Minor criteria are: pyrexia>38°C, association with an underlying hematologic or visceral malignancy, inflammatory disease or pregnancy or preceded by upper respiratory or gastrointestinal infection or vaccination, excellent response to treatment with systemic corticosteroids or potassium iodide and abnormal laboratory values at presentation (3 out of 4)- ESR>20mm/hr, positive CRP>8000 leukocytes and >70% neutrophils. To fulfil the diagnostic criteria both major and atleast 2 out of 4 minor criteria was needed to be satisfied for the diagnosis of idiopathic sweet syndrome.[2] In present case the patient satisfied both the major and 3 out of 4 of the minor criteria. Since we had not started the patient on corticosteroids only one minor criteria remained to be fulfilled.

Review of literature also showed that of the extra cutaneous manifestations of sweet syndrome ocular

inflammation is a common finding in 17-72% which included conjunctivitis, episcleritis, scleritis, limbal nodules, iritis, keratitis, glaucoma, choroiditis.[3]

The histopathologic findings typically described in sweet syndrome are: prominent erythema in superficial dermis, dense infiltrate of neutrophils in the upper and mid-dermis with sparing of epidermis, leukocytoclasis, endothelial swelling, absence of vasculitis.[4] So the final diagnosis of idiopathic sweet syndrome with episcleritis was made.

The treatment of sweet syndrome includes: corticosteroids-systemic and topical, oral prednisolone 0.5-1mg/kg/day which is tapered off weekly. Other first line agents include: colchicine- 1-1.5mg per day, dapsone- 25mg/day and potassium iodide- 300mg TID.[5] The skin lesions of the patient subsided within 1 week and she was started on corticosteroids on outpatient basis and there was rapid resolution of the lesions.

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