CASE REPORT

Sweet's syndrome.

Ma. Luisa Silva Mora,* Enrique Úraga, Juan Carlos Garcés*****

* *Dermatologist, of the Dermatological Center "Dr. Úraga" Guayaquil - Ecuador*
** *Director of the Dermatological Center "Dr. Úraga"*
*** *Pathologist - Dermatopathologist, Guayaquil - Ecuador*

Correspondence: malusilm16@gmail.com

Key words:

Reception date: 19/12/2022
Acceptance date: 27/03/2023

ABSTRACT

Sweet's syndrome is a rare dermatosis, with established clinical, histopathological and laboratory criteria for its diagnosis. The classic form is the most frequent, being predominant in women. Classically painful lesions occur on the extremities, face and neck as papules, plaques, erythematous-edematous nodules. The first line of treatment is high doses of systemic corticosteroids. We present the case of a patient with systemic symptoms and lesions of classic localization.

INTRODUCTION

Acute febrile neutrophilic dermatosis was first described by Robert Douglas Sweet in 1964, in eight women between 32 and 55 years of age who presented fever, neutrophilia, painful plaques on extremities, face and neck, accompanied by neutrophilic dermal infiltrate.[1] It eventually became the eponym of the disease also known as Sweet's Syndrome.[2]

It typically manifests as painful erythematous plaques and nodules on the extremities, trunk and face. Different clinical variants have been reported such as blistering, subcutaneous, cellulitis-like, necrotizing,[3] localized on the dorsum of the hands and generalized pustular.[2]

Although the etiology is unknown, mainly idiopathic, triggering factors have been associated with infections, neoplasms, some medications, as well as its development following certain vaccines.[4,5]

Su and Liu proposed in 1986 the diagnostic criteria for the classic form, which were later modified in 1994 by von den Driesch, requiring that the two major and two of the four minor criteria be met.[2]

We present the case of a 31-year-old female patient with lesions suggestive of Sweet's syndrome, with no associated triggering factors.

CLINICAL CASE

A 31-year-old woman with no significant personal history, consulted with a clinical picture of 8 days of evolution that began with an unquantified thermal rise, edema in the lower extremities. Physical examination revealed erythematous papules and vesicles, some isolated and others grouped, located on the extremities, dorsum and palm of the hands (Figure 1 and 2), accompanied by pruritus. With a diagnosis of Sweet's syndrome versus erythema elevatum diutinum, a skin biopsy was requested.

Histopathologic study revealed accentuated edema in papillary dermis with formation of subepidermal blisters containing plasma and neutrophilic exudate (Figure 3). The underlying dermis with lymphocytic, neutrophilic and eosinophilic inflammatory infiltrate with superficial and mid perivascular and interstitial inflammatory infiltrate with presence of karyorrhexis (Figure 4). Findings compatible with Sweet's syndrome.

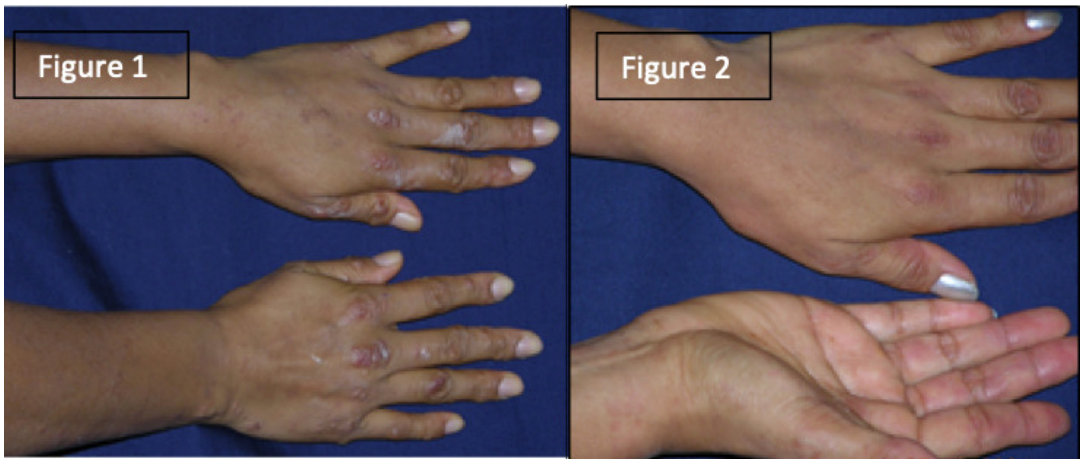

Figure 1: Papules, erythematous, edematous plaques, some with desquamation on the surface. With presence of vesicles.
Figure 2: Edema and erythema in the thumb of the fingers.

