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Case Report

Wells Syndrome: A Rare Recurrent Eosinophilic Dermatosis in a Tertiary Care Hospital

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Abstract

Wells syndrome, a rare eosinophilic cutaneous disorder, also called eosinophilic cellulitis, exhibits significant clinical heterogenicity. It typically presents as erythematous infiltrated plaques, primarily on the extremities. Initially, these lesions often exhibit similarities to erysipelas or cellulitis but do not respond to antimicrobial therapy. Contribution of IL-5, abnormal Th2 cells, and activated eosinophilic granulocytes suggests a nonspecific hypersensitivity reaction to exogenous or endogenous stimuli. Wells syndrome is a diagnosis of exclusion that can be recognised by its clinical course, and histological findings. Corticosteroids are the mainstay treatment, helps alleviate the ongoing, relapsing pattern. We are reporting a case of 50 years old male presented to us with complaint of itchy red raised lesions that has recurrent course. On physical examination, multiple erythematous plaques were identified over bilateral lower limb. Skin biopsy specimen was sent for histopathological examination which showed eosinophilic infiltration around peri-appendageal structures confirming the diagnosis of Wells syndrome. It is a rare, relapsing skin disorder of unknown cause which is difficult to diagnose specially on first episode. For mild cases, topical corticosteroids are often sufficient. However, in most of the cases, oral steroids show rapid resolution. Gradual reducing dosage of corticosteroids across a month is usually easily tolerated by the patients whereas low maintenance dose with corticosteroids helps in preventing recurrences.

Keyword

Wells Syndrome, Eosinophilic Cellulitis, Flame Figures

1. Introduction

Wells syndrome (Eosinophilic cellulitis) is a rare inflammatory skin condition characterized by recurrent, itchy and inflamed skin lesions that may resemble infections like cellulitis or erysipelas. Wells published it firstly in 1971 as "recurrent granulomatous dermatitis with eosinophilia" [1]. The disease typically presents with a sudden onset of one or more pruritic, erythematous plaques, most commonly affecting the extremities and resembling infectious cellulitis. However, the clinical presentation can be diverse, with different lesions morphologies appearing simultaneously in the same patients. Lesions may be localized or widespread, commonly occurring on the trunk and limbs but also affecting the face, eyelids, ear, scalp, axillae and groin [2].

Wells syndrome predominantly occurs in adults; however, it can affect individual of all ages, with peak incidence occurring in the second and third decades of life [3, 4]. Eosin-

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ophilic cellulitis (EC) shows no sex predilection in adults, though a male predominance has been reported in children [5]. There is no known association with any specific race or ethnicity. The pathogenesis of Wells syndrome remains unclear and is likely a reactive process triggered by various stimuli rather than a condition with a single definitive cause. It may involve a delayed-type (Type IV) hypersensitivity reaction, similar to allergic contact dermatitis [6, 7]. Eosinophil degranulation leads to the release of eosinophil cationic and major basic protein, which exert a harmful effect on tissue and further stimulating the activation of other immune cells [8, 9].

In histopathology, it may show presence of dermal edema, flame figures and marked dermal eosinophilic infiltrates. Flame figures consist of eosinophil major basic protein which surrounding collagen fibers [10]. An inflammatory infiltrate may also be present in superficial and deep, perivascular region. However, histopathological findings vary depending on the timing of the biopsy. The exact nature of Wells syndrome remains debated, with some experts considering it a distinct disorder characterized by a unique combination of clinical and histopathological features. However, many of its clinical and histological characteristics are not entirely specific [11, 12].

2. Clinical Case

A 50 years old male presented with multiple erythematous, edematous plaques on bilateral lower limb for last 10 days. The plaques were pinkish-red in color initially, that gradually became violaceous and resolved within 2 weeks, resulting slight hyperpigmentation. The differential diagnosis considered included bacterial cellulitis, insect bite hypersensitivity, and Wells syndrome. Past history disclosed multiple (10 to 12) similar episodes over last 20 months. Laboratory investigations, including total leukocyte count and hemoglobin, were within normal limits. The absolute eosinophil count was 450-cells/mm^3 (reference range: 40-440 cells/mm^3). Histopathological examination revealed moderately dense superficial and deep perivascular, as well as peri-appendageal infiltrate composed of eosinophils and lymphocytes. Several eosinophils were scattered in interstitium of reticular dermis and around deep vascular plexus. Based on clinical presentations and histopathological findings, diagnosis of Wells syndrome was made and patient was prescribed low dose oral steroid along with topical corticosteroid. The patient responded well to therapy over 2 weeks, with most lesions resolving and no new lesions developing. A tapering dose of oral steroid was prescribed for the continued course.

