

A Case of Eosinophilic Cellulitis (Wells' Syndrome)

BİR EOZİNOFİLİK SELÜLİT (WELLS SENDROMU) OLGUSU

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Abstract

Wells' syndrome or eosinophilic cellulitis is a rare inflammatory dermatosis with uncertain pathogenesis and recurrent lesions resembling cellulitis. Bullous lesions may be seen as well as pruritic, erythematous and indurated plaques and nodules. Lesions resolve spontaneously without scarring.

Typical histopathological picture shows dermal eosinophilic infiltration and characteristic "flame figures".

Herein we report a 47 year old man presenting with erythematous, indurated plaques on the arms, legs and body who is later diagnosed clinically and histologically as eosinophilic cellulitis.

Key Words: Eosinophilic cellulitis, Wells' syndrome, flame figures

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Özet

Wells sendromu veya eozinofilik selülit, patogenezi tam olarak bilinmeyen, tekrarlayıcı, klinik olarak selülit düşündürülen, nadir bir inflamatuvar dermatozdur. Kaşıntılı, eritemli, endüre plak ve nodüllere büller de eşlik edebilir. Lezyonlar skar bırakmadan spontan geriler.

Histopatolojik incelemede; dermiste eozinofil infiltrasyonu ve karakteristik 'alev şekilleri' görülmesi tipiktir.

Burada kol, bacak ve gövdesinde tekrarlayan eritemli, endüre plaklarla başvuran klinik ve histopatolojik özellikleriyle eozinofilik selülit tanısı alan 47 yaşında bir erkek olgu sunulmaktadır.

Anahtar Kelimeler: Eozinofilik selülit, Wells sendromu, alev şekilleri

Eosinophilic cellulitis (EC), first described by Wells in 1971, is a recurrent, inflammatory dermatosis characterized by a cellulitis-like eruption¹ Vesicles and bullae may develop as well as pruritic, erythematous, indurated plaques and nodules. Lesions resolve spontaneously without scarring.²⁻⁴ It affects predominantly adults.⁵ During exacerbations peripheral blood eosinophilia and an increase in total IgE levels can be detected.⁶

The disease has a benign course. In some cases systemic findings such as fever, fatigue and arthralgias may be seen.^{6,7}

Case Report

47 year old man presented with a complaint of edematous, erythematous and blistering eruption on his right forearm. The patient was diagnosed clinically as bacterial cellulitis and parenteral antibiotics were prescribed. When he came back, the lesions regressed almost completely without using any of the drugs. But he had similar plaques on his back and upper right arm for 2 days.

He had a 7-year history of similar lesions occurring periodically. His medical history was significant for lung tuberculosis, hypertension and depression. Systemic examination showed no abnormalities.

Dermatological examination revealed erythematous, indurated plaques with intact vesicles on the lateral aspect of the upper arm and on the back (Figure 1,2).

Histopathological examination revealed epidermal acanthosis and spongiosis and pronounced

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Figure 1. Erythematous indurated plaques with vesicles.



Figure 2. Erythematous edematous plaques with irregular borders.

perivascular and interstitial infiltration of eosinophilic polymorphous dermal infiltrate. Interstitial edema and focal areas of collagen degeneration consistent with flame figures were observed (Figure 3).

Laboratory studies revealed an elevated Total IgE level (324 IU/ml; normal:0-87 IU/ml). CBC, ESR, serum immunoglobulins, urinalysis, antinuclear antibody (ANA), serum electrolytes, liver function tests, kidney function tests, stool analysis for parasites, abdomen ultrasound and chest X-ray were within normal limits. HSV Type I IgG antibody was positive but antibodies for HSV Type I IgM and HSV Type II IgG and IgM were negative. Peripheral blood eosinophils and eosinophilic cationic protein levels were within normal ranges. Direct and indirect immunofluorescent examinations of the skin were negative.

Clinical and histopathological findings of the patient were consistent with eosinophilic cellulitis. A specific triggering factor could not be identified for our patient and was considered as idiopathic.

