

Single Case

## Wells' Syndrome Successfully Treated with Colchicine

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### Keywords

Wells' syndrome · Eosinophilic cellulitis · Colchicine

### Abstract

Eosinophilic cellulitis is an uncommon, inflammatory and chronic disorder of unknown etiology. Corticosteroids are currently considered as the first-line treatment but they are not without significant disadvantages such as contraindications in steroid-resistant cases and patients with frequent recurrences. We report a patient suffering from Wells' syndrome with a 24-year history of symptomatic and generalized skin lesions. After consultation in our department, treatment with colchicine 1 mg/day was prescribed resulting in large clinical improvement. No side effects have been recorded. To our knowledge, this is an original disease approach. Although small, our clinical experience supports the inclusion of colchicine in the drug armamentarium when treating patients suffering from Wells' syndrome. Indeed, its excellent safety profile makes it very attractive for patients with frequent recurrent episodes who need secure options for the medium- and long-term disease control.

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## Introduction

Wells' syndrome is a rare and idiopathic entity whose treatment can be challenging [1, 2]. We report the case of a woman with bullous eosinophilic cellulitis who was successfully treated with colchicine 1 mg/day.

## Case Report

An otherwise healthy 44-year-old woman presented with a 24-year history of recurrent pruritic lesions located on her limbs and face. Skin lesions lasted over 24 h and resolved without scarring. No triggers were identified. The patient had been repeatedly treated with systemic corticosteroids and antihistamines with bad disease control. Physical examination showed erythematous, edematous and blushing plaques partially covered by tense serous bullae. Occasionally, edema of lips and eyelids was present (Fig. 1). Laboratory studies showed no abnormalities except ANA+ (speckled pattern) and ASMA+ with levels of 1/160 and 1/80, respectively (normal ranges <1/40).

There were no findings in radiological images and prick tests were negative to common allergens. Direct and indirect immunofluorescences were both negative. Skin biopsies revealed edema in the upper dermis with subepidermal vesicles and a lymphocytic infiltrate with degranulated eosinophilic material forming flame figures (Fig. 2, Fig. 3).

Wells' syndrome was diagnosed. Colchicine 1 mg/day was prescribed and the lesions completely resolved after 4 weeks. Thenceforth, the patient has been treated with colchicine 1 mg/day for over 24 months. Since the introduction of daily colchicine, an excellent disease control has been achieved: recurrences have been very rare, involving a very limited body surface area, never developing into vesicles or bullae and healing rapidly. Moreover, no additional therapies, such as systemic steroids, were needed and no side effects have been recorded. At present, the patient continues with routine outpatient clinical follow-up.

## Discussion

Eosinophilic cellulitis is an uncommon, inflammatory and recurrent disorder which is more frequently observed in adults than in children [3, 4]. Despite its etiology remaining unknown, hypersensitive responses to different stimuli such as insect bites, fungal and viral infections, medications (including TNF inhibitors), vaccinations, eczema and hematologic malignant disorders, among others [3, 4], have been proposed.

It is clinically characterized by erythematous, painful plaques on the trunk and upper limbs that resolve without scarring [5, 6]. These lesions are usually preceded by itching and evolve developing vesicles or bullae. Wells' syndrome is often misdiagnosed as bacterial cellulitis and should be considered when encountering a case of cellulitis with an atypical presentation and a lack of response to appropriate antibiotic treatment [1, 6].

Diagnosis is based on clinicopathological findings. Histologic features include dermal edema, lymphocytic infiltrate with numerous eosinophils, body giant cells and histiocytes around collagen fibers [7]. Degranulated eosinophilic material can form flame figures which are not pathognomonic and can also be found in insect bites, bullous pemphigoid, eczema, and drug reactions, among others [5].

To our knowledge, all treatment options available are related to case reports or to small case series. To date, the use of colchicine is only mentioned in one case by Paquet et al. [8], who reported on two cases of Wells' syndrome. Systemic and topical corticosteroids are considered as a first-line treatment option, but they are not without well-known significant disadvantages in the medium and long term [2]. Although colchicine mechanisms of action are still unclear, effects such as inhibition of neutrophil chemotaxis, adhesion, mobilization, and superoxide production in addition to inhibition of macrophage inflammasome activation have been described. As in other autoinflammatory diseases, colchicine inhibition of microtubule polymerization and altered expression of adhesion molecules and chemotactic factors could explain adequate clinical response in our case owing to its anti-inflammatory effect [9, 10].

