Case Report

Two Cases of Wells Syndrome with Marked Swelling in the Hands

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Abstract

Wells syndrome or eosinophilic cellulitis is an idiopathic inflammatory dermatitis characterized by a benign but recurrent evolution. It often causes edematous urticarial plaques on the extremities. Herein, we report two rare cases of Wells syndrome with marked swelling in the hands. Both cases exhibited various clinical features. Case 1 was Wells syndrome with collagen disease-like sclerotic edema in the fingers and annular erythema on the trunk. Case 2 was Wells syndrome with diffuse plate-like hardening of the forearm mimicking cellulitis, which required fasciotomy due to suspected compartment syndrome at the emergency room. Wells syndrome should be included in the differential diagnosis of patients presenting with marked diffuse swelling in the hands.

Keywords: *Eosinophilic cellulitis, hands swelling, Wells syndrome*

Introduction

Wells syndrome is a relapsing eosinophilic dermatitis with variable clinical appearance. Due to symptoms of erythema and swelling of the limbs, it is often misdiagnosed as cellulitis or contact dermatitis. However, cases of Wells syndrome with extending swelling from the arms to hands are rare. Herein, we describe two cases of Wells syndrome with marked swelling in the hands.

Case Reports

Case 1

A 61-year-old woman with hemorrhoids and arterial hypertension presented to our hospital with painful swelling, erythema, purple spots, and blisters on both hands, from fingers to wrists. Hemorrhagic plaques were observed in the palms. She had difficulty in bending her fingers and had finger sclerotic edema, which exhibited sausage-like swelling [Figure 1a and b]. In addition, her trunk and extremities were erythematous with itching. The initial clinical diagnosis made by a local hospital had been contact dermatitis, and she was treated with oral prednisolone (10 mg/day), which improved her symptoms. However, she was referred to our hospital because of recurrence. Laboratory examination revealed an elevated C-reactive protein level (1.57 mg/dL), although complete blood count, liver function test, renal function tests and serum complements were normal. A biopsy specimen taken from the right thumb revealed inflammatory infiltrate of eosinophils in all layers of the dermis, and flame figures were observed in the dermis [Figure 2a and b]. There was no vasculitis. The patient was diagnosed with Wells syndrome and was successfully treated with betamethasone butyrate propionate ointment and olopatadine hydrochloride tablets.

Case 2

A 13-year-old boy presented with swelling and pain on his left arm. He had noticed insect bites on his left palm 10 days before the presentation. At an internal medicine hospital, he was suspected to have cellulitis and underwent treatment for it with oral antibiotics for 3 days. However, the swelling further extended to his left fingers and shoulder, and purpuric lesions were scattered on the flexion aspect of the forearm [Figure 3]. The patient was, therefore, admitted to our hospital with suspected necrotizing fasciitis. His white blood cell count was within normal limits, but his blood eosinophil count was elevated (33%). The compartmental pressure of his left forearm was elevated (50–60 mmHg), and the patient was diagnosed as having compartment

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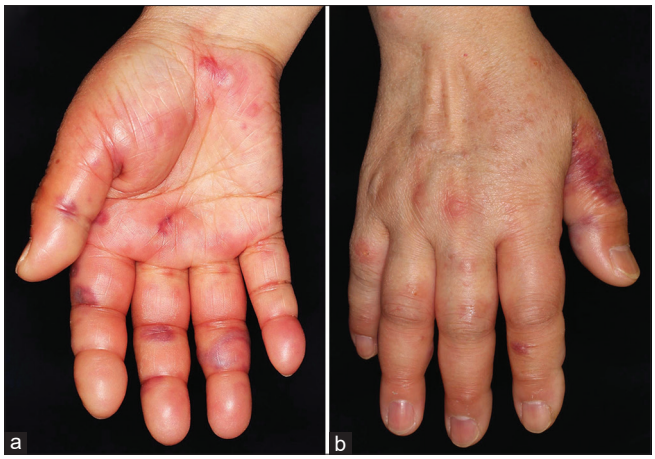

Figure 1: Clinical features of initial skin lesions. The fingers showed erythema and swelling. Some blisters and purpura were observed (a and b)

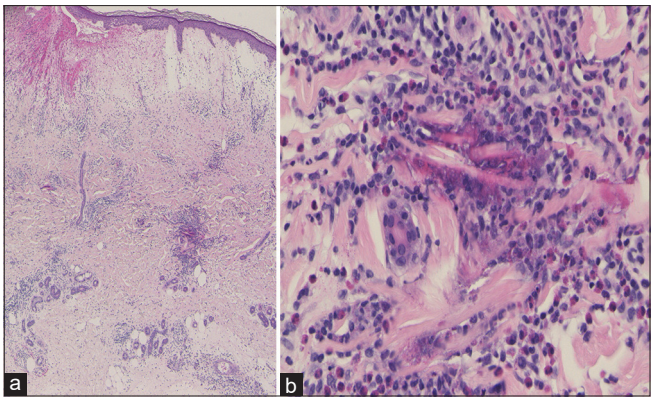

Figure 2: The dermis showed edema and inflammatory cell infiltration (hematoxylin-eosin: [a] ×40). Degeneration of collagen fibers and degranulation of eosinophils were observed; flame figures (hematoxylin-eosin: [b] ×200)