3. Discussion

Wells syndrome is a rare skin disorder of uncertain origin, characterized by a benign yet recurrent pattern. In previous reports, patients presented with various morphology including large erythematous plaques, vesicles, bullae, papules, and nodules [2]. Based on these findings, seven clinical subtypes have been mentioned: plaque-type, papulovesicular, bullous, annular granuloma-like, urticaria-like, fixed drug eruption-like, and papulonodular [13]. Cutaneous lesions may precede or occur simultaneously by itching and burning. They are often warm to touch but generally non tender and do not respond to antimicrobial treatment, unlike bacterial cellulitis [2]. In most cases, lesions heal within 2 to 8 weeks without leaving scars [14]. However, in some patients, the condition may persist, with lesions lasting for several months. Even after a full and prolonged remission, cutaneous lesions can reappear in the same location.

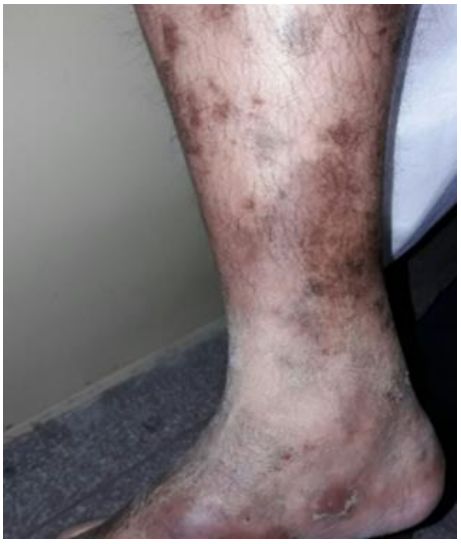

Figure 1. *Multiple erythematous, oedematous plaques involving lower limb with hyperpigmentation on resolving area.*

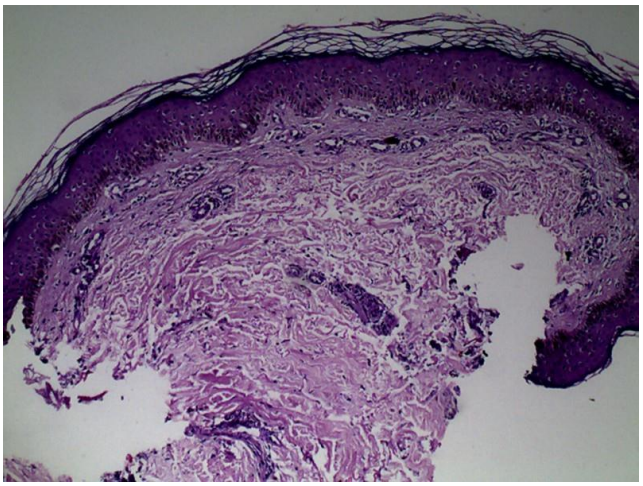

Figure 2. *H&E, x40 Histopathological examination revealed a moderately dense superficial and deep perivascular, as well as peri-appendageal, infiltrate composed of eosinophils and lymphocytes. Additionally, several eosinophils were interspersed throughout the interstitium of reticular dermis and surrounding the deep vascular plexus.*

Wells syndrome is considered as rare dermatological condition that is often difficult to diagnose, with many cases initially diagnosed inaccurately as bacterial cellulitis and treated with antimicrobial therapy. The disease should be assumed in patients with a clinical history of recurrent, pruritic, and cellulitic plaques that are non-tender and unresponsive to antimicrobial medication. A skin biopsy revealing characteristics histopathological findings, such as eosinophilic dermal infiltrate and flame figures, along with a complete blood count showing absolute eosinophilia, supports the clinical diagnosis. For further supporting the diagnosis, a series of clinical and pathologic criteria (consist of Major and minor criteria) has been proposed [12].

