

- of a new recombinant hirudin. *J Am Acad Dermatol.* 2000;42: 612-9.
8. Palacios Colom L, Alcántara Villar M, Anguita Carazo JL, Ruiz Villaverde R, Quiralte Enriquez J. Delayed-type hypersensitivity to heparins: different patterns of cross-reactivity. *Contact Dermatitis.* 2008;59:375-7.
9. Nino M, Patruno C, Zagaria O, Balato N. Allergic contact dermatitis from heparin-containing gel: use of scratch patch test for diagnosis. *Dermatitis.* 2009;20:171-2.

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Sweet Syndrome in a Pregnant Woman

Síndrome de Sweet asociado al embarazo

To the Editor:

A 33-year-old woman, a primigravid at 16 weeks' gestation, came to our outpatient clinic due to the sudden appearance of a hot, infiltrated, edematous erythematous plaque, with clusters of pustules, and areas of crusting and desquamation (Figure 1). The lesion had appeared on the anterior aspect of her right thigh 4 days earlier. The patient had no relevant past medical history and no record of abortion, and her pregnancy was developing normally. Coinciding with the appearance of the lesion, she had been feeling feverish, and complained of pruritus and pain in the area; she thought the lesion might have been the result of an insect bite. She was diagnosed with cellulitis and was prescribed an oral antibiotic, an aqueous solution of 3 sulfates (copper, zinc, and potassium), and topical corticosteroids. The thigh lesion gradually improved, but 10 days after the first visit, the patient returned to the clinic with 2 similar plaques (1 on a wrist and 1 on the abdomen), urticarial in appearance, and characterized by intense pruritus (Figure 2).

As possible diagnoses, we considered urticarial vasculitis, eosinophilic panniculitis, herpes gestationis, and Sweet syndrome. Blood tests revealed iron deficiency anemia, and leukocytosis of 12000 cells/ μ L, both considered to be consistent with pregnancy. Samples taken for pathology study confirmed Sweet syndrome. Intense edema was observed in the upper dermis, accompanied by a predominantly perivascular neutrophilic inflammatory infiltrate, and a dense band-like inflammatory infiltrate in the papillary dermis (Figure 3). Treatment with 45 mg/d of deflazacort resolved the lesions in 10 days. The dose of corticosteroids was tapered over the following month, but the patient—22 weeks pregnant, and receiving 7.5 mg/d of deflazacort—returned with identical lesions at the same sites. Deflazacort was again prescribed at a dose of 45 mg/d for 15 days. The lesions resolved, and treatment was reduced to 10 mg on alternate days until vaginal delivery at 39 weeks without complications. Two years later the patient remains asymptomatic.

Sweet syndrome is named after Dr. Robert Douglas Sweet, who first described the disorder in 1964. Also known as acute febrile neutrophilic dermatosis, it has a worldwide distribution and is most common in women aged 30 to 50 years. Five subtypes have been identified: classical or idiopathic (71%); infection- or autoimmune-associated

(15%), paraneoplastic (10%-20%); pregnancy-associated (2%); and drug-induced.¹

The etiology is unknown. The fact that Sweet syndrome is predominantly a disorder of women and is associated with pregnancy and oral contraceptives would suggest a hormonal origin. Elevated estrogen and progesterone levels during pregnancy may be responsible for the vascular, cellular, microbiological, and immunological changes linked to the pathogenesis of pregnancy-associated Sweet syndrome.² Diagnosis is complicated by the fact that skin lesions are not always accompanied by the typical triad of fever, anemia, and leukocytosis with neutrophilia and an increased erythrocyte sedimentation rate. For this reason, we are of the opinion that Sweet syndrome is probably underdiagnosed.

The skin lesions typically present as clearly circumscribed, infiltrated erythematous papules and plaques, with marked edema. Vesicles and blisters may also be observed. Around 33% of patients relapse—as happened with our patient—as corticosteroid treatment is tapered off. Lesions tend to occur mostly on the upper part of the body, although, as happened with our patient, they may also appear elsewhere.³



Figure 1 First episode. Hot, infiltrated, edematous erythematous plaque on the right thigh, with clusters of pustules, and areas of crusting and desquamation.

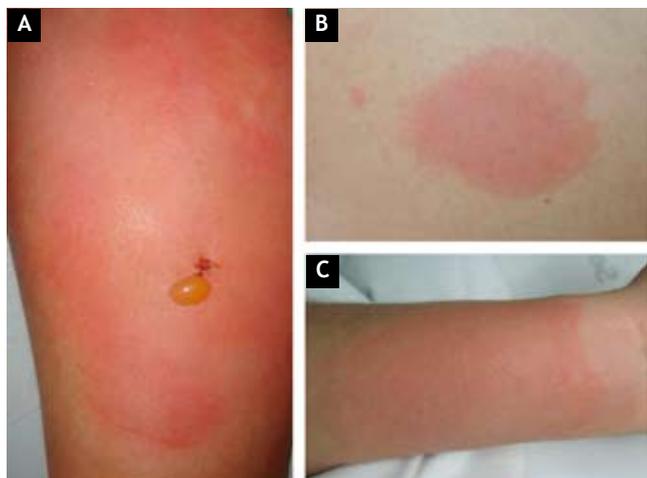


Figure 2 Recurrence. A, The plaque on the thigh reappeared with the same symptoms. B and C, Two new plaques appeared, on the abdomen (B) and on a wrist (C).

