Case Report

Not so sweet; severe Sweet's syndrome presenting as SIRS and pleural effusion

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Abstract

Acute febrile neutrophilic dermatosis (Sweet's syndrome) is a rare inflammatory condition which presents as abrupt onset of painful erythematous plaques or nodules, often associated with fever and leucocytosis. Many extracutaneous manifestations are described in literature, but pulmonary manifestations and systemic inflammatory response syndrome (SIRS) are rare. Here we report a case of a 35-year-old male who presented with SIRS and pleural effusion. The presence of vesiculobullous and pustular skin lesions raised the suspicion of Sweet's syndrome and it was confirmed by skin biopsy. Initiation of systemic glucocorticoids lead to complete resolution of symptoms.

Keywords: Glucocorticoids, inflammatory disorder, neutrophilic dermatosis, vesiculobullous lesions

Introduction

Acute febrile neutrophilic dermatosis (Sweet's syndrome) is a rare inflammatory condition which presents as abrupt onset of painful erythematous plaques or nodules, often associated with fever and leucocytosis. Extracutaneous manifestations such as arthralgias, myalgias, and ocular involvement are well-described in Sweet's syndrome. However, pulmonary manifestations and systemic inflammatory response syndrome (SIRS) are rare. Here we report a case of a 35-year-old male with Sweet's syndrome who had many atypical clinical features including SIRS and pleural effusion.

Case Report

A 35-year-old male, software engineer presented to our hospital with complaints of fever of 11 days' duration, pustular lesions of the lower limb, and respiratory distress. The patient was in his usual state of health when the symptoms started as sore throat and high-grade fever 11 days before presentation. He also had significant myalgia. There was no history of cough or breathlessness. There was no history of any drug intake before the onset of symptoms. Three days after the onset of his illness, he was admitted to a local hospital. His total leucocyte count at presentation to the hospital was 15,800 cells/mm^3 (90% polymorphonuclear cells), and erythrocyte sedimentation rate (ESR) was 55 mm in the first hour. His bilirubin was raised (total 2.95 mg/dL, direct 2.02 mg/dL); the rest of the liver function tests and renal function test were normal [Table 1]. The patient was started on IV meropenem from the referring hospital. However, his fever persisted. Workup for fever including dengue NS1 antigen, serology and polymerase chain reaction for leptospirosis and scrub typhus, peripheral smear, and antigen testing for malarial parasites came negative. Transthoracic echocardiogram, skiagram of the chest, and contrast-enhanced computed tomography scan of

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thorax and abdomen were also normal. After 8 days of fever, the patient developed painful pustular lesions involving the left popliteal region [Figure 1] and serpigenous vesiculobullous lesions involving dorsum of both the feet [Figure 2]. Despite continuation of the antibiotics, his fever persisted, total leucocyte count continued to rise, and he developed shortness of breath and was referred to our hospital.

Table 1: Results of laboratory investigations

	On admission to the first hospital	At presentation in our hospital (8 days later)
Hematocrit (%)	35.2	31
Hemoglobin (g/dL)	11.6	10.6
White cell count (per mm^3)	15,800	35,400
Differential count	N90L4	N93L3
Platelet count	321,000	297,000
ESR (mm in first hour)	55	61
Blood urea (mg/dL)	28	38
Creatinine (mg/dL)	0.83	0.4
Sodium (mmol/L)	136	145
Potassium (mmol/L)	4.1	3.69
Bilirubin (mg/dL)		
Total	2.95	3.0
Direct	2.02	0.2
Alanine aminotransferase (U/L)	44	76
Aspartate aminotransferase (U/L)	40	56
C-reactive protein (mg/L)		33
Serum procalcitonin (ng/mL)		0.9
Serum lactate (mmol/L)		1.9
Arterial blood gases		
pO_2 (mmHg)		86
pCO_2 (mmHg)		33.4
pH		7.48
HCO_{3-} (mmol/L)		25.5
Pleural fluid analysis		
White cell count (per mm^3)		6000
Differential count		N80L20
Protein (g/dL)		4.5
Glucose (mg/dL)		60

ESR: Erythrocyte sedimentation rate

At presentation, the patient was looking toxic and ill. The temperature was 104°F, pulse rate was 166/min, and respiratory rate was 26/min. His blood pressure was maintained at 140/70 mm Hg and SpO_2 on room air was 91%. Examination of the cardiovascular and respiratory system as well as the abdomen was unremarkable. His skin lesions were persisting.

