International Journal of Research in Dermatology
Lakamsani NP et al. Int J Res Dermatol. 2022 Mar;8(2):xxx-xxx
http://www.ijord.com

DOI: https://dx.doi.org/10.18203/issn.2455-4529.IntJResDermatol20220501

Case Report

An unusual presentation of Gomm-Button disease: a diagnostic enigma

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Received: 30 December 2021
Accepted: 02 February 2022

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ABSTRACT

Gomm-Button disease (Sweet syndrome) typically presents with abrupt onset of tender skin lesions accompanied by fever and neutrophilia. Histopathology shows the presence of dense neutrophilic infiltrates, oedema without vasculitis. One of the criteria for diagnosis of sweets syndrome is the absence of vasculitis. However, recent reports suggest that vasculitis should not exclude the diagnosis. We present a case of Sweet syndrome with an atypical clinical and histopathological presentation.

Keywords: Sweet syndrome, Tender subcutaneous and erythematous plaques, Neutrophilic infiltrate in the upper dermis, Leukocytoclastic vasculitis

INTRODUCTION

Dr. Robert Douglas Sweet originally described the Sweet syndrome in 1964.[1] It is an acute febrile neutrophilic dermatosis characterized by fever, neutrophilia and cutaneous lesions. Histologically, the disease typically shows an upper dermal infiltrate of mature neutrophils, with rapid improvement after the initiation of systemic corticosteroids.[2] The exact etiology of Sweet syndrome is not known. However, in many cases, the disease is preceded by viral infections of the upper respiratory tract, hematologic and visceral malignancies, autoimmune diseases, inflammatory bowel disease, pregnancy and medications.[3-5] Su and Liu established diagnostic criteria for Sweet syndrome, and later, Von den Driesch modified it by including abnormal laboratory values.[5,6]

CASE REPORT

A 60-year-old male patient presented with a sudden onset of asymptomatic red elevated lesions over the upper back of six days duration and painful, fluid-filled lesions over the right lower limb since two days. There was no similar history in the past, constitutional symptoms, or drug usage before the onset of lesions. There was no history of contact with cattle or exposure to arthropods. No history of comorbidities.

Cutaneous examination revealed four well-defined, erythematous, edematous, non-tender plaques with the peau-d'-orange appearance of sizes ranging from 0.5×1 cm^2 to 2×4 cm^2 over the upper back, tender bullae on an erythematous base of size 2×2 cm^2 over the dorsum of the right foot was noted (Figure 1).

Sensations over the plaques were normal. Blood investigations revealed neutrophilia, elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). Anti-nuclear antibodies (ANA) profile, anti-nuclear antibodies (ANCA) and slit skin smear for acid-fast bacillus (AFB) were negative.

Two skin biopsies were taken from the upper back and right lower limb and sent for histopathology examination. Multiple sections studied from both skin biopsy tissues shows intense neutrophilic dermal infiltrate and marked

papillary oedema in the reticular dermis, perivascular and peri-adnexal inflammation and vasculitis with plump endothelial cells with fibrinoid necrosis (Figures 3a-e).

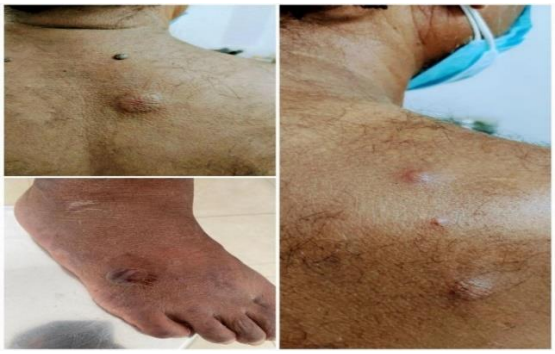

Figure 1: Well-defined, erythematous, edematous plaques with peau-d'-orange appearance over upper back; bullae on the erythematous base over dorsum of right foot.

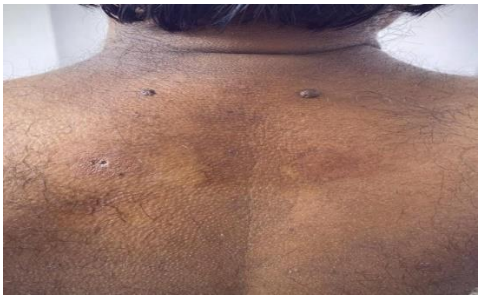

Figure 2: clinical improvement after initiation systemic corticosteroid therapy.

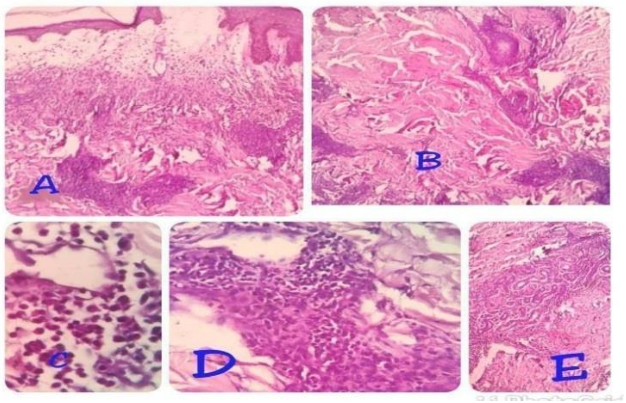

Figure 3: (a) H and E photomicrograph showing dense neutrophilic infiltrate and oedema in the reticular dermis; (b) H and E photomicrograph showing perivascular and peri adnexal (pilosebaceous unit) inflammation; (c) H and E photomicrograph showing perivascular inflammation comprising neutrophils and eosinophils; (d) H and E photomicrograph showing vasculitis with plump endothelial cells with fibrinoid necrosis; (e) H and E photomicrograph showing acute inflammation around peri adnexal glands.

