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Sweet Syndrome -A Case Report

Dr. Deepsheeka .G[1], Dr. Badrinath. A.K[2]
[1]Post Graduate, [2]Professor and Unit Chief,
Department Of General Medicine,
Sri Manakula Vinayagar Medical College and Hospital

***Corresponding Author:**
Dr. Deepsheeka . G
Post Graduate, Department Of General Medicine,
Sri Manakula Vinayagar Medical College and Hospital

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Abstract

Sweet syndrome is a rare disorder that is manifested by a constellation of clinical features, including fever, neutrophilic leucocytosis, raised painful plaques on skin and dermal infiltration by neutrophils.[1] Numerous etiological associations have been reported in the literature, including infections,autoimmune,malignancy or drug induced. We present a case of association of mixed connective tissue disorder with sweet syndrome . Here we describe a case of a man aged 45 years,who presented with complaints of fever and was evaluated as pyrexia of unknown origin and as Sweet's syndrome. He was successfully treated with systemic steroids.

Keywords: Sweet syndrome,Acute neutrophilic dermatosis ,MCTD,fever with rash,Connective tissue disorder,Pyrexia of unknown origin

Introduction

A 45 year old male came with complaints of fever for 1 week ,intermittent ,low grade fever not associated with chills and rigors.H/o multiple joint pain with significant left shoulder joint pain associated with restricted movements.H/o skin lesion present over the chest and upper back .H/o oral ulcers present.No h/o sorethroat,weight loss.No h/o cough with expectoration ,burning micturition.No h\o vomiting,loose stools,headache,multiple small joint pain .No h\o any recent or chronic drug intake .No h\o similar complaints in the past.No h\o any recent travel history.Patient has no comorbidities .Patient is not an alcoholic or smoker .Pateint was vitally stable.Systemic examination was found to be normal .On examination Ill defined erythematous to hyperpigmented plaques over upper chest and few skin coloured tender nodules were present over the upper back (Figure 1.1).Routine investigations were done showed Hb -10.7 g/dl,total counts of 8400cells/cu.mm with 71% neutrophils ,platelets-214 x 1000/uL,ESR - 120mm ,serum urea 16 mg\dl ,serum creatinine was 1.0 mg\dl ,RBS -125 mg\dl ,urine protein - trace ,urine WBC-2-3 /hpf .Liver function test was normal.ASO titre <200 IU/ml.RA Factor - <8 IU/ml .HIV,HbsAg,HCV - Non reactive .Serum LDH - 573 IU/L.Tropical fever workup(Dengue ,scrub) - Non reactive .Sputum AFB D1 and D2 - no AFB .Mantoux test - negative.Bone marrow aspiration showed hypercellular reactive marrow .Bone marrow C&S showed no growth .USG abdomen and pelvis done showed no significant abnormality .CT thorax was done given as normal study.Patient was initially started on Inj .Ceftriaxone along with Doxycycline but as frequent fever spikes were present stepped up to Inj Meropenam and started on Antimalarials .But patient still had persistent fever spikes .ANA [IFA]done came as 2+ positive - nuclear coarse granular pattern .ANA profile done showed U1 RNP and Smith antibodies positive suggestive of Mixed connective tissue

disorder .Skin biopsy done (Fig 1.2) showed Basket weave hyperkeratosis ,mild spongiosis . Perivascular and periadnexal neutrophilic infiltrate.Nerve conduction study was normal .EMG showed myogenic pattern of involvement in right vastus lateralis and muscle biopsy was planned on follow up .

Patient was started on T. Predinisolone 1 mg/kg and patient became symptomatically better and skin also resolved .(Figure 1.3).