His blood investigations showed a total leucocyte count of 35,400 cells/mm^3 with 93% neutrophils. The ESR was 61 mm in the first hour, and C-reactive protein level was high at 33 mg/L, bilirubin was 3 mg/dL, and serum transaminases were mildly raised. The arterial blood gas analysis revealed primary respiratory alkalosis. His electrocardiography showed multiple atrial premature complexes.

He fulfilled all the criteria for SIRS, and a possibility of sepsis was considered with bullous and pustular lesions as possible source of infection. He was put on noninvasive ventilation, and after drawing blood for culture and scraping from the pustular skin lesions for culture, vancomycin was added and meropenem was continued.

Despite antibiotics, his fever continued and blood culture results were reported to be sterile and pus cultures grew few colonies of skin commensals. His breathlessness increased, and on chest examination 48 h after admission, there was reduced intensity of breath sounds on the right side. Chest X-ray revealed bilateral pleural effusion (more on the right side) which on aspiration showed abundant neutrophils and no microorganisms. The culture of the pleural fluid turned out to be sterile.

In view of the high fever, neutrophilic leucocytosis, typical skin lesions, and poor response to antibiotics, a possibility of Sweet's syndrome was considered. A skin biopsy was taken and the patient was started on oral prednisolone 60 mg PO