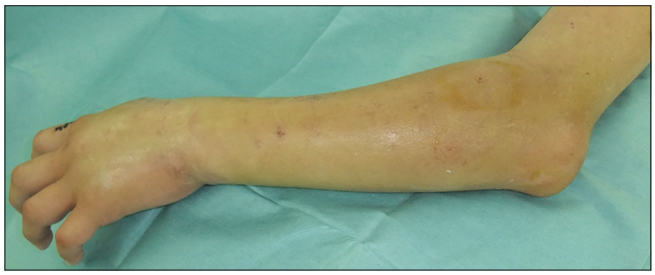

Figure 3: The patient's hands were swollen from his fingers to shoulders. Some blisters and erythema were observed

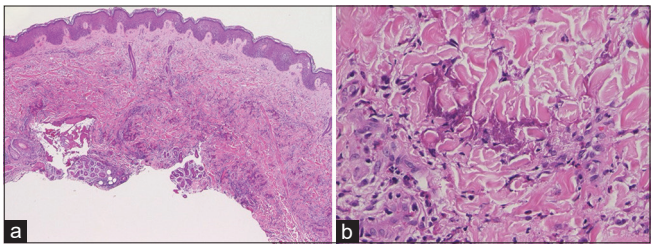

Figure 4: Infiltration of inflammatory cells was observed in the entire dermis (hematoxylin-eosin: [a] ×40), and there was an infiltration of lymphocytes and eosinophils. Some eosinophil degranulation was also observed (hematoxylin-eosin: [b] ×200)

syndrome. He underwent fasciotomy, and no necrosis was observed from the subcutaneous tissue to the muscle tissue. A biopsy specimen taken from the forearm revealed inflammatory infiltrate of eosinophils in all layers of the dermis, as well as flame figures [Figure 4a and b], and all bacterial cultures were sterile. He was successfully treated with prednisolone (25 mg/day), which was gradually tapered within 3 months.

Discussion

Wells syndrome affects both adults and children,[1] and presents with tender urticarial erythematous plaques, vesicles, bullae, or nodules.[2] To date, only six cases of Wells syndrome with marked swelling in the hands have been reported.[3-8] The patient characteristics of those cases, along with the present cases, are shown in Table 1. All patients had lesions on one side, and three patients had pruritic erythema on the trunk and extremities. All cases were successfully treated with topical and systemic corticosteroids, but two patients experienced recurrence. The usual course of the condition is spontaneous remission. In Case 1, spontaneously resolution may be considered. Both of our cases developed hemorrhagic plaques or purpuric lesions. Ratzinger *et al*.[9] suggested that eosinophilic leukocytoclastic vasculitis might form a bridge between Wells syndrome and Churg–Strauss syndrome. Eosinophilic leukocytoclastic vasculitis has a mild course and can be cured after an episode, while patients with Churg–Strauss syndrome have multiple flare-ups. Whether Wells syndrome is to be classified as vasculitis or not is still controversial; and in our cases, vasculitis was not observed histologically.

Case 1 additionally developed annular erythema on the right lower extremity, which may require the differentiation with eosinophilic annular erythema (EAE). In the original paper by Wells *et al*.,[10] various clinical features of Wells syndrome were reported, including cellulitis-like erythema with or without some blistering and annular or circinate erythematous plaques. Thereafter, several cases of Wells syndrome with an annular erythematous lesion have been reported. There are a number of inflammatory skin diseases presenting with annular lesions, and Wells syndrome should be included in such disorders. Some authors view EAE as a variant of Wells syndrome, while other authors believe that slight differences can make this differential diagnosis possible.[11,12] EAE histologically lacks flame figures and degranulation of eosinophils, whereas perivascular infiltration of mononuclear cells and eosinophils are observed in the superficial and deep dermis. Therefore, whether EAE is a distinct entity or not depends on further studies. Eosinophil-derived proteins, that is, major basic proteins play roles in allergic inflammation, tissue damage

Table 1: Six cases of Wells syndrome with hand swelling

Case	Age•Sex	Medical history	Lesion	Trigger	First diagnosis	Therapy	Recurrence
1. Gander et al.[3]	48•M	No	Left mid-humerus to finger tips	Unknown	Necrotizing fasciitis	Antibiotics Debridement Oral prednisone	no
2.Shodo et al. [4]	67•M	Pneumoconiosis	Right hand and forearm, neck, trunk, and thighs	Touching a potted plant	Bacterial cellulitis and contact dermatitis	Antibiotics Oral steroid	no
3. Lin et al. [5]	45•F	No	Right hand and forearm	Stung by a honeybee	Bacterial cellulitis	Antibiotics Methylprednisolone	no
4. Sakara et al. [6]	22•F	No	Right hand	Ulcerative colitis	Wells' syndrome	Oral prednisone	once
5. Yoshii et al. [7]	45•M	No	Hands and upper arm, feet and trunk	Coxsackie virus A6 infection	Wells' syndrome or non-episodic type of angioedema	Oral prednisolone	no
6 Brain et al. [8]	31•F	No	Right extremity	Unknown	Wells' syndrome	Systemic steroids Antihistamines	no
Present case	60•F	Hemorrhoid, hypertension	Hands Trunk, extremities	Unknown	Contact dermatitis	Topical steroid Antihistamine	no or once
Present case	13•M	No	Left hand and forearm	Insect bites	Necrotizing fasciitis	Antibiotics Debridement Oral prednisolone	no

and repair, remodeling, and fibrosis. The secretion of eosinophil proteins may play a role in the induction of prominent swelling of the hands with transient joint contracture. Wells syndrome should be included in the differential diagnosis of patients presenting with marked diffuse swelling in the acral sites.

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Conflicts of interest

There are no conflicts of interest.

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