Sweet syndrome with extracutaneous manifestation: a case report

Bhargavi Neela[1], Srinivas Pramod Junutula[1], Sheshagiri Sharvana Bhava Bandaru[2*]

European Journal of Medical Case Reports
Volume 5(1):22–25
https://doi.org/10.24911/ejmcr/173-1592581130

ABSTRACT

Background: Sweet syndrome (SS), also known as acute febrile neutrophilic dermatosis, is a rare dermatological disorder characterized by fever and the sudden onset of a rash, which consists of multiple tender, red or bluish-red bumps or lesions. These lesions usually occur on the arms, legs, trunk, face, or neck. In some cases, additional systems of the body can become involved.

Case Presentation: A 20-year-old male presented with chief complaints of fever, cough, and skin lesions. Laboratory investigations revealed an elevated erythrocyte sedimentation rate, C- reactive protein, neutrophils, and total leukocyte count. A skin biopsy was performed, and the patient was diagnosed with SS.

Conclusion: The patient was treated with oral corticosteroids and other medications to alleviate his symptoms. He was discharged on a tapering dose of prednisolone, and follow-up after a month showed that he remained afebrile with no relapse of skin lesions.

Keywords: Sweet syndrome, inflammatory disorder, neutrophilic dermatosis, erythrocyte sedimentation rate, corticosteroids.

Received: 19 June 2020 **Accepted:** 04 December 2020 **Type of Article:** CASE REPORT **Specialty:** Dermatology

Correspondence to: Sharvana Bhava Bandaru
*Department of Clinical Pharmacy & Pharm. D., Vaagdevi College of Pharmacy, Warangal, India.
Email: sharavanabhava6@gmail.com
Full list of author information is available at the end of the article.

Background

Sweet syndrome (SS), also known as acute febrile neutrophilic dermatosis, is a serious dermatological illness. Patients affected present with an abrupt onset of tender plaques or nodules and accompanying fever, arthralgia, ophthalmology manifestations, headaches, and in rare cases, oral or genital damage. SS has been associated with autoimmune processes, malignancies, infections, drug reactions, and gastrointestinal disorders, such as inflammatory bowel disease. Some authors have suggested that the pathophysiology of SS involves a dysregulation of immune function or immunomediated hypersensitivity [1].

The diagnosis of SS is confirmed by biopsy. The findings are significant for neutrophilic dermatosis with epidermal/dermal junction interface involvement. However, pulmonary manifestations are rare. Here we report a case of a 20-year-old male with SS who had an atypical clinical feature of pleural effusion.

Case Presentation

A 20-year-old male presented to the hospital with complaints of fever of 5 days duration and cough, followed by erythematous plaques on the forehead and neck. The patient was in his usual state of health when symptoms started as sore throat and high-grade fever. There was no history of drug intake before the onset of skin lesions and no history suggesting systemic involvement. After 5 days of fever, the patient developed painful erythematous plaques with elevated borders, involving the head and neck region, which were hyperpigmented at the time of admission (Figure 1). On examination his lesions were solid to the touch.

At presentation, the patient was conscious and coherent. The temperature was 102°F, pulse rate was 152/minute, and respiratory rate was 24/minute. His blood pressure was 130/70 mmHg. Examination of the cardiovascular system as well as the abdomen was normal. On chest examination, there were reduced breath sounds on the right side. His skin lesions were persisting.

His blood investigations showed a total leukocyte count of 14,400 cells/mm^3, with 80% neutrophils. The

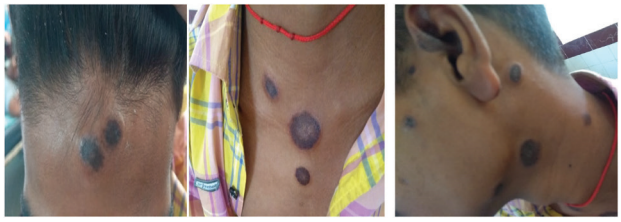

Figure 1. *Multiple hyperpigmented plaques with elevated borders distributed over forehead and neck region.*