Due to the rarity of Wells syndrome, no consensus exists on optimal management, as randomized controlled trials or large prospective studies have not been conducted. Consequently, the available evidence is primarily derived from case reports and small case series [2, 15]. For localized disease, medium to high potency topical corticosteroids are considered the first-line treatment, particularly in children [2, 6]. In cases where localized disease does not respond to topical corticosteroid or when the disease is generalized, a short course of systemic corticosteroid, such as prednisolone (0.5 to 1 mg/kg per day), may be administered for 5 to 7 days [2, 16]. Based on response, the dose of corticosteroids can be tapered off over two to three weeks.

Antihistamines are typically used as adjunctive therapy to manage pruritus but do not impact the skin lesions [2, 17]. In refractory or recurrent cases, other therapies that have been used include dapsone, azathioprine, cyclosporine, colchicine, minocycline, psoralen with ultraviolet A (PUVA) phototherapy, mepolizumab, omalizumab and dupilumab [18-26].

4. Conclusion

Wells syndrome is considered as chronic disorder comprising of recurrent course. Corticosteroids remain cornerstone treatment. Recurrence is a significant concern in both adults and children, especially in patients with involvement of multiple body areas and the spread of lesions beyond the initially affected sites. All patients with Wells syndrome require clinical follow-up to monitor treatment response and detect potential disease recurrence. Surveillance for the development of haematological malignancy is particularly recommended for individuals with chronic comorbidities or those presenting with systemic symptoms.

Abbreviations

IL	Interleukin
Th2	T helper 2
EC	Eosinophilic Cellulitis

Author Contributions

Both authors made significant contribution to the conceptualization and development of this manuscript. There contributions encompassed the initiation, drafting and refinement of the content. Ajay Kumar Vishwakarma contributed to the clinical management of this patient, while Tripti Sharma analysed pathology images.

Conflicts of Interest

The authors declare no conflicts of interest.

References

[1] Wells GC. Recurrent granulomatous dermatitis with eosinophilia. Trans St Johns Hosp Dermatol Soc 1971; 57: 46-6. PMID: 5570262.

[2] Sinno H, Lacroix JP, Lee J, et al. Diagnosis and management of eosinophilic cellulitis (Wells' syndrome): A case series and literature review. Can J Plast Surg 2012; 20: 91-7. https://doi.org/10.1177/229255031202000204

[3] Chan JK, Hui PK, Ng CS, et al. Epithelioid haemangioma (angiolymphoid hyperplasia with eosinophilia) and Kimura's disease in Chinese. Histopathology 1989; 15: 557-74. https://doi.org/10.1111/j.1365-2559.1989.tb01622.x

[4] Hui PK, Chan JK, Ng CS, et al. Lymphadenopathy of Kimura's disease. Am J Surg Pathol 1989; 13: 177-86. https://doi.org/10.1097/00000478-198903000-00001

[5] Reichel M, Isseroff RR, Vogt PJ, Gandour-Edwards R. Wells' syndrome in children: varicella infection as a precipitating event. Br J Dermatol 1991; 124: 187-90. https://doi.org/10.1111/j.1365-2133.1991.tb00431.x

[6] Koh KJ, Warren L, Moore L, et al. Wells' syndrome following thiomersal-containing vaccinations. Australas J Dermatol 2003; 44: 199-202. https://doi.org/10.1046/j.1440-0960.2003.00678.x

[7] Nacaroglu HT, Celegen M, Karkıner CS, et al. Eosinophilic cellulitis (Wells' syndrome) caused by a temporary henna tattoo. Postepy Dermatol Alergol 2014; 31: 322-4. https://doi.org/10.5114/pdia.2014.40951

[8] Rothenberg ME, Hogan SP. The eosinophil. Annu Rev Immunol 2006; 24: 147-74. https://doi.org/10.1146/annurev.immunol.24.021605.090720

[9] Slifman NR, Loegering DA, McKean DJ, Gleich GJ. Ribonuclease activity associated with human eosinophil-derived neurotoxin and eosinophil cationic protein. J Immunol 1986; 137: 2913-7.

