Case report

Sweet Syndrome secondary to infection: A case report

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Abstract

We present a case of a patient with Sweet Syndrome secondary to possible bacterial infection. The diagnosis of Sweet Syndrome was made based on his clinical and histopathological findings. Sweet syndrome is a rare inflammatory disorder characterised by tender erythematous skin lesions, often accompanied with fever, neutrophilia and leukocytosis. It is generally classified into classical (or idiopathic), drug-induced or malignancy-associated, each of which has its own features.

Keywords: Sweet Syndrome; acute febrile neutrophilicdermatosis; corticosteroids

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Introduction

Sweet syndrome (SS) is a rare inflammatory disorder characterised by tender erythematous skin lesions, often accompanied with fever, neutrophilia and leukocytosis.[1] Involvement of the extracutaneous organs may occur. SS is generally classified into classical (or idiopathic), drug-induced or malignancy-associated, each of which has its own features.[1] The pathogenesis of SS can be multifactorial and remains inconclusive. Corticosteroids either oral or topical play an important role in therapeutic options for SS which gives a quick improvement of both the skin lesions and other related symptoms.[1]

Case Report

A 62-year-old man with no underlying medical illness presented with skin rashes and tenderness mainly on his bilateral forearm, aggravated by sweating for a period of two months. He denied fever or joint pain. He had seen general practitioners and was given topical and oral medications with temporary relief. He had no medical problems and not on any medication. There was no family history of malignancy. He worked at a wood factory and smokes 5 cigarettes per day for more than 30 years.

On examination, he was alert, afebrile and comfortable. No fingers clubbing noted. There was presence of multiple tender erythematous non-pustular nodules over his bilateral arms (Picture 1). There was right anterior cervical solitary lymph nodemeasuring 1x1 cm and no axillary lymphadenopathy. There was no hepatosplenomegaly. Other physical examinations including per rectal examination revealed no abnormalities.

His blood investigations showed mild transaminitis with ALT 18 U/L and AST 25 U/L. His renal function test and glucose level were normal. His full blood count (FBC) showed WBC 9.05 x 10^9/L, Hb 13g/L, PLT 309 x 10^9/L with high neutrophil count 6.9 x

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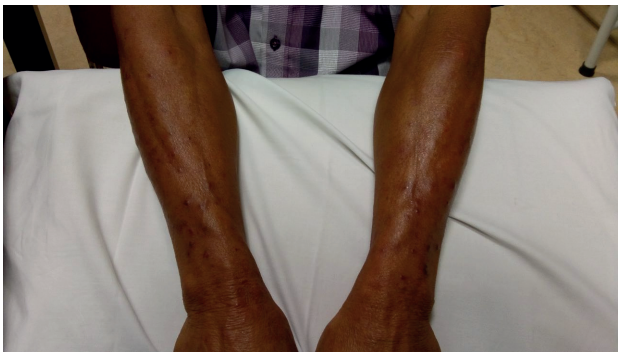
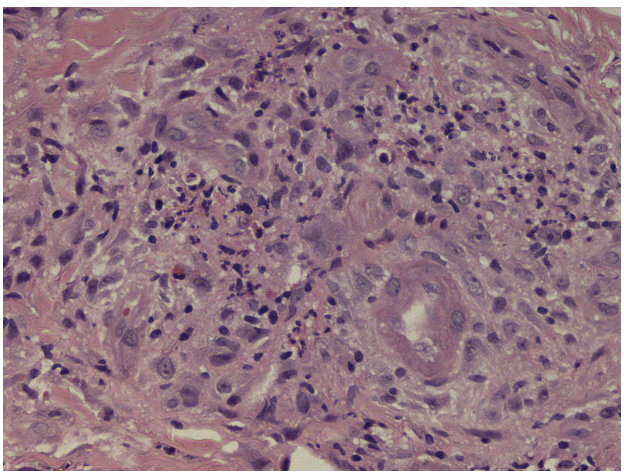


Picture 1: Multiple erythematous non-pustular nodules over the bilateral arms.

10^9/L.His swab culture and sensitivity grew *Staph aureus* sensitive to erythromycin.His full blood picture (FBP) revealed no significant hematological abnormality and his autoimmune screening test using anti-neutrophil antibody (ANA) was negative. The histopathological examination (HPE) reported perivascular infiltration of the dermis by lymphocytes and histiocytes with many cell debris and neutrophils are also seen. The inflammation extends into the dermis however no subepidermal fatty tissue included.There was no frank granuloma encountered andPAS/GMS stain for fungi were negative." The mycobacterium tuberculosis culture and sensitivity was negative. The skin biopsy features neutrophilic dermatosis consistent with SS (Picture 2).



Picture 2: The dermis exhibit perivascular infiltration by lymphocytes, histiocytes and neutrophils with many cell debris seen (x 40 microscopic magnification)

Adiagnosis of SS secondary to possible bacterial infection was made and he was treated with oral Prednisolone 25mg once daily and oral erythromycin succinate 400mg twice daily for one week. He was also investigated to look for other primary causes of this problem by undergoing chest x-ray and taking blood investigations including screening for malignancy, which were all normal.

After afew weeks of post-treatment, the patient reported complete resolution of his skin reactions and other related symptoms. He will be reviewed again in a couple of months under dermatology department for follow-up.

