

CASE REPORTS

Chronic Recurrent Neutrophilic Dermatitis: A Possible Variant in the Spectrum of Neutrophilic Dermatoses

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Abstract. Neutrophilic dermatoses constitute a clinically heterogeneous group of diseases that share a common histological substrate, consisting of a dense dermal inflammatory infiltrate of mature polymorphonuclear neutrophils and no evidence of vasculitis. We describe the case of a 56-year-old man with a 6-month history of painful generalized erythematous edematous plaques. Histopathology indicated neutrophilic dermatitis but the patient did not have fever, elevated white blood cell count, or systemic involvement. Tests to rule out possible inflammatory, neoplastic, or infectious processes were negative. We consider the term chronic recurrent annular neutrophilic dermatitis, first used by Christensen et al, to be the most appropriate to define this variant with clinical findings that differ from classic Sweet syndrome.

Key words: neutrophilic dermatitis, chronic disease, recurrence.

DERMATOSIS NEUTROFÍLICA CRÓNICA RECURRENTE: UNA POSIBLE VARIANTE EN EL ESPECTRO DE LAS DERMATOSIS NEUTROFÍLICAS

Resumen. Las dermatosis neutrofilicas constituyen un grupo clínicamente heterogéneo de enfermedades con un amplio espectro clínico que comparten un sustrato histopatológico común, consistente en un denso infiltrado inflamatorio dérmico de polimorfonucleares neutrófilos maduros sin evidencia de vasculitis. Describimos el caso de un varón de 56 años con una historia de 6 meses de evolución de placas eritematoedematosas dolorosas generalizadas, con un patrón histopatológico de dermatosis neutrofilica pero sin fiebre, leucocitosis ni afectación del estado general. Los estudios realizados para descartar una posible asociación con procesos inflamatorios, neoplásicos o infecciosos resultaron negativos. Creemos que el término de dermatosis neutrofilica anular crónica y recurrente acuñado por Christensen et al es el más adecuado para definir esta variante con hallazgos clínicos que difieren del clásico síndrome de Sweet.

Palabras clave: dermatosis neutrofilica, enfermedad crónica, recurrencia.

Introduction

Acute febrile neutrophilic dermatitis was first described by Sweet in 1964,¹ based on 4 main criteria: fever, peripheral blood leukocytosis, erythematous edematous plaques on the arms, face, and neck, and a dense histologic infiltrate of mature neutrophils. In general, given that it is currently accepted that both fever and leukocytosis are not always

present in this condition, whereas extracutaneous manifestations are common, and chronic and recurrent forms have been previously described,²⁻⁷ the term Sweet syndrome seems more appropriate than acute febrile neutrophilic dermatitis to refer to this entity.⁸

Case Description

We describe a 56-year-old man with no significant medical history, admitted to our clinic for symptoms that had started approximately 6 months earlier. These included recurrent outbreaks of edematous, infiltrated, annular, reddish-violet plaques, painful to the touch, located on the trunk (Figure 1), arms (Figure 2), neck and forehead, and legs (Figure 3), which progressed to residual hyperpigmented lesions. He did not present fever or malaise, although he did report

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Figure 1. Erythematous edematous annular plaques, of elastic consistency, on the upper trunk.



Figure 2. Reddish-violaceous annular lesions in the arm progressing to residual hyperpigmented macules.



Figure 3. Annular lesions with a violaceous center and erythematous halo on a thigh.

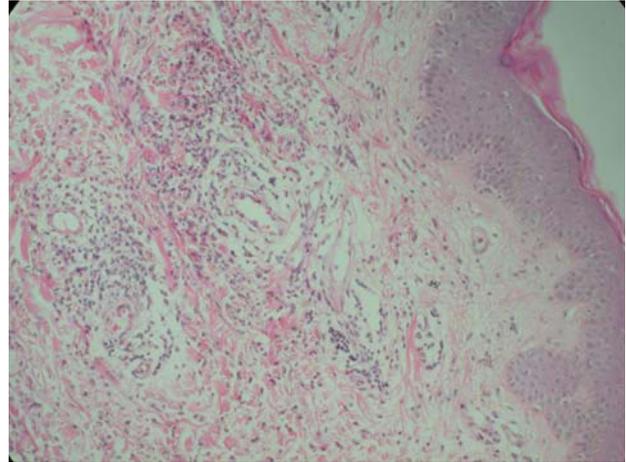


Figure 4. Edema and intense predominantly neutrophilic inflammatory infiltrate in the dermis (hematoxylin-eosin stain, $\times 250$).

