

## BRIEF ARTICLE

## Wells Syndrome with Sustained Response to Omalizumab

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## ABSTRACT

Wells syndrome (eosinophilic cellulitis), a rare dermatosis that arises from a multitude of triggers, may present with pruritic and erythematous plaques, papules, vesicles, or blisters.<sup>1</sup> We present a case of Wells syndrome that was successfully treated with omalizumab. A 39-year-old female presented with a three-year history of diffusely located pruritic papules. After a negative lab and infectious workup, a punch biopsy showed dense eosinophilic infiltrate in the dermis as well as “flame figure” formation. Unsuccessful trials of prednisone, cetirizine, hydroxyzine, and dapsone prompted the use of omalizumab, which yielded excellent control of symptoms. Omalizumab’s success in the Wells syndrome disease process may be explained by its anti-inflammatory effects on IgE autoantibodies and receptors.<sup>5</sup> Few cases of Wells syndrome treated with omalizumab have been reported. Thus, increased research to thoroughly elucidate omalizumab’s pharmacological mechanism of action in this particular disease process is warranted and would be a valuable contribution to the Wells syndrome literature.

## INTRODUCTION

Wells syndrome (eosinophilic cellulitis) is a rare dermatosis characterized by pruritic and erythematous plaques, papules, vesicles, or blisters that commonly appear on the extremities. Autoimmunity, insect bites, drugs, contact allergens, infections, and photosensitivity are triggers that have previously been associated with this diagnosis.<sup>1</sup> Treatment options for Wells syndrome include corticosteroids, antihistamines, antibiotics, immunosuppressants, and biologics.<sup>2</sup> We present a case of Wells syndrome that was unresponsive to corticosteroids, antivirals, antibiotics, antihistamines, and dapsone. This case reflects one of the few reported instances of Wells syndrome that was successfully treated with omalizumab.

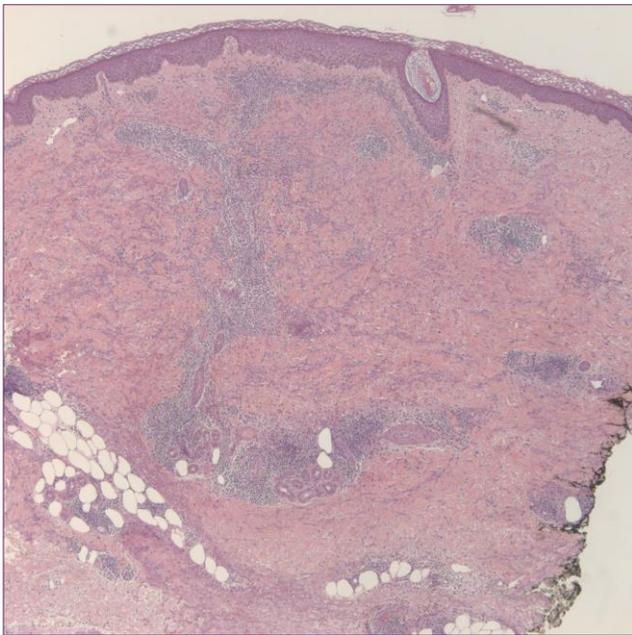
## CASE REPORT

A 39-year-old female with no significant past medical history presented with a three-year history of small pruritic papules located on the face, extremities, and genitals (Figure 1).



Figure 1.

Further review of systems was negative. Prior to initial presentation, she trialed valacyclovir, clobetasol, mupirocin, and loratadine without improvement. Previous labs showed negative ANA and a normal eosinophil count. At the first visit, she was prescribed betamethasone ointment and an increased dose frequency of loratadine from once to twice a day. A scraping for scabies was negative, which prompted a punch biopsy of the left lower arm. Biopsy sections showed a dense infiltrate in the dermis with abundant eosinophils (Figure 2)

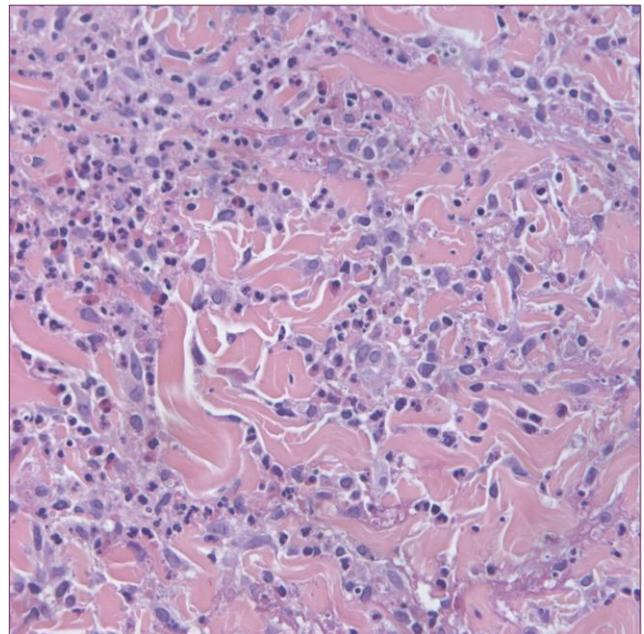


**Figure 2.**

as well as characteristic “flame figure” formation (Figure 3) that supported a diagnosis of Wells syndrome.

