



Unusual Presentation of Wells Syndrome: A Case Report

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ABSTRACT

Wells syndrome is an uncommon disease that typically presents as edematous erythematous plaques, usually preceded by burning or itching of the skin. Histopathological examination shows dense dermal eosinophilic infiltrates in an edematous dermis at the acute phase of lesions. Some of the identified triggering factors include infection, arthropod bites, hematological malignancies, thimerosal containing vaccines and drugs such as penicillin, lincomycin, tetracycline, minocycline and ampicillin. Here we describe a case of Wells syndrome in a 75-year-old woman that its outstanding feature was its large size. Although this case was resistant to our treatment, the condition improved spontaneously after several weeks without administering any other alternative treatments. On the other hand, despite its large size, this case had no identifiable trigger.

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Introduction

Wells syndrome is an uncommon inflammatory disease, initially described in 1971 by GC Wells (1). It is characterized by pruritic annular erythematous plaques with infiltrated borders which mainly affects the limbs or trunk, and has a relapsing and remitting course (2-6).

Typical lesions include a wide spectrum from a mild form of circinate red plaques to more severe forms such as blistering or a sudden eruption of cellulitis-like lesions, sometimes associated with burning or itching sensation. It may persist over months to years but usually resolves spontaneously without scarring (2, 3, 6, 7).

Wells syndrome usually does not have any systemic involvement, but peripheral blood eosinophilia is a common feature (5, 8). There are various therapeutic options, but low-dose oral corticosteroids are the first-line treatment in Wells syndrome (5, 9).

We describe a case of Wells syndrome in a 75-year-old woman that its outstanding feature was its large size; although this case was resistant to our treatment, the condition improved spontaneously after several weeks without administering other treatments. On the other hand, this case had no identifiable trigger.

Case Report

A 75-year-old Iranian woman presented with 1-month history of itching, and an infiltrating erythematous plaque on abdomen which initiated from periumbilical region but extended in size and spread bilaterally to the flanks.

It was the first episode. Having no similar or recurrent lesions before, she denied any systemic or cutaneous associated symptoms other than itching of the lesion. There was no history of any drug intake, insect bites or any known food or other substances as a trigger for the condition. Her medical history, family history and drug history were all negative.

Physical examination revealed an erythematous-violaceous, well-defined, annular, indurated plaque affecting abdomen skin, approximately 80 cm x 35 cm in size without any vesicles or bullae on the surface. The indurated plaque had a peau d'orange appearance. In addition, it was neither tender on palpation nor scaly on observation (Figure 1). General physical examination was completely normal. The patient was afebrile.

Laboratory results were unremarkable, except mild serum eosinophilia (6%, 384 mm^3). Complete blood count (CBC), erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), and other routine biochemical tests were within normal limits. A work up for possible underlying malignancy was negative, and there was no associated lymphadenopathy.



Figure 1. Erythematous, well-defined, annular, indurated plaque affecting abdomen and flank skin, approximately 80 cm x 35 cm in size.

A punch biopsy was taken with differential diagnosis of eosinophilic cellulitis, inflammatory morphea, granuloma annular, sarcoidosis, and interstitial granulomatous disease.

The biopsy of the plaque revealed a patchy infiltration of lymphocytes and eosinophils extending from superficial to deep dermis. There was also mild vasculopathy and scattered flame figures, which represented eosinophilic granules encrusted on dermal collagen (Figure 2). These findings were consistent with the diagnosis of Wells syndrome.

After two weeks, the centrifugal expansion of plaque was obvious by its discrete, raised, infiltrative, sharply red border along with the fading hyperpigmented center.

Prednisolone was initiated at a dose of 40 mg daily (0.5 mg/kg), betamethasone valerate cream along with emollient eucerin was applied topically, and oral cetirizine was administered for the pruritus.

However, the patient discontinued the therapy after 2 weeks since there was no improvement of the skin lesions.

