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Case Report

Diabetic patient presenting with Prurigo Nodularis like Eosinophilic Cellulitis (EC) Successfully Treated with Dupilumab

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Abstract

Prurigo Nodularis like Eosinophilic Cellulitis represents a unique and uncommon dermatologic condition characterized by pruritic nodules and inflammatory cellulitis, the aetiology of which remains elusive. The co-occurrence of these two entities poses diagnostic and therapeutic challenges. The clinical and histopathological features of Prurigo Nodularis like Eosinophilic Cellulitis, emphasizing the distinctive presentation of pruritic nodules alongside inflammatory cellulitis. The lack of a standardized treatment protocol underscores the complexity of managing this condition, with documented cases showing variable responses to oral and topical corticosteroids, immune suppressants, antihistamines, and other therapeutic modalities.

Keywords: *Eosinophilic Infiltrates, Refractory Skin Lesions, Wells' Syndrome, Eosinophilic Cellulitis, Dupilumab.*

Case Report

This case report presents a 37-year-old Indian diabetic male who developed severely pruritic and painful skin lesions accompanied by lower leg swelling following COVID-19 vaccination. Initial misdiagnosis and treatment for deep vein thrombosis (DVT) exacerbated his condition. Subsequent examination revealed hyperpigmented, tender papules, and nodules with both lower legs swelling (Fig 1). Histopathological analysis confirmed Eosinophilic Cellulitis (EC) characterized by hyperkeratosis, parakeratosis, fibrosis, and a polymorphous perivascular infiltrate rich in eosinophils. (Fig 2).