Figure 1.1 Day 1 of admission

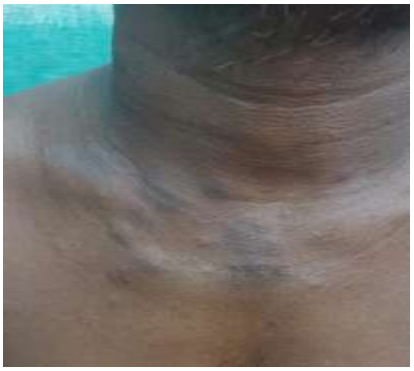


Figure 1.2 Skin biopsy

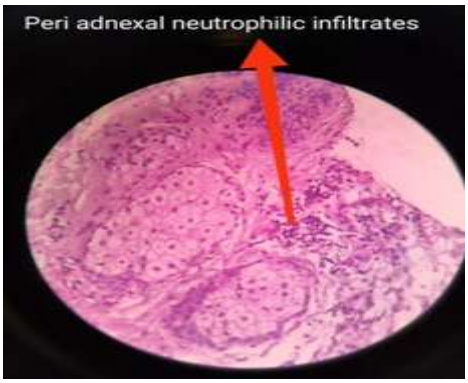
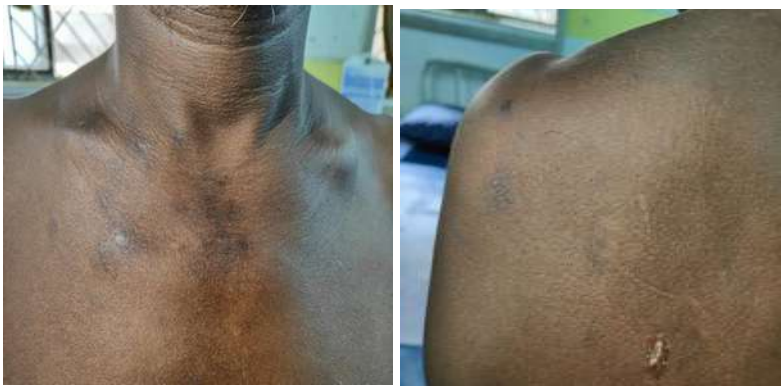


Figure 1.3 Day of discharge



Discussion:

Sweet syndrome or Acute febrile Neutrophilic dermatosis or Gomm Button disease has a 4:1 female to male predominance and average age group involved is 30 to 60 years .

Etiology can be

1. Infections - Streptococcal Upper respiratory , GIT, Mycobacterial , sporotrichosis, crytococcosis
2. Autoimmune : Collagen vascular disorders, immune thrombocytopenic purpura ,pemphigus vulgaris, autoimmune thyroiditis , IBD
3. Drug induced - G-CSF,trans retinoic acid, imatinib, bortezomib, OCP's, propylthiouracil
4. Pregnancy associated .

Pathophysiology is unknown but a few hypotheses include dysregulation of cytokine secretion ,defect in transcriptional regulation of PTPN 6 gene ,mutations of MEFV gene (alteration in proinflammatory cell signalling pathways).

Histopathological findings include in the Epidermis : Sub corneal pustules ,epidermal spongiosis ,frank intradermal or subepidermal vesiculation ;Dermis : Diffuse nodular and perivascular neutrophilic infiltrate ,leukocytoclasia ;Subcutaneous fat : Extension of dermal infiltrate into subcutis .Classical clinical appearance - erythematous ,tender,non pruritic,papules ,nodules or plaques with predominant

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head,neck and trunk distribution pattern. Genital and oral ulcerations are uncommon but aphthous ulceration can be present .There are various subtypes such as bullous, cellulitis like, necrotising .

Systemic involvement may present as Fever, Arthralgias or arthritis (asymmetric ,non erosive and sterile) , Neutrophilic alveolitis, Multiple focal osteomyelitis, Mesangial glomerulonephritis, Acute myositis , Hepatitis, pancreatitis, Colitis, Neuro sweet's disease .

Diagnostic criteria for sweet's syndrome(2 major +2 minor) :

Major criteria:

- Abrupt onset of typical cutaneous lesions
- Histopathology consistent with Sweet syndrome.

Minor criteria:

- Preceded by one of the associated infections or vaccinations, accompanied by one of the associated malignancies or drug exposure or pregnancy
- Fever >38 C
- Abnormal laboratory values at presentation (3 out of the 4 required) a. erythrocyte sedimentation rate (ESR)>20mm/hour, b. leukocytes>8000/mm3, c. neutrophils>70%, d. Elevated C reactive protein (CRP).
- Dramatic response to systemic corticosteroids or potassium iodide (KI).[2]

Treatment for sweet's syndrome in case of mild localised disease by potent topical or intralesional corticosteroids For systemic disease : 0.5 mg to 1 mg per kg prednisolone for 4 to 6 weeks .If remission not acheived by 3 months a second line steroid sparing agent such as Dapsone (50 to 100 mg /day),colchicine (0.5 mg 3 times daily) can be given .Other drugs which have been tried include Acitretin, Baricitinib, Adalimumab, Anakinra, Infliximab, Rilonacept, Tocilizumab, Potassium iodide, Thalidomide.

Conclusion :Sweet syndrome is mostly a presentation of an underlying condition , thus patient should be throughly evaluated and treated accordingly .

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