Table 1. *Results of laboratory investigations.*

HEMOGLOBIN (G/DL)	12
White cell count (per mm^3)	14,400
Differential count	
Neutrophils	80%
Lymphocytes	48%
Eosinophils	2%
Monocytes	5%
Platelet count (mm^3)	3,90,000
ESR (mm in first hour)	60
Random blood sugar (mg/dl)	62
Blood urea (mg/dl)	10
Creatinine (mg/dl)	0.7
Sodium (mmol/l)	137
Potassium (mmol/l)	3.8
Chlorine (mmol/l)	96
Total Bilirubin (mg/dl)	0.71
Alanine aminotransferase (U/l)	10
Aspartate aminotransferase (U/l)	32
Total serum proteins (g/dl)	7.2
CRP (mg/l)	36

erythrocyte sedimentation rate (ESR) was 60 mm in the first hour, and C-reactive protein (CRP) level was high at 36 mg/l. The patient's renal and hepatic profiles were normal (Table 1). Chest X-ray revealed bilateral pleural effusion (more on the right side; R>L) and right middle lower lobe consolidation. He also tested positive for antistreptolysin "o" test which is indicative of a recent strep infection. The patient was started on IV antibiotics [amoxicillin/potassium clavulanate 1.2 g/bis die (two times a day) (BD)]. Despite the continuation of antibiotics, his fever persisted. A possibility of SS was considered, in view of the neutrophilic leukocytosis and typical skin lesions.

A skin biopsy was taken and the histopathologic sections revealed keratinized stratified squamous epithelium; underlying dermis showed edematous with acute inflammatory cells infiltrating the karyorrhexis of neutrophils, perivascular and periadenexal neutrophil infiltrations. The patient was finally diagnosed with classical/idiopathic SS. The patient was started on oral prednisolone 60 mg/omne die (once daily) (OD) after 5 days of his initial presentation. His other medications included Inj. amoxicillin/potassium clavulanate 1.2 g/BD, tablet acetaminophen 500 mg/ter in die (three times a day), tablet azithromycin 500 mg/OD, and nebulization with salbutamol and budesonide to alleviate his symptoms. His condition improved with the corticosteroid's treatment. His leukocyte count reduced to normal and pleural effusion was resolved. Antibiotics were stopped and the patient was discharged on a tapering dose of prednisolone. The patient was followed-up after a month, and he remained afebrile with no relapse of skin lesions.

Discussion

Acute febrile neutrophil dermatosis is also known as SS, following Dr. Robert Douglas' first classical explanation and description in 1964 [2]. This is a rare inflammatory condition characterized by the abruptly painful, erythematous appearance and indurated papules, plaques, or nodules on the skin with frequent fever and leukocytosis.

SS is traditionally classified as classical (or idiopathic), malignancy associated, and drug-induced [3]. The disease is most frequent in women between the age of 30 and 60 years [4], with an antecedent history of upper respiratory tract infection or associated pregnancy or inflammatory bowel disease [2,3]. For establishing the diagnosis of classical SS, a set of criteria is suggested below, of which the presence of both major criteria and two of the four minor criteria is required [5].

Major criteria

1. Abrupt onset of painful erythematous plaques or nodules.
2. Histopathology 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): ESR >20 mm/hours, positive CRP, >8,000 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 young male, and the lesions were on the forehead and neck region. However, a large 18-year retrospective study from the Mayo Clinic with 77 patients has reported that male patients to dominate (56%), and head and neck skin lesions were present in 29% and 25% of the cases, respectively. The same study also reported a lower association of malignancy in case the patients were non-anemic [6]. In another retrospective study conducted over a 20-year period included 90 cases from a tertiary care center included 75 women (three were pregnant) and 15 men and the mean age of patients at the time of diagnosis was 46.5 years [7].

Although upper respiratory infection is the most common presentation preceding SS skin lesions, it is important to not discard other types of viral, bacterial, and even fungal infections. Other infectious diseases recently thought to be associated with SS include chlamydophila,

pneumoniae, osteomyelitis, sporotrichosis, and leptospirosis; however, the association of these conditions with SS must be confirmed [8].

The skin lesions (plaques) that we have encountered in our patient have been described in about 51% of the patients in the Mayo Clinic study [6]. The histopathologic report of our patient was like the retrospective study conducted by Amouri et al. [7], in which histological examination of all patients were found to exhibit the characteristic features of SS, which include diffuse dermal nodular and perivascular neutrophilic infiltrate with varying degrees of edema.

SS has been related to many extracutaneous manifestations. Myalgias, arthralgias, conjunctivitis, and uveitis are relatively common; however, it can present with uncommon features like myocarditis, aseptic meningitis, hepatosplenomegaly, sterile osteomyelitis; may be even pulmonary infiltrates and pleural effusion [9]. An in-depth review of 34 cases on pulmonary involvement in SS showed that skin involvement precedes pulmonary involvement, and bilateral or unilateral pulmonary infiltrates were the foremost common radiological feature [10]. Pleural effusion in SS that we have encountered in our patient was also present during a case reported by Vettakkara et al. [11].

