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Case Study

Case report on Acetaminophen induced Gomm - Button disease

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ABSTRACT

The Gomm-Button disease/Sweet's syndrome is a rare and reactive disorder so often idiopathic, sometimes associated with diseases and drugs rarely. Herein, we report a case of a 49 year old female patient, presented to the dermatology department with a 3 day history of low grade fever and 1 day history from congestion of eyes, along with development of multiple itchy and painful bullous vesicular skin lesions distributed over face and upper limbs. Lab Examination revealed elevated white blood cells and negative IgE. Histopathological and cytology reports showed neutrophilic infiltrates which were suggestive of Gomm-Button disease.

Key words: Sweet's syndrome, acute febrile dermatosis, Acetaminophen, Drug Induced Sweet Syndrome (DISS)

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INTRODUCTION

Gomm-Button disease, also known as acute febrile neutrophilc dermatosis or Sweet's syndrome (SS), was first described by the scientist Robert Sweet in 1964[1]. It is a rare and uncommon systemic inflammatory disorder characterized by constellation of symptoms and findings such as high fever, neutrophilic leukocytosis and abrupt onset of tender erythematous, painful and itchy cutaneous lesions. SS is associated with a range of disorders and is categorized into classical/ idiopathic, malignancy-associated and drug induced based on the etiological factors.

It is crucial to understand the pathophysiology of the disease for the management of neutrophilic-dermatosis. Classical/idiopathic Sweet's syndrome (CSS) (≥70%) affects predominantly middle aged women (30-60 years old) and have an antecedent upper respiratory tract infection especially due to streptococcus.[2,3] Other underlining conditions associated with CSS are viral/bacterial GI infection, recent vaccination, pregnancy and IBD. A few case reports revealing the association of pulmonary tuberculosis with Sweet's syndrome have also been reported in India which is rare [4]. Malignancy associated Sweet's syndrome (20%) can occur before, after or concurrently with hematologic malignancy (85%) of which Sweet's syndrome associated with Acute Myelogenous Leukaemia is the commonest or a solid tumour (15%) and patients with Hodgkin's disease[5]. This subtype tends to affect older patients with an average age of about 68 years at diagnosis.[3]

The management of patients with Sweet's syndrome can be performed in 3 steps: assessment, workup and treatment. Assessment includes the identification of the type of lesions, the existence of possible extracutaneous site, and the search for associate disease. It is important to rule out infection and to have precise drug therapy history. Laboratory evaluation for complete blood count including total leukocyte count and platelet count should be a part of the workup plan. Evaluation for acute phase reactant reactants like ESR, CRP and serum chemistries to evaluate renal and hepatic function, and a urinalysis can be performed.[2] Consistent laboratory evaluations in Gomm-Button disease include total leukocyte count, peripheral leucocytosis with neutrophilia and an elevated ESR. To confirm a clinically suspected Sweet's syndrome, a skin lesion biopsy for routine histopathological examination is performed. it is mandatory to perform the in depth analysis of chest radiographs, SPECTs, computerized axial tomography, electroencephalograms, magnetic resonance imaging and even cerebrospinal fluid, to rule out the extracutaneous involvement.[2]

CASE REPORT

A 49 year old female patient with no relevant medical history was admitted to the ward with a 1 day history of erythematous lesions on face and mostly distributed over the upper limbs associated with 3 day history of low grade fever (100.8°F) and congestion of eyes. New lesions and bullae developed over feet and palm.The rest of her systemic examination was unremarkable. Admission laboratory results revealed erythrocyte sedimentation rate (78 mm/hr) on the first day and (113 mm/hr) on the second day, neutrophil count (78%), urinalysis showed significant bacteriuria. Antibiotic therapy was initiated with amoxicillin clavulunate for 3 days suspecting Erythema multiforme, no significant clinical response was observed. Patient developed a progression of erythematous bullae over feet and palm. Skin biopsy for immune fluorescence showed IgG, IgM, IgA and C_3 to be negative. Epidermal region showed hyperkeratosis and keratotic plugging, subepidermal region showed congestion, melanophages and infiltrate of lymphocytes and plasma cells while there was dense neutrophilic infiltrates in the deeper serial section of subepidermal region around the capillaries during histological examination which were suggestive of Sweet's syndrome. Antibiotic therapy was stopped. Patient was treated with dexamethasone (1g IV for 3 days) followed by oral prednisone.