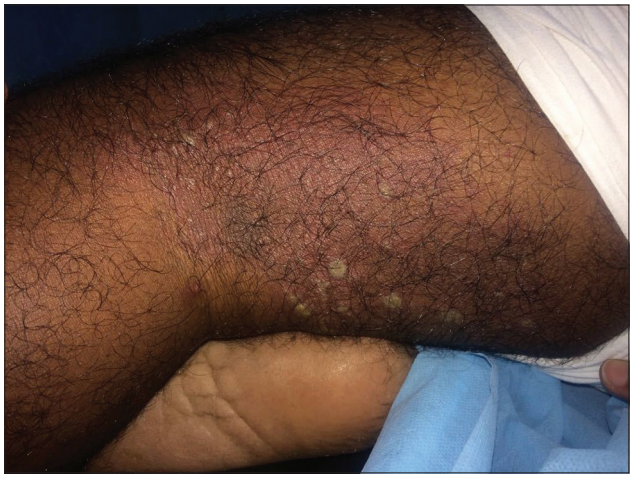

Figure 1: Painful pustular lesions in the left popliteal region

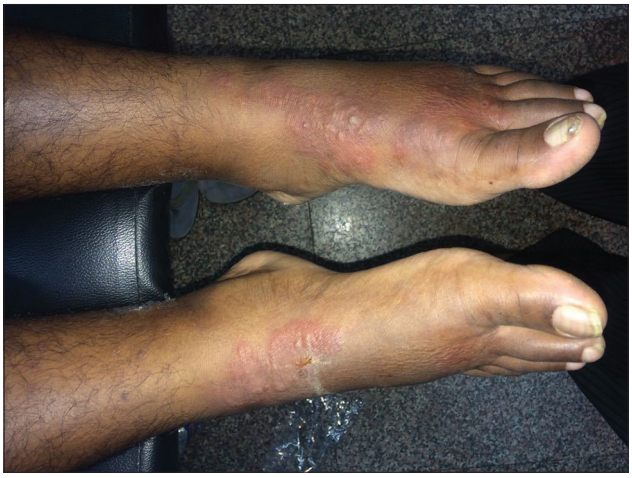

Figure 2: Serpigenous vesiculobullous lesions involving dorsum of both feet

OD after 11 days of his initial presentation. His condition dramatically improved, with fever, tachycardia and tachypnea subsiding within two days of starting corticosteroids. His total leucocyte count gradually returned back to normal after 5 days. Repeat chest X-ray after 3 days showed resolution of pleural effusion. The lesions healed without ulceration and with mild hyperpigmentation Histopathology of the skin biopsy showed neutrophilic dermatoses without any evidence of vasculitis suggestive of Sweet's syndrome. Antibiotics were stopped, and the patient was discharged on tapering dose of prednisolone. At 2 months follow-up, the patient remained afebrile, with no relapse of the skin lesions and normal blood counts.

Discussion

Acute febrile neutrophilic dermatosis is also known as Sweet's syndrome after the first classic description by Dr. Robert Douglas Sweet in 1964.[1] It is an uncommon inflammatory disorder characterized by the abruptly appearing painful, erythematous and indurated papules, plaques, or nodules on the skin with frequent company of fever, and leukocytosis.

Sweet's syndrome is traditionally classified into classical (or idiopathic), malignancy-associated, and drug-induced.[2] Classical or idiopathic Sweet's syndrome has been described mostly in young or middle-aged women with an antecedent history of upper respiratory tract infection or associated pregnancy or inflammatory bowel disease.[1,2]

For establishing the diagnosis of classical Sweet's syndrome, a set of criteria are suggested as follows, of which the presence of at least two major and two minor criteria are given below:[3]

Major criteria: (1) abrupt onset of painful erythematous plaques or nodules; (2) histopathologic evidence of a dense neutrophilic infiltrate without evidence of leukocytoclastic vasculitis.

Minor criteria: (1) pyrexia >38°C; (2) association with an underlying hematologic or visceral malignancy, inflammatory disease, or pregnancy, or preceded by an upper respiratory or gastrointestinal infection or vaccination; (3) excellent response to treatment with systemic corticosteroids or potassium iodide; (4) abnormal laboratory values at presentation (three of four): erythrocyte sedimentation rate >20 mm/h, positive CRP,>8000 leukocytes, and >70% neutrophils.

In our case, both the two major criteria and all the minor criteria were fulfilled. Unlike the classical descriptions, our patient was a middle-aged male and the lesions were in the inferior extremity. However, a large 18 years' retrospective study from the Mayo Clinic with 77 patients has reported male patients to dominate (56%) and lower extremity skin lesions to be present in 55% of cases. The same study has also reported lower association of malignancy, in case the patients were nonanemic.[4] The vesiculobullous lesions that we have encountered in our patient have been described in few cases in the literature[5] and in about 14% cases in the Mayo Clinic report.[4]

Sweet's syndrome has been associated with a multitude of extracutaneous manifestations. Arthralgias, myalgias, conjunctivitis, and uveitis are relatively common; however, it can present with uncommon features such as aseptic meningitis, myocarditis, hepatosplenomegaly, sterile osteomyelitis, or even pulmonary infiltrates and pleural effusion.[6] An extensive review of 34 cases on pulmonary involvement in Sweet's syndrome showed that skin involvement precedes pulmonary involvement, and bilateral or unilateral pulmonary infiltrates were the most common radiological feature.[7] Pleural effusion accompanying lung infiltrates was uncommon and was present only in seven cases. Bronchoalveolar lavage and lung biopsy may aid in the diagnosis but is not always essential.

SIRS is usually associated with sepsis and rarely described in Sweet's syndrome, with only a handful of case reports.[8,9] SIRS is a marker of poor prognosis as it can rapidly progress to multiorgan dysfunction. Systemic steroids should be initiated in such cases as soon as possible.

Primary care physicians must be aware of various noninfectious causes of SIRS. Severe Sweet's syndrome though rare should be considered as an important differential diagnosis of SIRS in the presence of characteristic skin lesions. A high index of suspicion can help in early diagnosis and early initiation of steroids leading to prompt clinical response.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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