Based on both clinical and histopathological examination, a diagnosis of sweet syndrome was made. The patient started on oral prednisolone and rapid clinical improvement within three days after initiation of systemic corticosteroid therapy (Figure 2).

DISCUSSION

Sweet syndrome can present heterogeneously, and the diagnosis can be challenging. A biopsy shall always be necessary to confirm the diagnosis. However, patients may usually present with abrupt onset of tender plaques or nodules, fever, arthralgias, ophthalmologic manifestations, headaches, and, rarely, oral or genital lesions. Pustular, vesicular, bullous, and targetoid lesions can occur rarely. Lesions are most commonly seen on the upper extremities and can also be present on the face, neck, chest, back, and lower extremities. Oral or genital lesions are rare. The role of dermatologists and pathologists are crucial in confirming the diagnosis. The widely used diagnostic criteria proposed by von den Driesch needs to fulfil two major and two minor criteria.[6]

Diagnostic criteria for sweet syndrome[7]

Major criteria

It includes: patients with sudden onset of tender erythematous plaques or nodules; and dense neutrophilic infiltrate on biopsy.

Minor criteria

It includes: patients with fever, temperature >38 ^{o}C association with an underlying hematologic malignancy, inflammatory disease, or pregnancy or preceded by an upper respiratory or gastrointestinal tract infection; excellent response to treatment with systemic corticosteroids; abnormal laboratory values at presentation (3 of 4); erythrocyte sedimentation rate >20 mm/h; positive CRP; leukocyte count >10×10^{3}/µl; and >70% neutrophils.

In most cases of Sweet syndrome, the absence of leukocytoclastic vasculitis is considered a criterion. However, studies have shown that leukocytoclastic vasculitis can be seen concurrently in sweet syndrome, and that it represents an epiphenomenon rather than a primary immune complex-mediated disease (i.e. vasculitis resulting from vascular transmigration of inflammatory cells). Slone et al showed and concluded that vasculitis is not a primary immune-mediated process in Sweet syndrome but can occur secondary to toxic products released from neutrophils.[8] A common pathway for vessel wall damage is the release of toxic metabolites and proteases from activated neutrophils. The longer the exposure to the toxic substance, the more damage to the vessel wall. Pathogenic factors other than the duration of lesions are at play since studies done by Malone et al showed not all lesions of long duration contained damaged

vessel walls, but one of the lesions of short duration (<7 days) also showed evidence of vasculitis.[8] We concur with the theory proposed by Jordaan and championed by others that vasculitis in Sweet syndrome is not a result of immune complex-mediated injury but represents secondary vessel wall damage due to toxic metabolites released by activated neutrophils.[9,10]

CONCLUSION

We report this case for its varied presentation, i.e. presence of asymptomatic edematous plaques over upper back and tender bullae over lower limbs and histologically presence of vasculitis with peri-vascular dense neutrophilic infiltrate. Nevertheless, the presence of vasculitis does not rule out Sweet syndrome.

ACKNOWLEDGEMENTS

Authors would like to thank department of dermatology and venereology and department of pathology for providing all the facilities to carry out this work.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Sweet RD. An acute febrile neutrophilic dermatosis. Br J Dermatol. 1964;76:34956.
2. Cohen PR, Kurzrock R. Sweet's syndrome and cancer. Clin Dermatol. 1993;11:14957.
3. Liu CI, Hsiao CH, Wu JT, Tsai TF. Sweet syndrome with histiocytoid infiltrates and neutropenia: a rare combination. J Am Acad Dermatol. 2009;61:882-4.
4. Cooper PH, Innes DJ Jr, Greer KE. Acute febrile neutrophilic dermatosis (Sweet's syndrome) and myeloproliferative disorders. Cancer. 1983;51:1518-26.
5. Su WP, Liu HN. Diagnostic criteria for Sweet's syndrome. Cutis. 1986;37:167-74.
6. Von den Driesch P. Sweet's syndrome (acute febrile neutrophilic dermatosis) J Am Acad Dermatol. 1994;31:535-56.
7. Honigsmann H, Cohen PR, Wolff K. Acute febrile neutrophilic dermatosis (Sweet's syndrome). In: Freedberg IM, Eisen AZ, Wolff K, Austen KF, Goldsmith LA, Katz SI, Fitzpatrick TB, editors. Fitzpatrick's Dermatology in General Medicine. 5th edition. New York: McGraw-Hill Health Professions Division. 1999;1117-23.
8. Malone JC, Slone SP. Vascular inflammation (vasculitis) in Sweet syndrome, A clinicopathological study of 28 biopsy specimens from 21 patients. Arch Dermatol. 2002;138:345-9.
9. Jordaan HF. Acute febrile neutrophilic dermatosis: a histopathological study of 37 patients and a literature review. Am J Dermatopathol. 1989;1199-211.
10. Von den Driesch P. Sweet's syndrome and vasculitis [letter]. J Am Acad Dermatol. 1996;34539.

Cite this article as: Lakamsani P, Sridevi K, Ramamurthy DVSB, Senthil AL, Salecha AJ. An unusual presentation of Gomm-Button disease: a diagnostic enigma. Int J Res Dermatol 2022;8:282-4.