In conclusion, we present a patient with Wells' syndrome successfully treated with colchicine 1 mg/day with very good clinical response and no side effects. Despite being limited [2, 8], our clinical experience supports colchicine 1 mg/day as an alternative therapeutic both on acute onset and as maintenance treatment due to its excellent safety profile.

### Statement of Ethics

The subject of this case report has given his informed consent.

### Disclosure Statement

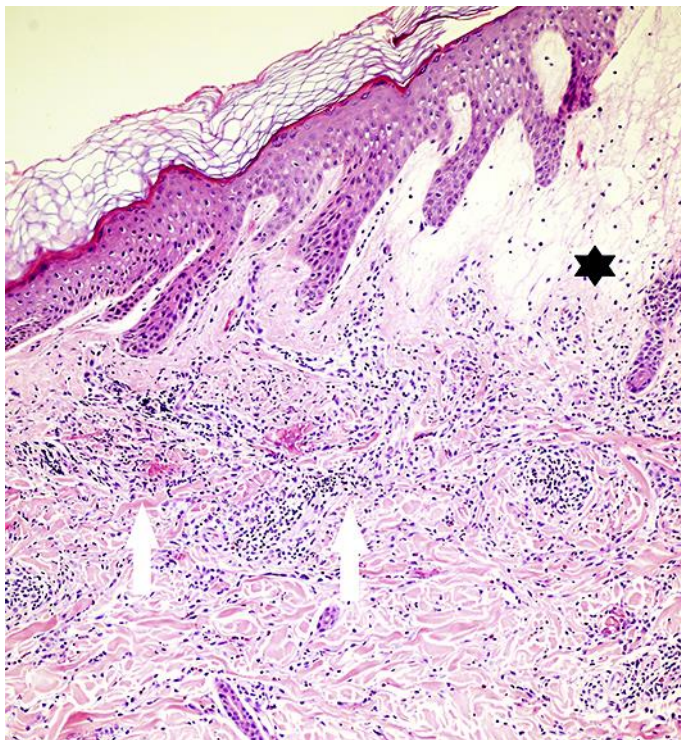
The authors have no conflicts of interest to disclose.

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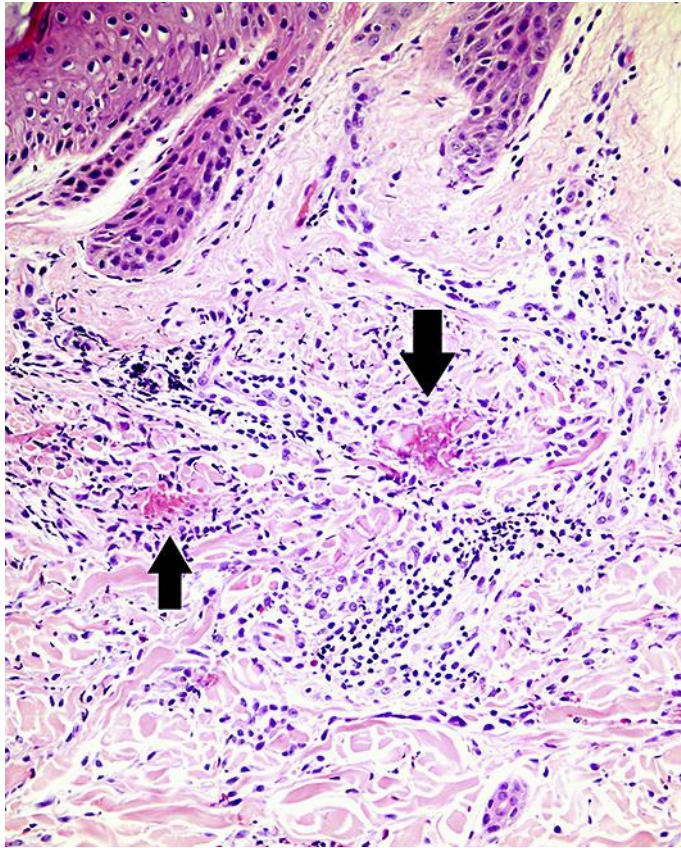


**Fig. 1.** Erythematous, edematous and blushing plaques partially covered by tense serous bullae.



**Fig. 2.** Dermal edema with intraepidermal vesicles (asterisk) and a lymphocytic infiltrate with numerous eosinophils (white arrows). HE. ×20.





**Fig. 3.** At higher magnification. Degranulated eosinophils forming flame figures (black arrows). HE.  $\times 50$ .