Discussion

SS was firstly described as "acute febrile neutrophilicdermatosis" by Dr. Robert Douglas Sweet who recognised the association of an acute inflammatory skin eruption with fever and leukocytosis in eight female patientsin 1964.[2]

An abrupt onset of the painful inflammatory papules, plaques and nodules are typical appearances of SS.[1] Significant superficial dermal edema may lead to a pseudovesicular quality and some pustules may also present. The distribution of the cutaneous eruption is often asymmetrical, commonly appears over the upper extremities and may develop on the head, neck, trunk and lower extremities as well.[1,3] However, these patient's skin lesions were symmetrically distributed to involve both of his arms and it was associated with tenderness.[3]Arthralgias, headache, malaise and myalgias are additional symptoms that often occur in SS.[1]

SS is associated with a wide range of disorders and mainly divided into three subtypes that are classical, drug-induced and malignancy-associated.[1] The classical or idiopathic SS comprises the common cases of SS and may occur in many kinds of medical conditions. The most frequently encountered conditions associated with classical SS are infections, inflammatory bowel diseases (Crohn's disease and ulcerative colitis) and pregnancy.[4] Infection is one of the most important disorders not to be missed in the patient with clinical and/or histological findings suggestive of SS. The upper respiratory tract and gastrointestinal infections are the commonly associated infections.[4] In this particular patient the source of infection was unable to be determined however the predominant neutrophilia count in the FBC suggest that infection do play a role in this case. On the other hand, it is important to highlight that fever, leukocytosis and cutaneous lesions characterised by neutrophil-dense infiltrates may occur in bacterial sepsis as in SS.[1]

The diagnosis of SS is made based upon the consistent clinical and laboratory features along with the exclusion of disorders that may have similar

clinical findings. A quick response to corticosteroid therapy also supports the diagnosis. The diagnostic criteria for classical SS were initially proposed in 1986 by Su and Liu[5] which were then modified in 1994 by von den Driesch.[6]The current diagnostic criterianeed both major criteria (1 and 2) and two of four minor criteria (3 to 6) to diagnose a classical or malignancy-associated SS[7].For drug-induced SS, a separate set of criteria was proposed[7].

Table 1: Diagnostic criteria for classical SS versus drug-induced SS.[7]

Classical	Drug-induced
1. Abrupt onset of painful erythematous plaques or nodules	A. Abrupt onset of painful erythematous plaques or nodules
2. Histopathologic evidence of a dense neutrophilic infiltrate without evidence of leukocytoclastic vasculitis	B. Histopathologic evidence of a dense neutrophilic infiltrate without evidence of leukocytoclasticvasculitis
3. Pyrexia >38°C	C. Pyrexia >38°C
4. Association with underlying hematologic or visceral malignancy, inflammatory disease or pregnancy, OR preceded by upper respiratory infection, gastrointestinal infection, or vaccination	D. Temporal relationship between drug ingestion and clinical presentation, or temporally-related recurrence after oral challenge
5. Excellent response to treatment with systemic glucocorticoids or potassium iodide	E. Temporally-related resolution of lesions after drug withdrawal or treatment with systemic corticosteroids
6. Abnormal laboratory values at presentation (three of four of the following: erythrocyte sedimentation rate >20 mm/hour, positive C-reactive protein, >8000 leukocytes, >70% neutrophils)	

*The presence of both major criteria (1 and 2), and two of the four minor classical SS. ** All five criteria (A, B, C, D, and E) are required for the diagnosis of drug-induced SS.

The detection of consistent histological findings is a major diagnostic criterion for SS. The common histologic characteristics of SS include prominent oedema in the superficial dermis, dense neutrophilic infiltration in the middle and upper dermis with sparing of the epidermis, endothelial swelling, leukocytoclasis and absence of vasculitis.[6] Laboratory evaluation is useful for identifying findings consistent with SS and signs of associated diseases or extracutaneous involvement. Anaemia and platelet abnormalities are commonly detected in SS related to drugs or malignancy, but are infrequently found in classical SS cases.[1]Abnormal urinalysis or complete metabolic panel may indicate kidney or liver involvement[1]

Pertaining to the patient's case, he had an abrupt onset of painful erythematous nodular skin lesions, histological evidence of neutrophilic infiltrate without evidence of leukocytoclastic vasculitis, responded well to systemic glucocorticoids and elevated erythrocyte sedimentation rate (ESR). Also, as he was not on any medication prior to the onset of the skin lesions, these all fit into the classical SS.

The treatment of SS is tailored individually and mainly targeting the specific symptoms that are prominent in the affected individuals. Oral corticosteroids are the mainstay of treatment and have proven effective in most cases by eradicatingthe symptoms.[1] The same positive response was seen in our patient. Topical or intra-lesional corticosteroids injections may be used for localised lesions. Several drugs have been used to treat patients with SS which include potassium iodide, colchicine, indomethacin, dapsone, cyclosporine and clofazimine can be tried as alternative options.[1] These drugs are commonly used for patients who cannot tolerate corticosteroids or if corticosteroids were found ineffective. With timely diagnosis and proper treatment, the cutaneous lesions of SS can resolve without scarring and major complications.

Conclusion

This case report illustrated a rare acute febrile neutrophilicdermatosis which presents with symptoms and signs that are commonly encountered and may mimic many acute inflammatory skin disorders. It is very important for health professionals to recognize this condition as it can cause significant negative effect on patients quality of life unless properly treated.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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