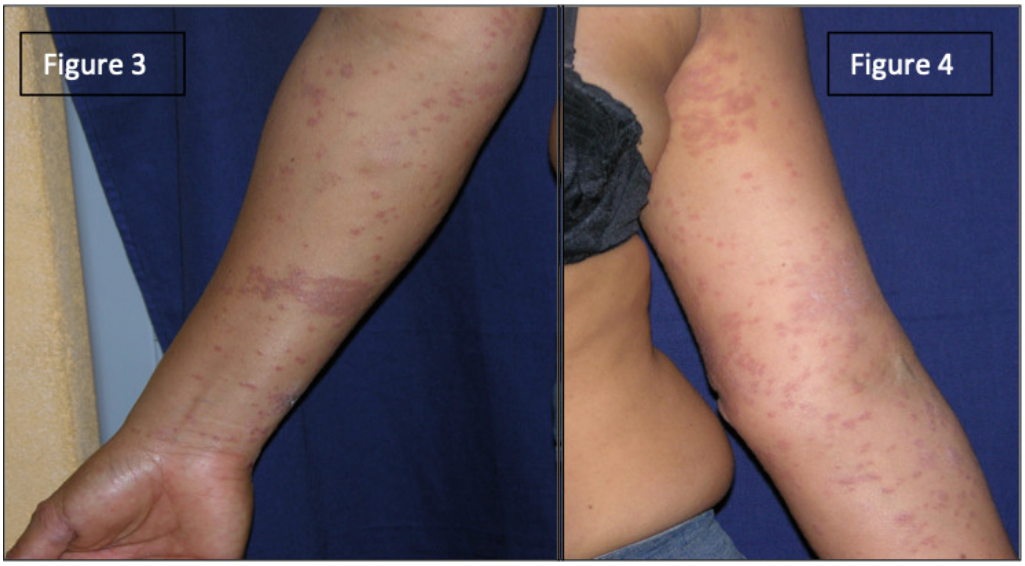

Figure 3 and 4: Erythematous papules and plaques on upper extremities.

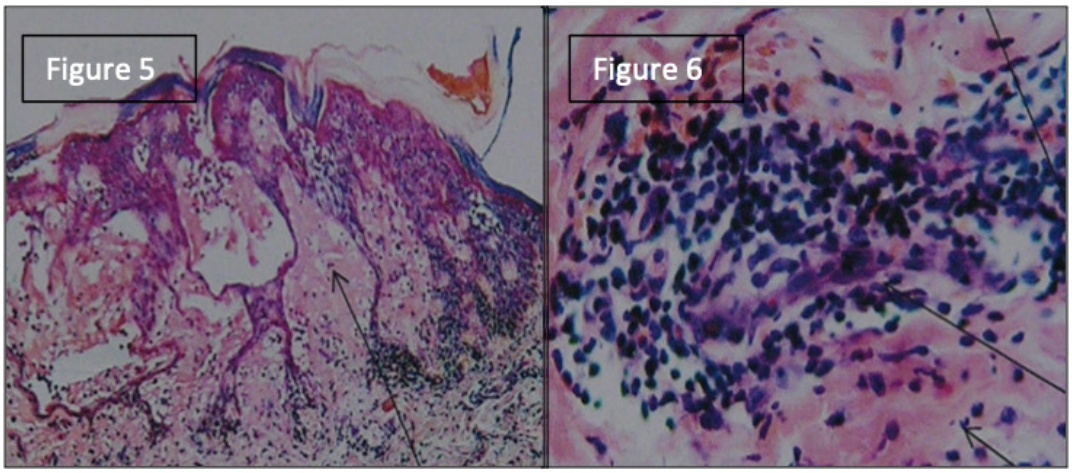

Figure 5: Presence of intense edema in papillary dermis with subepidermal blisters.
Figure 6: Perivascular neutrophilic, lymphocytic and eosinophilic inflammatory infiltrate.