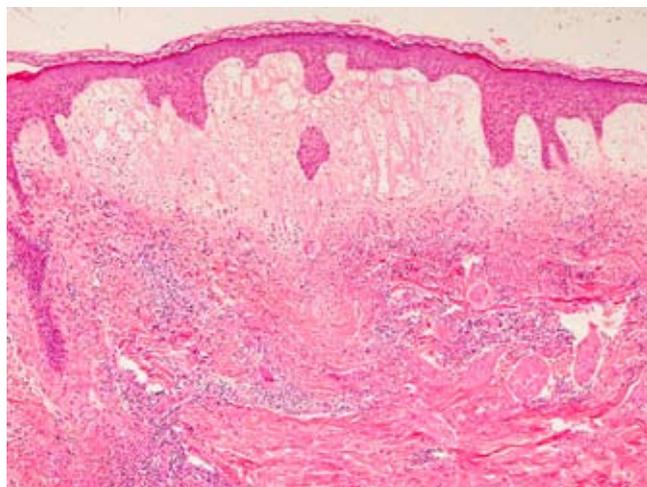


Figure 3 Histology study (hematoxylin-eosin staining, original magnification $\times 10$). Band-like, dense inflammatory infiltrate in the papillary dermis and edema in the upper dermis accompanied by a predominantly perivascular neutrophilic infiltrate.

Although infrequent, bone, joint, central nervous system, ophthalmological, hepatic, renal, intestinal, and cardiopulmonary complications have been reported. These complications all respond to corticosteroid treatment.

Pregnancy-associated Sweet syndrome typically resolves after delivery. Corticosteroids—rated by the US Food and Drug Administration as category B drugs (with no apparent risk, as no studies have demonstrated a risk to humans)⁴—should be considered as treatment, under medical supervision, for recurring, very extensive, and systemic cases. Treatment should commence with 1 mg/kg/d or less, in a single dose taken in the morning, and should be tapered off until the minimum effective dose is determined. Combination with other immunosuppressant drugs is contraindicated, as ciclosporin, azathioprine, and methotrexate are rated as category C, D, and X drugs, respectively. Topical corticosteroids (most commonly clobetasol propionate

0.05%) are indicated for one-off treatments or as adjuvant treatment when lesions are very localized.

For our patient in her second trimester, with symptomatic and recurrent lesions that failed to respond to local treatment, the use of systemic corticosteroids was assessed in collaboration with the obstetrics department. Treatment was started with a low dose (45 mg/d) of corticosteroids, with the intention of determining the minimum effective dose in as short a time as possible. Obstetric monitoring was performed as for an at-risk pregnancy. When there is no response to systemic corticosteroids, other treatment options to be considered include colchicine and indomethacin, both category B drugs. Potassium iodide is contraindicated during pregnancy (category X), as are the new biologic agents.⁵

We report a new case of pregnancy-associated Sweet syndrome. Only 8 such cases were found in the English-language literature, and none in the Spanish literature.⁶⁻¹⁰

We draw particular attention to the diagnostic difficulty arising from the low frequency of pregnancy-associated Sweet syndrome, the very broad differential diagnosis, and laboratory alterations that are compatible with those associated with gestation.

References

1. Cohen PR, Kurzrock R. Sweet's syndrome revisited: a review of disease concepts. *Int J Dermatol*. 2003;42:761-78.
2. Ambros Rudolph CM, Mullegger RR, Vaughan Jones SA, Kerl H, Black MM. The specific dermatoses of pregnancy revisited and reclassified: results of a retrospective two-center study on 505 pregnant patients. *J Am Acad Dermatol*. 2006;54:395-404.
3. 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;88:601-6.
4. Fármacos y embarazo. Clasificación FDA según riesgo de embarazo. Available from: <http://www.cfnavarra.es/salud/PUBLICACIONES/Libro%20electronico%20de%20temas%20de%20Urgencia/22.Ginecologicas/Farmacos%20y%20embarazo.pdf>.
5. Yamauchi PS, Turner L, Lowe N. Etanercept as a new therapeutic option in Sweet's Syndrome. *J Am Acad Dermatol*. 2006;54:123-6.
6. Giovanna Brunasso AM, Massone C. Clinical images. Sweet syndrome during pregnancy. *CMAJ*. 2008;179:967.
7. López-Sánchez M, García-Sánchez Y, Marín AP. An unusual evolution of a pregnancy-associated Sweet's Syndrome. *Eur J Obstet Gynecol Reprod Biol*. 2008;140:283-5.
8. Hussain W, Craven N, Agarwal M, Kumar S. A painful cutaneous eruption in a pregnant woman. *Clin Exp Dermatol*. 2007;32:227-8.
9. Cohen PR. Pregnancy-associated Sweet's syndrome: world literature review. *Obstet Gynecol Surv*. 1993;48:584-7.
10. Satra K, Zalka A, Cohen PR, Grossman ME. Sweet's syndrome and pregnancy. *J Am Acad Dermatol*. 1994;30:297-300.

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