arthromyalgia and presented mild bilateral conjunctivitis. Complete blood count showed 4100 white blood cells/ μL with 74% polymorphonuclear neutrophils. The erythrocyte sedimentation rate was 31 mm/h. The antinuclear antibody test was positive at low titers (1/40). Anti-DNA antibody tests were negative. Chest X-ray, abdominal ultrasound, upper gastrointestinal endoscopy and colonoscopy, and bone-marrow aspiration and biopsy showed no abnormalities. All blood tests performed to rule out associated infections, including hepatotropic virus, human immunodeficiency virus, cytomegalovirus, syphilis, and herpes simplex were negative. Histopathology of a pectoral skin biopsy indicated a dense neutrophilic inflammatory infiltrate in the upper dermis without findings suggestive of leukocytoclastic vasculitis (Figure 4). Cutaneous lesions improved after beginning treatment with oral prednisone (50 mg/d), with gradual tapering of the dose. Occasional mild recurrences were reported.

Discussion

Sweet syndrome has been associated with a wide range of processes and situations, including infectious and inflammatory diseases, neoplastic disease, drug use, vaccination, and even pregnancy.⁹⁻¹¹ Nevertheless, these conditions can only be identified in approximately 30% of cases. An occult neoplasm can be identified in 20% of cases, mainly hematologic neoplasia (85%), the most frequent types being acute myelogenous leukemia and lymphomas.¹² The absence of factors commonly associated with triggering classic Sweet syndrome (upper respiratory tract infections, drugs, etc), male predominance, presence of more severe generalized cutaneous lesions, frequent extracutaneous disorders, absence of neutrophilia, presence of anemia or

abnormal platelet count and, in particular, the high rate of recurrence, are clinical observations that frequently appear in the context of malignancy-associated Sweet syndrome.⁸ On the other hand, in these cases, cutaneous lesions can precede the onset of neoplastic processes even by several years.¹³

In 1989, Christensen et al⁶ described 2 cases of patients with chronic and recurrent outbreaks of generalized annular erythematous, edematous cutaneous plaques, with histopathological findings suggestive of Sweet syndrome, but without fever or general symptoms. Although some cases of recurrent neutrophilic dermatosis in isolation had been described previously,²⁻⁵ Christensen et al were the first to regard this as an entity in itself, coining the term chronic recurrent annular neutrophilic dermatosis to refer to this variant of neutrophilic dermatosis with histopathological findings typical of this entity, but with a chronic and recurrent course differing from classic Sweet syndrome and no evidence of associated systemic disease.

Subsequently, Romero et al⁷ reported 4 cases of patients with lesions clinically or histologically compatible with Sweet syndrome, but with an afebrile and recurrent course lasting several years, without presenting leukocytosis or peripheral neutrophilia at any time. They referred to these cases as chronic recurrent afebrile neutrophilic dermatosis and, unlike Christensen et al,⁶ regarded this process as a variant of Sweet syndrome and not as a separate clinical entity.

From the clinical standpoint, the absence of fever and leukocytosis is the norm in most chronic and recurrent cases, although these criteria may be present in some instances.^{2,5} On the other hand, some of the forms of chronic recurrent neutrophilic dermatosis described present as single lesions, especially on the face.^{2,4} Furthermore, this chronic variant does not seem to respond so well as classic Sweet syndrome to systemic corticosteroids, and recurrence is frequent, as in our case.⁵⁻⁶

Our patient shares several of the aforementioned clinical criteria that are more often associated with Sweet syndrome. However, to date, all the tests performed to diagnose this disease have been negative. Thus, we believe that the term chronic recurrent annular neutrophilic dermatosis best describes our patient's disease. In any case, the fact that cutaneous lesions can precede the diagnosis of malignancy by several years suggests that chronic recurrent annular neutrophilic dermatosis can be an early manifestation of

systemic disease such as cancer and thus clinical follow-up in these patients is recommended.

In conclusion, we describe a case of neutrophilic dermatosis of atypical, chronic, and recurrent evolution. Further studies on a large series of similar cases may help us to understand whether we are facing a true variant within the spectrum of neutrophilic dermatosis or, on the other hand, whether the condition is a severe form of Sweet syndrome as an early manifestation of a systemic disease.

Conflicts of Interest

The authors declare no conflicts of interest.

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