

# ACTAS Derma-Sifiliográficas

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## CASE FOR DIAGNOSIS

### Recurrent Blistering Rash

#### Erupción ampollosa recidivante

#### Medical History

A 42 year-old man with no relevant past history consulted for lesions on the trunk and arms present for more than 24 hours. He reported several similar outbreaks in the last 9 months that had resolved following treatment with corticosteroids and oral antihistamines.

#### Physical Examination

Physical examination revealed a number of well-defined indurated erythematous and edematous plaques on the back, some in a curvilinear distribution (Figure 1). Similar plaques covered in multiple vesicles and occasional blisters containing a serous fluid were observed on both arms (Figure 2).

#### Histopathology

Histopathology showed an intact epidermis and intense edema of the dermis associated with a diffuse infiltrate consisting mainly of lymphocytes and eosinophils. The deep dermis presented foci of eosinophilic degenerative collagen (Figure 3).

#### Additional Tests

The results of blood tests including complete blood count, basic blood chemistry, protein electrophoresis, autoimmunity, and peripheral blood smear were all normal. Immunoglobulin E (IgE) levels were slightly elevated. A fecal parasite test was negative.



Figure 2



Figure 1

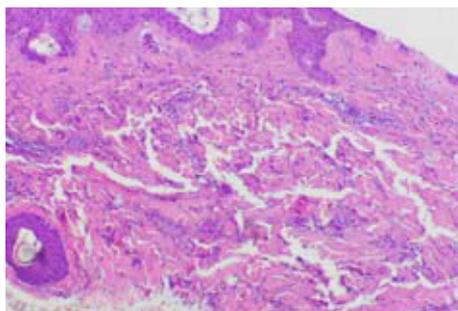


Figure 3 Hematoxylin-eosin, original magnification, 100.

**What Is Your Diagnosis?**

## Diagnosis

Wells syndrome or eosinophilic cellulitis.

## Clinical Course and Treatment

The patient received treatment with a tapering course of oral prednisone at an initial dosage of 0.5 mg/kg/d. The lesions disappeared in less than 48 hours. There has been no recurrence in 1 year of follow-up.

## Comment

The first 4 cases of Wells syndrome (WS) were described by George Wells<sup>1</sup> in 1971 and Wells and Smith<sup>3</sup> renamed the condition eosinophilic cellulitis in 1979. This is a rare inflammatory dermatosis that presents marked clinical polymorphism, a characteristic histology, and a clinical course that is usually self-limiting and recurrent. It is seen most often in adults, although it is increasingly common in children and even among newborns.<sup>3</sup> The pathogenesis is unknown and although appearance of the disease has been related to many factors, in about 50% of cases no trigger is identified.<sup>4</sup>

Classic WS presents as a series of recurrent prodromes characterized by an itching or burning sensation followed by the appearance of highly edematous nodules and annular or curved plaques with violaceous borders. Vesicles and blisters may occasionally appear.<sup>5,6</sup> The initial lesions are a bright red color, fading to a greenish, brownish or slate-gray tone and healing in 2 to 8 weeks. They are most common on the legs, trunk, and upper arms. Less than 25% of cases are associated with fever and 50% present peripheral blood eosinophilia during the active phase of the disease.<sup>4</sup>

Histopathology shows 3 phases<sup>1-3,5,6</sup>: an acute phase characterized by dermal edema and a dense infiltrate consisting fundamentally of eosinophils; a subacute phase with the appearance of "flame figures" consisting of collagen fibers covered by the products of eosinophil degranulation (although a trait of WS, these figures are non-specific as they are also found in many other diseases associated with significant eosinophil activation and degranulation); and a healing phase characterized by a

gradual disappearance of eosinophils but with a persistence of histiocytes and the appearance of giant cells around the collagen deposits.

Differential diagnosis is principally with bacterial cellulitis. Diagnosis is supported by a lack of response to antibiotics, recurrences, local prodromal symptoms, eosinophilia, and lesions less hot and tender than is usual in bacterial lesions.

Initial treatment consists of oral corticosteroids and these tend to produce a spectacular improvement within a matter of days. Etiological treatment can also be effective when a clear trigger is identified.<sup>4,6</sup>

## Conflict of Interest

The authors declare that they have no conflict of interest.

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J. Alonso-González, J. García-Gavín, J. Toribio\*

*Departamento de Dermatología, Complejo Hospitalario Universitario, Facultad de Medicina, Santiago de Compostela, Spain*

\*Corresponding author.

*E-mail address:* jaime.toribio@usc.es (J. Toribio).