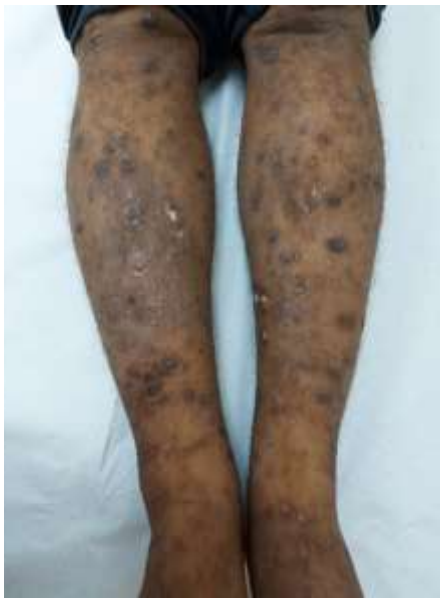

Figure 1

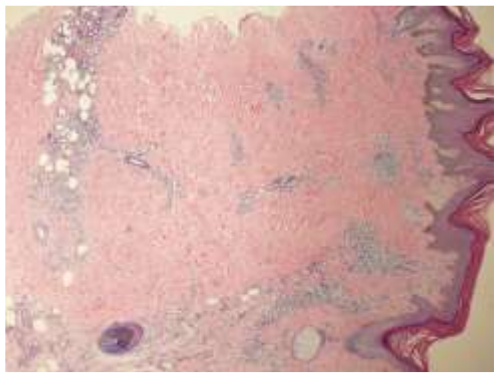

Figure 2

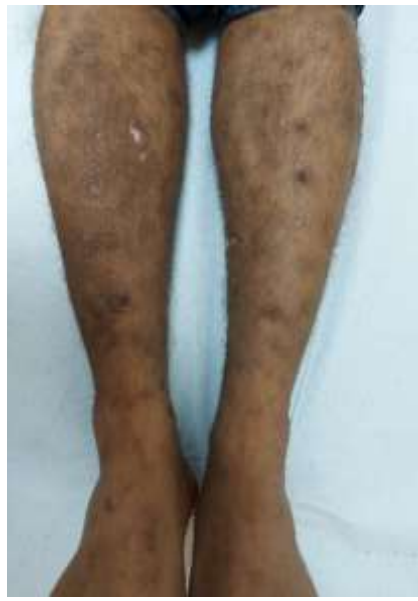

Figure 3

Despite initial therapy with glucocorticosteroids and doxycycline, the patient experienced only marginal improvement. Methotrexate was subsequently administered, resulting in a partial response and subsequent relapse upon cessation. The breakthrough in treatment occurred with the initiation of subcutaneous Dupilumab, administered at a loading dose of 600 mg followed by 300 mg every two weeks, in conjunction with topical emollients. Within three months, the patient exhibited significant improvement, with near-complete resolution of active lesions and nodules, maintaining this response on a maintenance dose of 300 mg Dupilumab every two weeks for 6 month (Fig 3) without recurrence.

Discussion

This case underscores the significance of clinic pathological correlation for diagnosing atypical presentations of Eosinophilic Cellulitis (EC), particularly in the context of challenging treatment scenarios. Our patient, a diabetic male, presented with pruritic and painful skin lesions on the lower legs following COVID-19 vaccination, resembling Prurigo Nodularis. The diagnostic journey, marked by initial misdiagnosis and inconsistent responses to conventional therapies, reflects the complexity associated with managing EC.

Wells' syndrome typically manifests on the extremities, trunk, and face,[1-4] making it crucial to consider in the differential diagnosis of localized recalcitrant inflammatory skin lesions. Histopathological analysis revealed the characteristic infiltration of reactive eosinophils and histiocytes within the affected dermis, aligning with the diagnostic criteria for Wells' syndrome.

The challenges in diagnosing EC arise from its infrequency and the absence of distinct markers. This complexity is reflected in the inconsistent responses to standard therapies, necessitating further investigation and personalized treatment strategies. Limited availability of large-scale clinical trials adds to the reliance on case reports and observational studies for guidance.

Standard treatments for EC, including oral and topical corticosteroids, methotrexate, and cyclosporine, exhibit varying efficacy.[5] Antihistamines are engaged for alleviating associated itching, while specific cases have shown successful resolution with treatments such as colchicines.[6] However, our case introduces an innovative therapeutic approach with subcutaneous Dupilumab. The patient displayed significant improvement and near-complete resolution of active lesions and nodules, a response comparable to previous cases reported by Traidl al.[7] Dupilumab received FDA approval in April 2017 for treating adult patients with moderate-to-severe atopic dermatitis, specifically when topical prescription therapies prove insufficient or are deemed inadvisable.[8] Furthermore, its application has extended to the treatment of prurigo nodularis (approved in September 2022) [9, 10] , bullous pemphigoid, [11] eosinophilic esophagitis, [12] among other conditions. Dupilumab, approved for atopic dermatitis and prurigo nodularis, showcases its potential as a versatile treatment option for EC.

The proposed mechanism of Dupilumab involves blocking IL-4 and IL-13 receptors, crucial in treating EC. Its efficacy in managing EC may stem from its ability to regulate eosinophil migration to the dermis and suppress IL-5 release through an IL-4 mediated pathway within eosinophils. This mechanism aligns with the observed enhancement of EC through Dupilumab, marking a notable advancement in addressing this complex dermatological ailment. [7, 13]

The lack of response to mepolizumab, targeting IL-5, suggests the involvement of immune mechanisms beyond IL-5 in EC development. Dupilumab's success in treating EC adds to the accumulating evidence of its effectiveness in confronting the complexities associated with this condition. [7, 14, 15]

Highlighting the patient's resistance to conventional treatments provides valuable insights into the potential efficacy of Dupilumab as a tailored therapeutic approach for cases where standard interventions fall short. In instances where standard treatments prove ineffective, Dupilumab emerges as a promising candidate for addressing refractory EC cases, presenting a novel avenue for therapeutic exploration. Further studies are warranted to explore the efficacy of Dupilumab in this context and its potential role in immune-mediated dermatological conditions post-vaccination.

Ethics Statement: Written informed consent was obtained from the participant's statutory guardian for the publication of any potentially identifiable images or data included in this article.

Conflict of Interest:

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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