

## Case Report

# Eosinophilic cellulitis (wells syndrome): a case report

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### ABSTRACT

Eosinophilic Cellulitis is also known as Wells syndrome is uncommon dermatitis, characterized by the infiltration of eosinophils in the dermis. The exact etiology of the disease is unknown. Clinically, it is highly varied but commonly the presentation is pruritic erythematous plaque. We report a case of one and half years old healthy boy who developed itchy bullae on the dorsum of his hand with multiple erythematous papules over his extremities that started immediately after his vaccines. Histopathological examination of the lesion showed infiltrate eosinophils with typical flame figures. The case was successfully treated with corticosteroid course. This report aims to present a case of eosinophilic cellulitis (Wells syndrome) triggered by the vaccine with a literature review of the disease. In conclusion, atypical presentation of any inflammatory disorder or that not responding to appropriate treatment should trigger suspicion of eosinophilic cellulitis. Up to now, systemic corticosteroids are the most successful treatment of eosinophilic cellulitis.

**Keywords:** Eosinophilic cellulitis, Wells syndrome, Flame figures.

### INTRODUCTION

Eosinophilic cellulitis (Wells syndrome) is a rare inflammatory disorder, characterized by the eosinophilic accumulation in specific organs such as skin and lungs. It is a skin disorder characterized by recurrent pruritic or tender skin lesions resembling cellulitis. It has broad diagnostic categories and variable presentations; the most common documented presentation is large painful erythematous plaque.<sup>1</sup>

However, plaques pass through two phases over weeks. The first phase starts as erythematous centrifugally spreading plaques, usually following a prodrome of pain and burning sensation, within 1-3 weeks these plaques will progress into the second phase which is characterized

by induration that resolves by 4-8 weeks with hyperpigmentation or morphea-like skin atrophy.<sup>2,3</sup>

Despite the fact that the etiology of wells syndrome is unknown, certain precipitating factors have been described in the literature, including parasitic infections, vaccines, contact dermatitis, arthropod bites, hematologic disorders, myeloproliferative disease, systemic disease, malignancy and various medications.<sup>4</sup>

Peripheral eosinophilia (mild or moderate-profound) is present in more than 50%.<sup>5</sup> Although, eosinophilic cellulitis (Wells syndrome) has an excellent prognosis, it tends to resolve in weeks or months, usually without scarring. It occasionally recurs. In these recurrent cases, it can take years to ultimately resolve.<sup>5</sup>

The aim of this report was to present a case of eosinophilic cellulitis triggered by the vaccine with a literature review of the disease.

### CASE REPORT

One-and-half years old healthy boy brought by his parents complaining of very itchy recurrent skin lesions for 1-week duration, this is his second attack. The first attack was one year ago, that was generalized all over the body that persists for 2 months and resolved after a course of prednisolone.

The current attack started one week in the upper extremities then progress to the lower extremities, 2-days ago one of lesion on the dorsum of the hand gets worse associated with pain and hand swelling (Figure 1).



**Figure 1: Hemorrhagic bulla associated with edematous erythematous right hand.**

The lesion was associated with low grade fever 38°C started 3 days after his 18 months scheduled vaccines. He was otherwise healthy, no preceding history of arthropod bites or trauma, unremarkable medical and family history.



**Figure 2: The hemorrhagic bulla after incision and drainage.**

Regarding patient antenatal and postnatal history, he is a product of uneventful pregnancy, spontaneous vaginal delivery, full term on breastfeeding, and formula milk he started his weaning at 6 months of age. No known drug or

food allergy. The patient misdiagnosed as an abscess by a pediatric surgeon who did an incision and drainage for him and put him on oral antibiotic (Figure 2).

Skin examination revealed multiple erythematous crusted papules scattered in all extremities (Figure 3, 4).

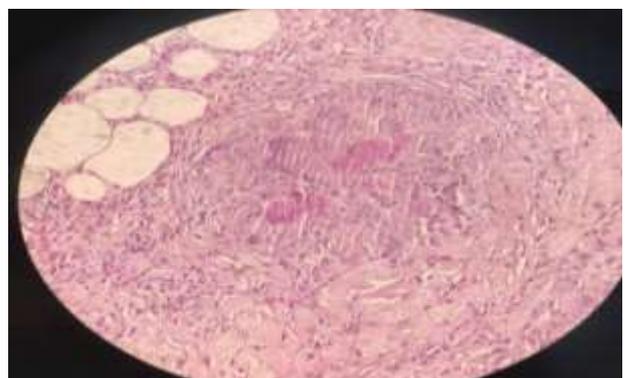


**Figure 3: Erythematous crusted papules over the left arm.**



**Figure 4: Erythematous crusted papule over the left thigh.**

In the dorsum of the right-hand middle finger there was necrotic ulcerated plaque this lesion was hemorrhagic bulla that ruptured and left an ulcerative necrotic plaque.