[10] Peters MS, Schroeter AL, Gleich GJ. Immunofluorescence identification of eosinophil granule major basic protein in the flame figures of Wells' syndrome. Br J Dermatol 1983; 109: 141-8. https://doi.org/10.1111/j.1365-2133.1983.tb07074.x

[11] Aberer W, Konrad K, Wolff K. Wells' syndrome is a distinctive disease entity and not a histologic diagnosis. J Am Acad Dermatol 1988; 18: 105-14. https://doi.org/10.1016/s0190-9622(88)70016-x

[12] Heelan K, Ryan JF, Shear NH, Egan CA. Wells syndrome (eosinophilic cellulitis): Proposed diagnostic criteria and a literature review of the drug-induced variant. J Dermatol Case Rep 2013; 7: 113-20. https://doi.org/10.3315/jdcr.2013.1157

[13] Caputo R, Marzano AV, Vezzoli P, Lunardon L. Wells syndrome in adults and children: a report of 19 cases. Arch Dermatol 2006; 142: 1157-61. https://doi.org/10.1001/archderm.142.9.1157

[14] Anderson CR, Jenkins D, Tron V, Prendiville JS. Wells' syndrome in childhood: case report and review of the literature. J Am Acad Dermatol 1995; 33: 857-64. https://doi.org/10.1016/0190-9622(95)90423-9

[15] Räßler F, Lukács J, Elsner P. Treatment of eosinophilic cellulitis (Wells syndrome) - a systematic review. J Eur Acad Dermatol Venereol 2016; 30: 1465-79. https://doi.org/10.1111/jdv.13706

[16] Gandhi RK, Coloe J, Peters S, et al. Wells syndrome (eosinophilic cellulitis): a clinical imitator of bacterial cellulitis. J Clin Aesthet Dermatol 2011; 4: 55-7. PMID: 21779422.

[17] Aroni K, Aivaliotis M, Liossi A, Davaris P. Eosinophilic cellulitis in a child successfully treated with cetirizine. Acta Derm Venereol 1999; 79: 332. https://doi.org/10.1080/000155599750010841

[18] Marks R. Eosinophilic cellulitis--a response to treatment with dapsone: case report. Australas J Dermatol 1980; 21: 10-2. https://doi.org/10.1111/j.1440-0960.1980.tb00132.x

[19] Moossavi M, Mehregan DR. Wells' syndrome: a clinical and histopathologic review of seven cases. Int J Dermatol 2003; 42: 62-7. https://doi.org/10.1046/j.1365-4362.2003.01705.x

[20] Herr H, Koh JK. Eosinophilic cellulitis (Wells' syndrome) successfully treated with low-dose cyclosporine. J Korean Med Sci 2001; 16: 664-8. https://doi.org/10.3346/jkms.2001.16.5.664

[21] Paquet P, Laso-Dosal F, de la Brassinne M. Wells' syndrome: report of 2 cases. Dermatology 1992; 184: 139-41. https://doi.org/10.1159/000247523

[22] Stam-Westerveld EB, Daenen S, Van der Meer JB, Jonkman MF. Eosinophilic cellulitis (Wells' syndrome): treatment with minocycline. Acta Derm Venereol 1998; 78: 157. https://doi.org/10.1080/000155598433610

[23] Diridl E, Hönigsmann H, Tanew A. Wells' syndrome responsive to PUVA therapy. Br J Dermatol 1997; 137: 479-81. https://doi.org/10.1111/j.1365-2133.1997.tb03772.x

[24] Terhorst-Molawi D, Altrichter S, Röwert J, et al. Effective treatment with mepolizumab in a patient with refractory Wells syndrome. J Dtsch Dermatol Ges 2020; 18: 737-9. https://doi.org/10.1111/ddg.14151

[25] Coattrenec Y, Ibrahim Yasmine L, Harr T, et al. Long-term Remission of Wells Syndrome with Omalizumab. J Investig Allergol Clin Immunol 2020; 30: 58-9. https://doi.org/10.18176/jiaci.0436

[26] Traidl S, Angela Y, Kapp A, et al. Dupilumab in eosinophilic cellulitis (Wells' syndrome) - case report of a potential new treatment option. J Dtsch Dermatol Ges 2021; 19: 1653-5. https://doi.org/10.1111/ddg.14598