CASES FOR DIAGNOSIS

Girl With Recurrent Erythematous Edematous Plaques on Her Legs

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Patient History

A 4-year-old girl with a history of surgery for strabismus was assessed in the emergency department for pruritic lesions on the right leg that had appeared 3 days previously. Although the patient was afebrile and in good general health, she was diagnosed with bacterial cellulitis and admitted to the hospital for intravenous antibiotic therapy. Improvement was very slow and the patient required a switch from amoxicillin-clavulanic acid to cefotaxime plus clindamycin; she was discharged 11 days later.

A week later, she was taken back to the emergency room due to the appearance of a similar lesion on the contralateral ankle. She was afebrile and in good general health.

Physical Examination

The physical examination showed well-delimited erythematous edematous plaques that affected the dorsum, side, and heel of the right foot (Figure 1). She also had blisters, some of them with bloody content (Figure 2). The thighs and legs showed papular lesions of similar characteristics.

Additional Examinations

The only relevant finding was 10.8% eosinophilia. All other tests (biochemistry, urine, antinuclear antibodies, protein analysis, complement, circulating immune complexes, rheumatoid factor, C-reactive protein, erythrocyte sedimentation rate, immunoglobulin E, antibodies against *Borrelia burgdorferi*, and fecal parasites) were normal or negative. A punch biopsy of the skin revealed a perivascular and interstitial inflammatory infiltrate predominantly made up of eosinophils (Figures 3 and 4). Flame figures were

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Manuscript accepted for publication June 7, 2007





Figure 1.

Figure 2.



Figure 3. Hematoxylineosin, ×200



Figure 4.

common, but the epidermis showed no abnormalities in any of its layers. No immune deposits were observed by direct immunofluorescence.

What is your diagnosis?

Diagnosis

Eosinophilic cellulitis or Wells syndrome

Course

A topical corticosteroid was prescribed; the patient progressed favorably with complete resolution of the lesions and no recurrences at the time of writing.

Comment

Eosinophilic cellulitis is a rare condition first described in 1971 by Wells, who called it "recurrent granulomatous dermatitis with eosinophilia." Wells and Smith described new cases using the term "eosinophilic cellulitis."¹

The condition occurs most often in adulthood, although about 30 cases have been reported among children. It is characterized by recurrent episodes of erythematous, edematous lesions with well-defined borders; the lesions may also be blistered. They are found most often on the trunk and limbs. The patient's general health is not usually affected and systemic complications (fever, joint pain, pericarditis, enlarged liver and spleen, superinfection, etc) are uncommon, but occur more often in children than adults.¹

During resolution, the lesions acquire a brownish, morphea-like appearance. Complete remission occurs in 4 to 8 weeks with no residual scarring. Recurrences are common and may last for years.¹⁻⁴

Abnormalities are rarely found in the results of laboratory tests, except for eosinophilia in peripheral blood and bone marrow in 50% of cases.

Histopathology reveals various phases according to the stage in which the biopsy is done. The first phase includes considerable edema along with a predominantly eosinophilic infiltrate. Later, in the subacute phase, the typical flame figures are formed by collagen degeneration secondary to eosinophilic degranulation. The last (granulomatous) phase shows numerous histiocytes surrounding the flame figures.⁵⁻⁷ There are numerous conditions in which flame figures can be found (insect bites, parasitosis, bullous pemphigoid, herpes gestationis, tinea infection, allergic eczemas, eosinophilic ulcer of the oral mucosa, drug reactions, eosinophilic folliculitis, etc). As a result, eosinophilic cellulitis as a defined entity has been questioned.

The etiology of Wells syndrome is unknown. It has been related to multiple triggering factors, such as insect bites, parasitosis, myeloproliferative or lymphoproliferative disorders, viruses, drugs, tinea infection, solid tumors, etc, and these should always be ruled out.⁸ It is currently considered to be a hypersensitivity reaction triggered by several factors. Chemotaxis and activation of eosinophils appears to be mediated by interleukin 5, and eosinophil degranulation causes dermal collagen to degenerate and the classic flame figures to form.^{9,10}

Eosinophilic cellulitis resembles various processes, both in terms of symptoms and pathology. Therefore, clinical and pathologic findings should be adequately correlated to obtain an accurate diagnosis.

Because this is a benign, self-limiting condition, it is difficult to assess the efficacy of the various available treatments, of which corticosteroids (topical and systemic) are the therapy of choice for use in children.²

Conflicts of Interest

The authors declare no conflicts of interest.

References

- Van der Straaten S, Wojciechowski M, Salgado R, Menten G, Mees N, Hagendorens M. Eosinophilic cellulitis or Wells' syndrome in a 6-year-old child. Eur J Pediatr. 2006;165:197-8.
- Áparicio S, Torrelo A, Medeiro IG, Zambrano A. Síndrome de Wells en la infancia. Presentación de un caso y revisión de la literatura. Actas Dermosifiliogr. 2000;91:343-8.
- Gilliam AE, Bruckner AL, Howard RM, Lee BP, Wu S, Frieden IJ. Bullous "cellulitis" with eosinophilia: case report and review of Wells' syndrome in childhood. Pediatrics. 2005;116:149-55.
- Kuwahara RT, Randall MB, Eisner MG. Eosinophilic cellulitis in a newborn. Pediatr Dermatol. 2001;18:89-90.
- Moossavi M, Mehregan DR. Wells' syndrome: a clinical and histopathologic review of seven cases. Int J Dermatol. 2003;42:62-7.
- Consigny S, Courville P, Young P, Richard C, Gauthier V, Maillard V, et al. Histological and clinical forms of eosinophilic cellulitis. Ann Dermatol Venereol. 2001;128:213-6.
- Ríos-Martín JJ, García-Escudero A, González-Cámpora R. Aproximación histológica al diagnóstico de las dermatitis con eosinófilos. Piel. 2005;20:405-13.
- Fujii K, Tanabe H, Kanno Y, Konishi K, Ohgou N. Eosinophilic cellulitis as a cutaneous manifestation of idiopathic hypereosinophilic syndrome. J Am Acad Dermatol. 2003;49:1174-7.
- Yagi H, Tokura Y, Matsushita K, Hanaoka K, Furukawa F, Takigawa M. Wells? syndrome: a pathogenic role for circulating CD4+??CD7- T cells expressing interleukin-5 mRNA. Br J Dermatol. 1997;136:918-23.
- España A, Sanz ML, Sola J, Gil P. Wells? syndrome (eosinophilic cellulitis): correlation between clinical activity, eosinophil levels, eosinophil cation protein and interleukin-5. Br J Dermatol. 1999;140:127-30.