DISCUSSION

Sweet's syndrome, also called acute febrile neutrophilic fever dermatosis, is a rare disorder, with an annual incidence of 2.7 - 3 cases per 1000000.[6] It occurs mainly in women aged 30-60 years sometimes associated with triggering factors,[7] although cases have also been reported in children.[6,8]

The diagnosis of Sweet's syndrome is based on typical clinical features, laboratory and histopathological findings.[5]

The diagnostic criteria were initially described by Su and Liu, being modified by von den Driesch in 1994, requiring the presence of neutrophilic infiltrate for its diagnosis, although there are reports of Sweet's syndrome without predominant neutrophilic infiltrate and the fulfillment of two major and two minor criteria is required (Table 1).[2]

On the other hand, in 1996, Walker and Cohen established the criteria for drug-induced Sweet's Syndrome, requiring the fulfillment of all criteria for its diagnosis.[2]

Clinically, fever is the most common symptom. Lesions usually appear as erythematous, edematous, typically painful papules, plaques or nodules. They can appear anywhere on the body, with the face, neck and upper extremities being the most frequent sites.[9]

Cutaneous lesions are accompanied by systemic symptoms such as fever, malaise, myalgias and arthralgias. On the other hand, it can manifest with extracutaneous lesions in the central nervous system, kidneys, liver, lungs and heart.[5]

Although its etiology is unknown, and it is generally idiopathic, many associated or triggering factors have been described in the literature, such as malignancies, infections mainly of the upper respiratory and gastrointestinal tract, pregnancy, drugs such as azathioprine, vaccines, autoimmune diseases such as Behcet syndrome, Sjogren syndrome, systemic lupus erythematosus, rheumatoid arthritis and inflammatory bowel disease, thyroid disease.[9,10]

Table 1: Diagnostic criteria of Sweet's Syndrome.

SWEET'S SYNDROME CRITERIA	
MAJOR CRITERIA	**MINOR CRITERIA**
1. Abrupt onset of painful erythematous plaques or nodules. 2. Presence of dense neutrophilic infiltrate without leukocytoclastic vasculitis on histopathology.	3. Temperature higher than 38C 4. Association of underlying visceral or hematologic malignancy, inflammatory disease or pregnancy, or being preceded by upper respiratory or gastrointestinal infection or vaccination. 5. Excellent response to systemic corticosteroids or potassium iodide. 6. Elevation of three of the following four laboratory parameters: a. Erythrocyte sedimentation rate higher than 29mm/h b. C-reactive protein positive c. Leukocytosis greater than 8000 d. Neutrophilia greater than 70%.

This entity is classified into three categories: classical or idiopathic SS, being the most frequent form, SS associated with malignancies and drug-induced SS.[5]

Sweet's syndrome in the context of neoplasias occurs in 10-20% of cases,[6] it constitutes a rare paraneoplastic syndrome that usually occurs in hematological malignancies, mainly acute myeloid leukemia, being its association with solid tumors infrequent. It can be a manifestation of an undiagnosed malignancy or present as an initial sign of cancer recurrence.[5,11]

A wide variety of drugs have been reported in the literature as triggers of SS such as antibiotics, antineoplastics, granulocyte colony stimulating factors, anticonvulsants,[5] non-steroidal anti- inflammatory drugs, and associated with vaccines such as BCG, influenza, pneumococcal and recently with the vaccine for COVID-19 Pfizer, Astrazeneca,[4] Moderna.[9]

Histopathology typically shows a dense neutrophilic infiltrate with edema without vasculitis.[11] In some cases lymphocytes and eosinophils are found, with the presence of spongiosis.[6] Histopathological variants have been described as: subcutaneous, eosinophilic, histiocytoid and lymphocytic.[6]

The approach and management of SS should be carried out systematically. Upon suspicion, it is necessary to identify extracutaneous involvement, associated diseases, infections and possible association with drugs. At this point it is necessary to rule out hematological diseases and associated neoplasms according to the symptomatology of each patient.[12]