**Figure 5: Histopathology picture shows flame figures, and dermal infiltration by eosinophils consistent with Wells'.**

Skin biopsy was taken, histopathology showed normal epidermis apart from crust formation. In the dermis there was dense mixed cellular infiltrate throughout the dermis extending deeply composed mainly of eosinophils with numerous flame figures (Figure 5).

In the base of the above clinicopathologic finding the patient was diagnosed with eosinophilic cellulitis (Wells syndrome). We started the patient on Prednisolone 10 mg OD x 2 weeks, then stopped after clearance of all lesions but recurred again, so he restarted again on Prednisolone 10 mg once daily for 2 weeks then tapered to 5 mg once daily for 2 weeks then stopped with the disappearance of all skin lesions. No new recurrence until the time of this case report.

## DISCUSSION

Wells syndrome was first described by George Wells in 1971 as “granulomatous dermatitis with eosinophilia”. The term “eosinophilic cellulitis” was established later.<sup>4</sup> Over 100 cases have been reported to date with varying ages range, the exact etiology is unknown, however, various possible triggers have been reported, including drugs such as penicillin or infliximab, solid and hematological malignancies, arthropod bites, vaccinations, varicella, and parvovirus infections.<sup>1</sup>

Clinical presentation of eosinophilic cellulitis had broad and variable presentation, it commences initially with the prodromal phase of mild tender localized or diffuse cutaneous erythematous plaques with burning or pruritic sensation associated with cutaneous edema. After weeks later, these lesions progressively disappear that can result in some cases atrophic, annular-like plaques.<sup>6</sup> Systemic symptoms frequently present such as fever, malaise, headache, arthralgia, and myalgia.<sup>1</sup>

Histopathology varies according to the phase of infiltration; the acute stage is characterized by marked dermal eosinophilic infiltration surrounding by edema in papillary dermis. Later in the subacute stage may exhibit small groups of eosinophils with their cellular debris collect around collagen bundles producing “flame figures”. Over the course of 1–3 weeks in resolution stage gradual disappearance of the eosinophils occur, leading to typical flame figures to develop.<sup>6</sup> However, half of the patients, their skin manifestations are accompanied by peripheral blood eosinophilia.<sup>7</sup>

Proposed diagnostic criteria for eosinophilic cellulitis by Heelan et al in 2013 require 2 major criteria: clinical picture include any reported variants (plaque-like, urticaria-like, papulovesicular, bullous, papulonodular, granuloma annulare-like and fixed drug eruption-like), relapsing remitting course, no evidence of systemic

disease, or histologic eosinophilic infiltrate with no vasculitis. In addition to at least 1 minor criteria: flame figures histology, granulomatous change, peripheral eosinophilia not persistent and not greater than >1500/ $\mu$ l or triggering factors.<sup>8</sup>

Eosinophilic cellulitis is successfully treated with topical and/or systemic corticosteroids with a 92% success rate, the initial dose of 2 mg/kg over 1–2 weeks, followed by tapering over 2–3 weeks. Attempt of dapsone, cyclosporine, as well as antihistamines can be made if corticosteroids are not sufficiently effective or long-term treatment is required.<sup>9</sup> Recently, successful use of mepolizumab (anti-IL-5 antibody) has been reported.<sup>6,7</sup>

However, eosinophilic cellulitis prognosis is excellent, lesions tend to heal without scarring and resolve in weeks or months, it may heal with slight hyperpigmentation resembling morphea, and patients have a high probability of recurrence (56%).<sup>6</sup>

## CONCLUSION

Eosinophilic cellulitis (Wells' syndrome) is inflammatory dermatitis that is often misdiagnosed due to its similarity with other presentation which leads to delay of correct treatment and inappropriate use of medication. To our knowledge, eosinophilic cellulitis should be kept in mind as part of the differential diagnosis and rising suspicion of any atypical presentation of skin inflammatory disorder that not responsive to appropriate treatment. Correlation of clinical features and histopathological examination of a skin biopsy is necessary to obtain a definitive diagnosis. That the most recommended treatment for eosinophilic cellulitis is an oral steroid. However, the prognosis for eosinophilic cellulitis patients is excellent. It tends to resolve in weeks or months, usually without scarring or with slight hyperpigmentation.

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