On taking medication history interview she had a relevant history of taking paracetamol tab from fever.

DISCUSSION

SS can present as one of the 3 clinical variants; classical/idiopathic, malignancy associated and drug induced.[3] In drug induced Sweet's syndrome, there is always a temporal relationship between medication administration and the development of symptoms. The diagnostic criterion for DISS was described by Walker and Cohen in 1996 is being explained in (Table 1).[2]

1	Abrupt onset of tender or painful erythematous plaques or nodules
2	Dense dermal neutrophilic infiltrate without vasculitis
3	Fever > 38°C
4	Temporal relationship between drug administration and clinical manifestations
5	Temporally relationship between resolution of lesions with drug withdrawal or treatment with systemic corticosteroids

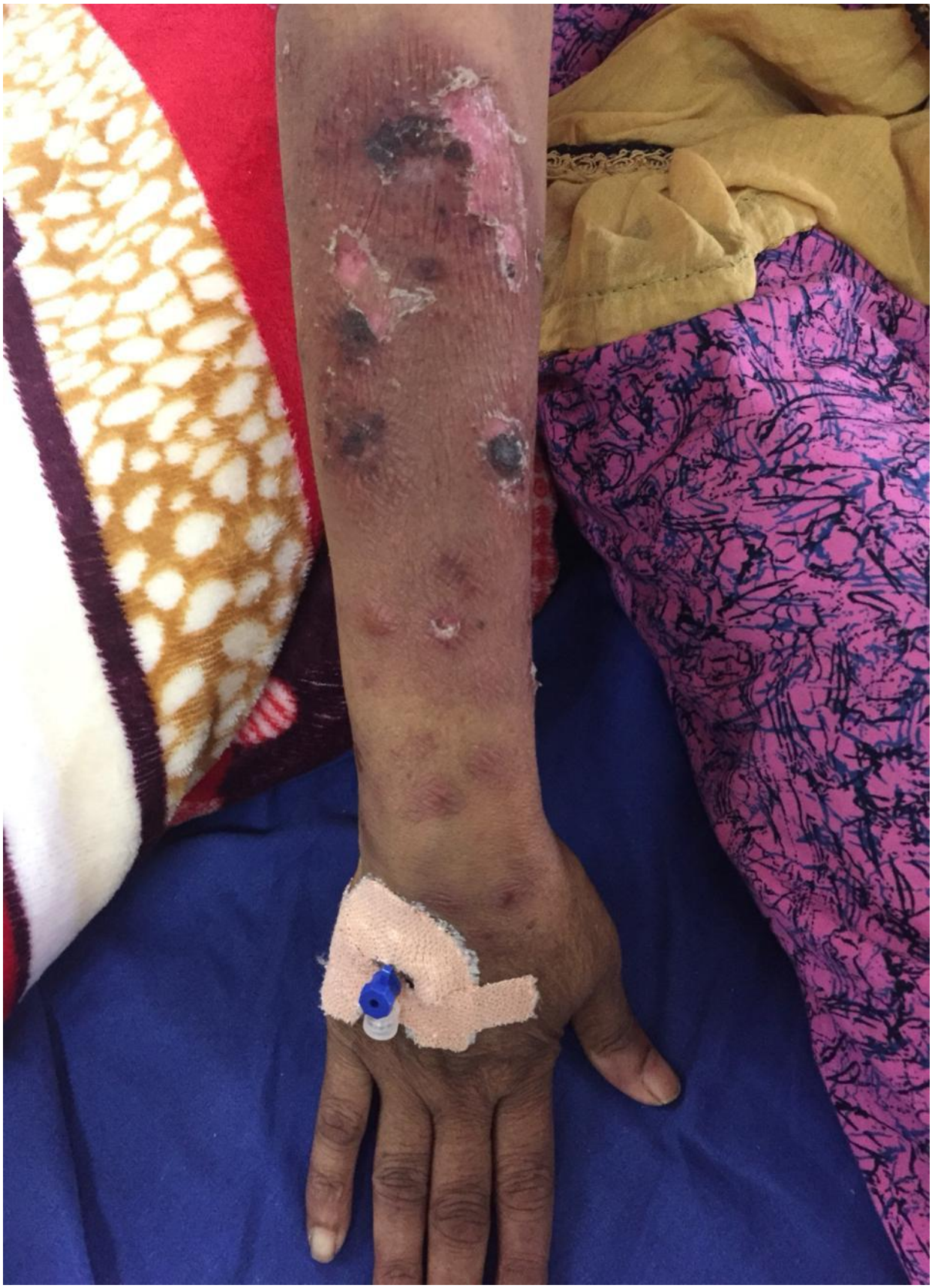
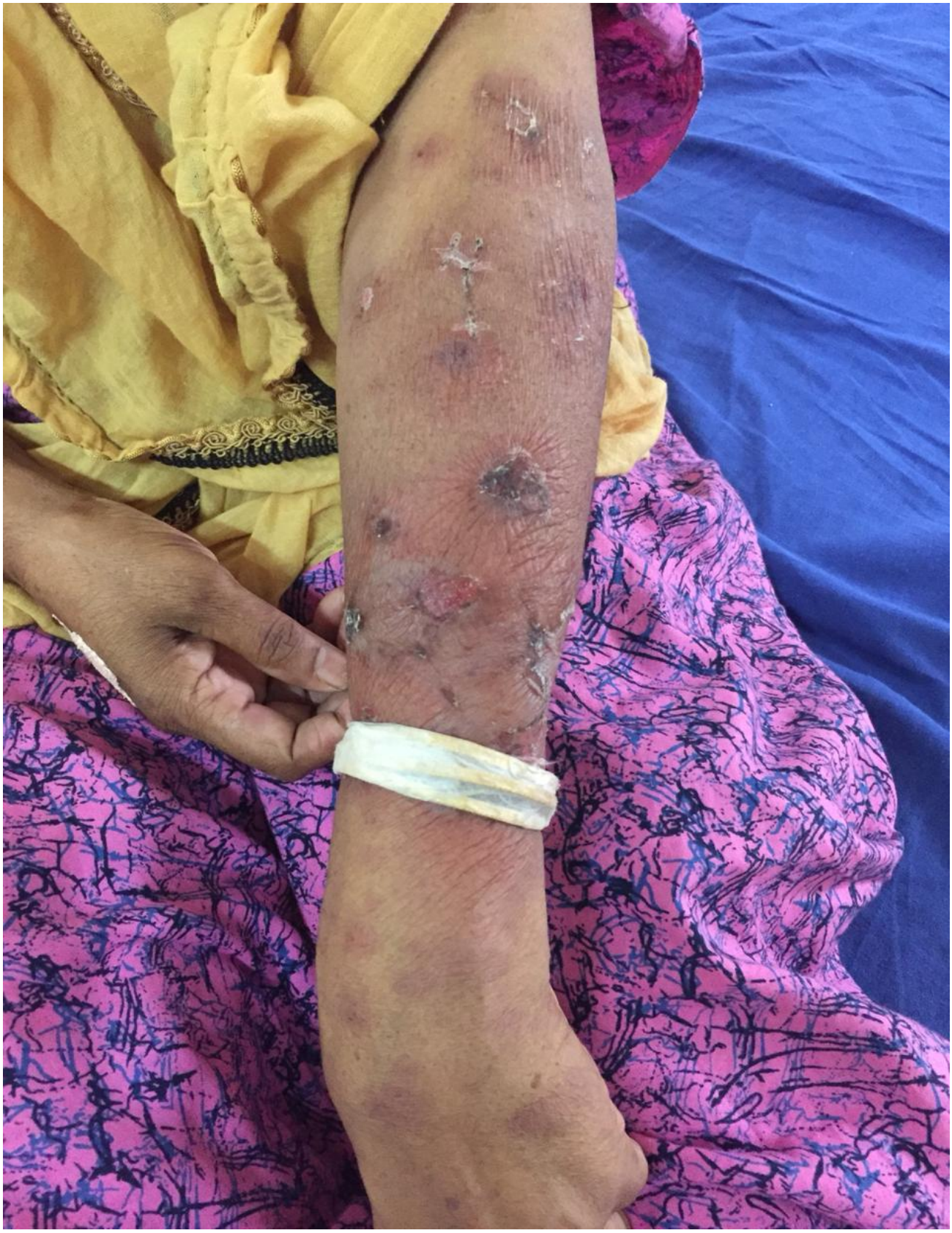