Discussion

Once referred as a distinct entity, currently EC is considered as a histopathological reaction pattern characterized by intense infiltration of eosinophils.^{6,8}

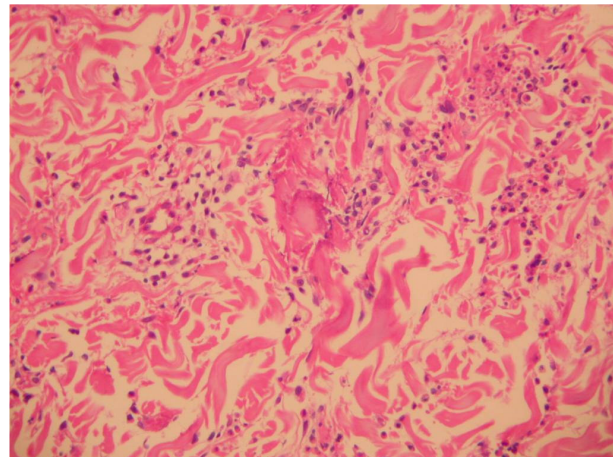


Figure 3. Diffuse eosinophil infiltration and flame figures (H&Ex40)

The most common presentation is solitary or multiple urticarial edematous plaques or erythematous papules and nodules located on the body or extremities.¹ Clinical picture has two phases; first phase is characterised by localised erythema and edema of the skin. Papules and bullae may be observed. The second phase consists of lesions that may last for 6 weeks and resolve without scarring.^{1,9,10} Lesions may be painful and itchy. Although systemic findings such as fever, fatigue, arthralgia may complicate the picture, the course is usually benign but recurrences may last for years.^{7,10} Apart from this classical form, there are reported cases of EC with indurated borders and

circinate lesions.^{11,12} Our patient had a 7 year history of recurrent, cellulitis-like plaques with vesicles located on the upper and lower limbs and the body.

Peripheral blood eosinophilia is a common finding but not obligatory for establishing the diagnosis. There are reported cases of bone marrow eosinophilia.³ Elevated IgE levels are occasionally found.⁵ Our patient had no peripheral blood eosinophilia but elevated levels of IgE.

Histopathologically; in the acute phase of cellulitis-like lesions, edema and diffuse infiltration of eosinophils are observed in the dermis, this infiltrate may occasionally extend to subcutaneous tissue and fascia. Where the epidermis is substantially involved, spongiotic intraepidermal vesicles develop. In the subacute or granulomatous phase; the characteristic flame figures appear. The core of these flame figures consist of granular deposition of eosinophilic material on the collagen bundles and the periphery is composed of degranulated eosinophils and major basic protein. The flame figures may develop a palisade of macrophages and sometimes giant cells. Resolution phase reveals phagocytic histiocytes around the flame figures. Necrobiosis may develop within the palisading histiocytic reaction. Vasculitis is an unexpected finding.^{1,2,13}

Flame figures are diagnostic but not specific for EC. They may also exist in bullous pemphigoid, prurigo, eczema, insect bites, drug reactions, parasitic and dermatophytic infections as a consequence of eosinophil degranulation.¹⁴ Therefore the diagnosis of EC should not be made based on only histopathologic findings. It should be made on the basis of recurrent skin lesions and clinical and histological picture.⁸ Typical flame figures observed on the light microscopy with recurrent clinical findings ensured the diagnosis of EC in the present case.

Although direct immunofluorescent is usually negative, there have been reported deposition of fibrin and C3, IgM, IgA and IgG on the basement membrane and dermis.^{2,3,10} The direct immunofluo-

rescent study we performed in our case was also negative.

The etiology of EC is unknown. Currently it has been hypothesized that it is an abnormal eosinophilic response to a variety of agents like drugs, insect bites, fungal, viral and parasitic infections.^{2,5} It may be idiopathic or associated with myeloproliferative diseases.^{2,5,6}

EC is suggested as a cutaneous manifestation of systemic hypereosinophilic diseases that are characterised by eosinophilia, urticarial plaques and erythematous papulonodules.⁶

Eosinophil activation and degranulation are associated with many dermatosis. Although the mechanisms of eosinophil activation and the skin edema are unknown, the granular cationic proteins are accused. Since blood levels of eosinophilic cationic protein, IL-5 and eosinophilia are reported to be associated with clinical activity, the role of these cytokines in the pathogenesis of the disease has gained significance.^{15,16}

The therapy of choice is oral corticosteroids.^{2,7} Dapsone, griseofulvin, minocycline and cetirizine are reported to be succesful.^{2,9,12}

Despite the finding that in some cases staphylococcus and streptococcus are isolated from the lesions, systemic antibiotics and acyclovir were found to be unaffactive.³ The first attack of our patient resolved without therapy, but the second attack was controlled with systemic corticosteroids and antihistamines.

In conclusion; EC is characterised clinically by recurrent cellulitis like lesions and histopathologically by flame figures and has to be considered in differential diagnosis of cellulitis.

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