Laboratory tests should include in addition to a complete blood count, acute phase reactants, renal and hepatic function, urinalysis, in certain cases, antistreptolysin O antibodies, rheumatoid factor, thyroid function, tumor markers may be necessary. Skin biopsy is indispensable and is routinely performed. If extracutaneous involvement is suspected, other diagnostic tests such as imaging studies are requested.[12]

In the classic form of SS, lesions may persist for weeks or months and then disappear. The first line of treatment is high doses of systemic corticosteroids of 0.5-1.5 mg/kg/day.[7] Other frequently used options are dapsone at doses of 100-200 mg/day, potassium iodide with doses of 900 mg/day and colchicine with doses of 1.5 mg/day.[7] Lesions usually improve 72 hours after the start of treatment.[12] On the other hand, the use of antibiotics such as doxycycline, as well as metronidazole, indomethacin, clofazimine, etretinate, thalidomide, TNF alpha and IL 1 antagonists in refractory cases have also proven to be effective.[11]

CONCLUSION

Sweet's syndrome also called acute febrile neutrophilic febrile dermatosis, is a rare disorder clinically manifested with fever, painful erythematous plaques and nodules, and leukocytosis with neutrophilia. It is divided or classified into a classic form of presentation, associated with mainly hematologic malignancies, and the drug-induced form. In our case no triggering or associated factors were found, so it is a classical or idiopathic form of the disease. The diagnosis is clinical, supported by diagnostic criteria, laboratory and histopathological findings. The mainstay of treatment is based on the use of systemic corticosteroids.

REFERENCES

1. Sweet R. AN ACUTE FEBRILE NEUTROPHILIC DERMTOSIS. Br J Dermatol. 1964;7(8–9):349–56.
2. Joshi TP, Friske SK, Hsiou DA, Duvic M. New Practical Aspects of Sweet Syndrome. Am J Clin Dermatol [Internet]. 2022;23(3):301–18. Available from: https://doi.org/10.1007/s40257-022-00673-4
3. Heath MS, Ortega-Loayza AG. Insights into the pathogenesis of Sweet's syndrome. Front Immunol. 2019;10(MAR).
4. Majid I, Mearaj S. Sweet syndrome after Oxford-AstraZeneca COVID-19 vaccine (AZD1222) in an elderly female. Dermatol Ther. 2021;34(6):3–5.
5. Khan U, Rizvi H, Ali F, Lebovic D. Sweet syndrome: A painful reality. BMJ Case Rep. 2016;2016:1–3.

6. PĂTRAŞCU V, GEOLOAICA LG, CIUREA RN. Recurrent Idiopathic Sweet Syndrome - Case Report and Literature Review. [Internet]. Vol. 46, Current Health Sciences Journal. 2020. p. 90–8. Available from: http://10.0.50.65/CHSJ.46.01.12%0Ahttps://search.ebscohost.com/login.aspx?direct=true&db=edb&AN=143215114&amp%0Alang=pt-pt&site=eds-live&scope=site
7. Shahid Z, Kalayanamitra R, Patel R, Groff A, Jain R. Refractory Sweet Syndrome Treated with Anakinra. Cureus. 2019;11(4).
8. dos Santos TBP, Sales BCG, Sigres M, Rosman F, de Cerqueira AMM. Sweet syndrome in childhood. An Bras Dermatol. 2015;90(4):567–9.
9. Pelchat F, Fournier C, Perron E, Gilbert M, Delisle B. Sweet syndrome following Moderna COVID-19 vaccine: A case report. SAGE Open Med Case Reports. 2022;10:1–4.
10. Sleiman J, Hitawala AA, Cohen B, Falloon K, Simonson M, Click B, et al. Systematic Review: Sweet Syndrome Associated with Inflammatory Bowel Disease. J Crohn's Colitis. 2021;15(11):1864–76.
11. Zheng S, Li S, Tang S, Pan Y, Ding Y, Qiao J, et al. Insights Into the Characteristics of Sweet Syndrome in Patients With and Without Hematologic Malignancy. Front Med. 2020;7(February):1–5.
12. Villarreal-Villarreal, D.D. Ocampo-Candiani J., Villarreal-Martinez, A. Sweet Syndrome: A Review and Update. Actas Dermo-Sifilográficas. 2016; 107 (5): 369-378