Figure no:1 Right upper limb

Figure no:2 Left upper limb



Rapid resolution of the bullous lesions was observed from systemic corticotherapy (I.V Dexamethasone 1 cc) (Figure 1 and Figure 2).

Our patient fulfilled all the four main criteria while the re-challenging option was not initiated due to ethical consideration. Most commonly reported DISS is with Granulocyte colony-stimulating factor. Several other drugs such as tretinoin[2], cotrimoxazole[5], diclofenac[4,5,6] and anti-cancer agents like bortezomib and azathioprine have shown to cause SS including paracetamol.

The biological mechanisms have remained elusive.[7]. Additional autoimmune features have been reported with SS. Patient's biopsy examination showed hyperkeratosis & keratotic plugging in epidermal region and congestion in subepidermal region. Melanophages, mixed leukocytic infiltrates along with dense neutrophilic infiltrates in the deeper serial section of subepidermal region. The histo-pathological changes explained in the literature review elucidates the presence of neutrophilic infiltrates with absence of vasculitis, linear deposition of immunoglobulins (IgA, IgG) at the basement membrane zone.[7] The cytokine mediated hypersensitivity followed by neutrophilic infiltration that are probably activated by (IL)-1.The factors such as dermal dendrocyte, immune complexes, HLA Serotypes, circulating autoantibodies as well as leukotactic-mechanisms contribute to the pathogenesis of SS.[2,7]..

The manifestations resolve without any therapeutic intervention in some patients with Idiopathic Sweet's syndrome, while in other forms of SS, the symptoms and cutaneous manifestations can persist without treatment for weeks to months.[7] And spontaneous improvement results from discontinuation of the associated medication in DISS. Patient's manifestations started to resolve with the withdrawal of causative drug (paracetamol).

Although there are no guidelines for the treatment of SS, the first line therapy for all variants is systemic corticosteroids. [8] Patient was treated with Inj. Dexamethasone at a dose of 2mg/day twice a day, which helped in the rapid resolution of the inflammatory lessons. In corticotherapy contraindicated cases, oral therapy with Potassium Iodide (900 mg/day) or Colchicine (0.5-1.5 mg/kg/day) can be initiated which results in rapid resolution of SS manifestations within the 72 hours of the start of the therapy.[8] Non complicated SS can be treated with oral prednisone with a dose of 0.5-1.5 mg/kg/day. Topical preparations of high potency steroids can be beneficial for localized lesions. Second line therapeutic agents include dapsone (100-200 mg/day), clofazimine (100-200 mg/day), indomethacin (50-150mg/day) and cyclosporin (2-4 mg/kg/day).[9,10]Refractory cases of Sweet's syndrome has been managed by IL-1 blocking agent, anakinra whose efficacy is proven and published in literature.[3]

CONCLUSION

Sweet Syndrome is a rare skin disease and its association with paracetamol is rarely reported. The number of DISS cases continues to rise in literature. Importance of this report lies in educating healthcare professionals alike on the newly recognized adverse event associated with acetaminophen - one of the popular and safest analgesic-antipyretic.

Consent: We obtained written informed consent from our patient for the publication of the case.

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