At follow up, the patient noted progression of her symptoms with a new plaque on the right hip and new pruritic papules arising on a weekly basis. She was started on prednisone 60 mg daily with a month-long taper, daily cetirizine and nightly hydroxyzine. Although her skin cleared with

the medications, her symptoms flared once the prednisone dose was tapered to 10 mg.



**Figure 3.**

She was then started on dapsone 50 mg daily, which decreased the tenderness of her lesions and the frequency of her flares.

Nine months after taking dapsone, the patient reported decreased grip strength in her right hand, which affected her work as an aesthetician. To avoid worsening of musculoskeletal sequelae, dapsone was discontinued and she was referred to neurology. The patient was then started on omalizumab 150 mg subcutaneously every 28 days.

The patient tolerated the omalizumab injections well, reporting no more than one flare of her skin in between injections and an overall improvement without any reported side effects. This patient has been on omalizumab for over 9 months now with excellent control of her symptoms.

**Table 1.** Summary of Wells syndrome cases successfully treated with Omalizumab. There are seven reported cases to date, including the present case report. Data from this table are from Egeland et al<sup>2</sup>, Ogueta et al<sup>3</sup>, and Coattre nec et al<sup>6</sup>.

Case Report	Age	Sex	Presenting Symptoms	Mean Time to Presentation	Medications Tried	Omalizumab Regimen	Mean Time to Resolution	Treatment Length
Egeland et al	45	Female	Painful and pruritic deep erythematous noduli and plaques (several ulcerated) on ears, back, hands, and scalp	10 years	Antihistamines Prednisolone Cyclosporine Dapsone Colchicine	300 mg every 4 weeks	3 months	13 months
Ogueta et al	71	Male	Acute cutaneous eruption of pruritic and painful erythematous and edematous plaques	15 years	Bilastine 80 mg Prednisone Colchicine Dapsone Cyclosporine	300 mg every 4 weeks	6 weeks	Asymptomatic after 20 months
Ogueta et al	62	Female	Painful erythematous and indurated plaques on trunk and legs	20 years	Bilastine 40 mg	300 mg every 4 weeks	8 weeks	Asymptomatic after 18 months
Ogueta et al	71	Female	Persistent plaques on trunk and extremities, and hyperpigmented macules	1 year	Bilastine 80 mg Prednisone 30 mg	300 mg every 3 weeks + Bilastine 60 mg daily, then 300 mg every 4 weeks + Bilastine 20 mg daily	16 weeks	Free of lesions on Omalizumab 300 mg every 4 weeks + Bilastine 20 mg daily
Ogueta et al	61	Female	Deep pruritic and hot erythematous plaques	15 years	Prednisone 30 mg Deflazacort 30 mg Rupatadine 20 mg Dapsone 100 mg	300 mg every 4 weeks, then 300 mg every 6 weeks	1 week	Asymptomatic after 1 year
Coattre nec et al	67	Male	Erythema and occasional edema of the limbs, tongue, and face	30 years	Antihistamines Corticosteroids (topical and systemic) Azathioprine 150 mg Tranexamic Acid 1000 mg	300 mg every 4 weeks, then 300 mg every 8 weeks	Dramatic improvement after a few days	Asymptomatic after 24 months
Surjanto et al	39	Female	Pruritic papules on face, extremities, and genitals Plaque on hip	3 years	Valacyclovir Clobetasol Mupirocin Loratadine Prednisone 60 mg, tapered to 10 mg Cetirizine Hydroxyzine Dapsone 50 mg	150 mg every 28 days	8 weeks	Asymptomatic for over 9 months

## DISCUSSION

Wells syndrome is an uncommon dermatosis believed to be a type IV hypersensitivity reaction.<sup>2,3</sup> Although many variants of Wells syndrome have been observed, characteristics such as pruritic and tender erythematous plaques on gross observation, along with dermal infiltrate of eosinophils with “flame figures” on histology, tend to be suggestive of the diagnosis.<sup>2,3</sup> Said “flame figures” represent aggregates of major basic protein released from eosinophils which disintegrate the structural integrity of surrounding collagen and subsequently destroy local tissue.<sup>1</sup> In addition to characteristic histological features, Wells syndrome criteria that were seen in our patient also include a recurrent course of cutaneous manifestations, no evidence of triggering factors, and inconsistent blood eosinophilia.<sup>4</sup>

Previously reported treatments for Wells syndrome are vast, ranging from corticosteroids to biologics.<sup>2</sup> However, there are only a handful of papers that report the use and efficacy of omalizumab. Omalizumab is a monoclonal anti-immunoglobulin E (IgE) antibody that was first approved for the treatment of persistent allergic asthma, then later for the treatment of refractory chronic spontaneous urticaria.<sup>5</sup> The current knowledge of omalizumab on cutaneous inflammatory diseases is that the drug decreases IgE levels, IgE receptors, mast cells’ ability to release contained substances, and the action of IgE autoantibodies against autoantigens.<sup>5</sup> Despite ongoing speculation on the drug’s anti-inflammatory pathways, treatment of Wells syndrome with omalizumab has been shown to be effective.<sup>2,3,6</sup>

## CONCLUSION

To date, our case is the seventh case of Wells syndrome that was successfully treated with omalizumab (Table 1). Although it is imperative that more research be done to elucidate omalizumab’s pharmacological mechanism of action on the specific pathology of Wells syndrome, omalizumab may be an effective treatment option for refractory cases of this disease. This case report aims to highlight another instance of omalizumab’s success to add to the Wells syndrome literature.

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