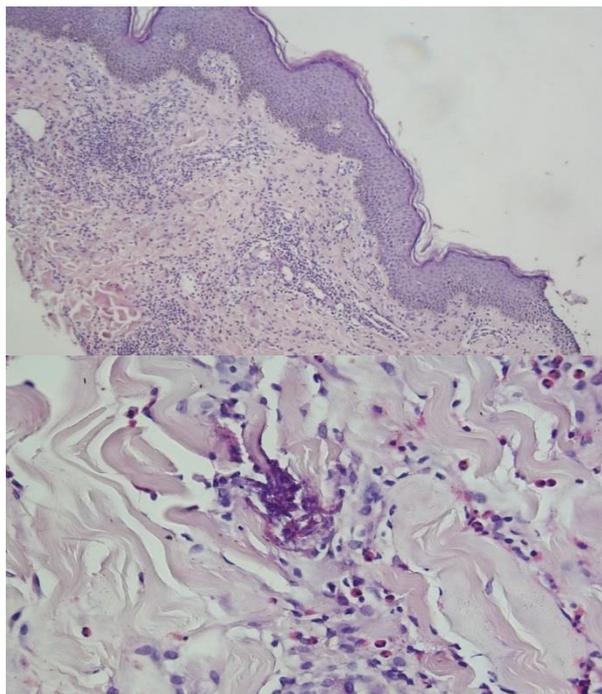


Figure 2. Patchy infiltration of lymphocytes and eosinophils extending from superficial to deep dermis

Although there was no additional medical intervention, the lesions began to clear spontaneously after three weeks without scarring or dyspigmentation.

Discussion

Wells syndrome (eosinophilic cellulitis) is a rare condition of unknown etiology (4, 5, 10). It typically presents as erythematous patches or non-scaly edematous plaques, usually preceded by burning or itching of the skin. The severity of the symptoms varies from a mild to severe pruritus as well as tenderness of the lesions (4, 6, 9). There are also various manifestations reported in the literature including vesicles, bullae, granuloma-like multiple arcuate erythematous plaques, as well as widespread papulonodular or even fixed drug eruption-like lesions (2, 4, 5, 9).

The typical course of disease often includes a central involution initially, and then turning from a red-brown color to a grey-blue color while becoming indurated in later stages, frequently the lesions involute over a period of two to eight weeks, and finally the skin clears without scarring. However, it sometimes results

in morphea-like atrophic hypopigmented patches (4, 6, 8). It occasionally may be associated with systemic involvement such as fever, arthralgia, lymphadenopathy, and rarely anterior uveitis (10).

Although the etiology of the disease remains unclear, a type IV hypersensitivity reaction and abnormal eosinophilic response to several exogenous and endogenous causative agents is considered to be the main underlying pathophysiology of eosinophilic cellulitis (4, 10). Some of the aforementioned triggering factors include infection, arthropod bites, hematological malignancies, thimerosal containing vaccines and drugs such as penicillin, lincomycin, tetracycline, minocycline, and ampicillin (5, 9, 10).

Histopathologically, there are three stages: first, acute lesions show dense dermal eosinophilic infiltrates in an edematous dermis; second, subacute lesions characterized by giant cells, histiocytic, eosinophils and flame figures in the dermis; and the last stage with micro granulomas consisting of foreign body type giant cells (8, 10).

Here, we report a case of idiopathic Wells syndrome in a 75-year-old woman, who presented with an exceptionally large size pruritic, erythematous and annular plaque on her abdomen extending to her flanks. Despite the large size of the lesion that was not previously reported in other literature, there was neither associated systemic involvement nor triggering factors. Furthermore, considering the age of our patient and the fact that there are some association between Wells syndrome and solid malignancies (8), we performed a complete malignancy work-up for the patient which revealed negative results.

The most confusing clinical differential diagnosis of Wells syndrome is acute infectious cellulitis. Other clinical differential diagnoses include erythema chronicum migrans, arthropod bites, hypereosinophilic syndrome, granuloma annular, and chronic idiopathic urticaria (3, 8).

Although, flame figures are not pathognomonic for Wells syndrome, and may

be detected in other dermatoses such as bullous pemphigoid, tineapedis, bite reactions, severe prurigo, eczema and follicular mucinoses (3, 10), we regarded this case as an eosinophilic cellulitis because its histopathologic features together with the clinical presentation as well as peripheral blood eosinophilia and negative result of drug history, bite reaction and malignancy work-up were all consistent with Wells syndrome rather than other diseases in the differential diagnosis of flame figure.

Treatment options include topical corticosteroids, antihistamines, antimicrobial agents, dapsone, griseofulvin, colchicine, minocycline, nicotinamide, cyclosporine, azathioprine, interferon alpha, ultraviolet light, and systemic corticosteroids, which is usually considered as the first-line treatment in Wells syndrome; but many cases resolve spontaneously without therapy (5, 8), as what actually happened in our case who did not respond to topical and systemic corticosteroid while the lesion began to disappear after a period of three weeks following discontinuation of prednisolone.

To our knowledge, this is the first case of Wells syndrome with such a large size of single lesion (80 cm x 35 cm) extending from abdomen to flanks bilaterally. Interestingly, despite the large size, we did not find any associated systemic symptom or underlying disease, and the lesion did not respond to topical and systemic corticosteroid.

Conflict of Interests

Authors have no conflict of interests.

Acknowledgments

None.

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