Conclusion

After promptly confirming the diagnosis of SS, systemic steroids should be initiated as soon as possible. It is a distinctive disorder with some clinical and histological characteristics that can be associated with a variety of systemic disorders and medications. Physicians who are involved in the diagnosis and care of SS patients should consider the broad range of possible underlying conditions related to this skin disease, and thus should always explore the likelihood of association with a neoplastic disorder or a drug.

What is new?

The patient was diagnosed with a case of idiopathic SS, which is a rarity in male patients, who usually present with drug-induced or paraneoplastic variant of SS and he was presented with extracutaneous manifestations with pulmonary involvement, which is uncommon.

List of Abbreviations

CRP	C-reactive protein
ESR	Erythrocyte sedimentation rate
SS	Sweet syndrome

Funding

None.

Conflict of interests

The authors declare that there is no conflict of interests regarding the publication of this case report.

Consent to participate

Written informed consent was taken from the patient.

Author details

Bhargavi Neela[1], Srinivas Pramod Junutula[1], Sheshagiri Sharvana Bhava Bandaru[2]

1. Student, Doctor of Pharmacy (Pharm.D), Department of Clinical Pharmacy & Pharm.D, Vaagdevi College of Pharmacy, Warangal, India
2. Associate Professor, Department of Clinical Pharmacy & Pharm.D., Vaagdevi College of Pharmacy, Warangal, India

References

1. Corazza M, Lauriola MM, Borghi A, Marzola A, Virgili A. Sweet's syndrome: a retrospective clinical, histopathological and immunohistochemical analysis of 11 cases. Acta Derm Venereol. 2008;88(6):601–6.
2. Sweet RD. An acute febrile neutrophilic dermatosis. Br J Dermatol. 1964;76:349–56. https://doi.org/10.1111/j.1365-2133.1964.tb14541.x
3. Cohen PR. Sweet's syndrome-a comprehensive review of an acute febrile neutrophilic dermatosis. Orphanet J Rare Dis. 2007;2:34. https://doi.org/10.1186/1750-1172-2-34
4. Borges da Costa J, Silva R, Soares de Almeida L, Filipe P, Marques Gomes M. Sweet's syndrome: a retrospective study of 42 admitted patients in a Portuguese hospital. Int J Dermatol. 2009;48(9):953–55. https://doi.org/10.1111/j.1365-4632.2009.04098.x
5. Walker DC, Cohen PR. Trimethoprim-sulfamethoxazole-associated acute febrile neutrophilic dermatosis: case report and review of drug-induced Sweet's syndrome. J Am Acad Dermatol. 1996;34(5 Pt 2):918–23. https://doi.org/10.1016/S0190-9622(96)90080-8
6. Rochet NM, Chavan RN, Cappel MA, Wada DA, Gibson LE. Sweet syndrome: clinical presentation, associations, and response to treatment in 77 patients. J Am Acad Dermatol. 2013;69(4):557–64. https://doi.org/10.1016/j.jaad.2013.06.023
7. Amouri M, Masmoudi A, Ammar M, Boudaya S, Khabir A, Boudawara T, et al. Sweet's syndrome: a retrospective study of 90 cases from a tertiary care center. Int J Dermatol. 2016;55(9):1033–39. https://doi.org/10.1111/ijd.13232
8. Anzalone CL, Cohen PR. Acute febrile neutrophilic dermatosis (Sweet's syndrome). Curr Opin Hematol. 2013;20(1):26–35. https://doi.org/10.1097/MOH.0b013e32835ad132
9. Cohen PR, Hongsmann H, Kurzrock R. Acute febrile neutrophilic dermatosis (Sweet syndrome). In: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K, editors. Fitzpatrick's dermatology in general medicine. 8th ed. New York, NY, McGraw Hill; 2012. vol. 1, p 362.
10. Fernandez-Bussy S, Labarca G, Cabello F, Cabello H, Folch E, Majid A. Sweet's syndrome with pulmonary involvement: case report and literature review. Respir Med Case Rep. 2012;6:16–9. https://doi.org/10.1016/j.rmcr.2012.08.004
11. Vettakkara KMN, Banerjee S, Mittal A, Goel P, Kumar P, Baitha U et al. Not so sweet; severe Sweet's syndrome presenting as SIRS and pleural effusion. J Family Med Prim Care. 2018;7(6):1584–87. https://doi.org/10.4103/jfmpc.jfmpc_289_18

Summary of the case

1	**Patient (gender, age)**	Male, 20-year-old
2	**Final diagnosis**	SS with right lung consolidation with right-sided pleural effusion.
3	**Symptoms**	Fever, cough, skin lesions.
4	**Medications**	Oral symptomatic medications were prescribed.
5	**Clinical procedure**	A skin biopsy was done.
6	**Specialty**	Dermatology