Clinical, Cosmetic and Investigational Dermatology

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CASE REPORT

Rapid Improvement of Refractory Cutaneous Hypereosinophilic Syndrome with Abrocitinib Plus Methylprednisolone: A Case Report

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Abstract: Hypereosinophilic syndrome (HES) is a rare disorder characterized by persistent eosinophilia and end-organ damage. Cutaneous manifestations are frequently refractory to conventional therapies, including corticosteroids and immunosuppressants, creating a significant clinical challenge. The Janus kinase (JAK)-STAT pathway has been implicated in eosinophil activation and survival, suggesting a potential role for JAK inhibitors in management. In this context, we present the case of a 58-year-old female with a 10-year history of refractory generalized erythema, papules, and severe pruritus progressing to erythroderma with scaling. Previous treatments including antihistamines, tripterygium glycosides, glycyrrhizin, sodium thiosulfate, and topical glucocorticoids had failed. Laboratory investigations revealed leukocytosis with severe hypereosinophilia (peak 8.65×10^9/L, 52.6% of total WBC), hypoalbuminemia, elevated lactate dehydrogenase, and transaminitis. Skin biopsy demonstrated spongiotic edema and eosinophil-rich perivasculitis. Bone marrow examination confirmed eosinophilic hyperplasia (32.5% eosinophils) without evidence of clonality. Comprehensive parasitic and secondary causes were excluded. Following diagnosis of HES, the patient was initiated on methylprednisolone (40 mg/day) combined with abrocitinib (100 mg/day). Within one week, eosinophil count reduced significantly (1.24×10^9/L, 7%) with concurrent improvement in liver enzymes. Complete cutaneous remission was achieved at 2-month follow-up, enabling substantial steroid reduction. This case underscores that the combination therapy of abrocitinib and methylprednisolone can offer a promising approach for the HES.

Keywords: hypereosinophilic syndrome, JAK inhibitor, abrocitinib, corticosteroid, refractory, cutaneous manifestations, eosinophilia, targeted therapy

Introduction

Hypereosinophilic syndrome (HES) occurs when hypereosinophilia is associated with a clinically relevant organ damage attributable to eosinophils. He cause encompass a broad spectrum of etiologies and HES can be sub-classified in reactive, clonal, familiar, and idiopathic form. This condition can affect multiple organ systems, frequently leading to significant damage in organs such as the heart, lungs, skin, gastrointestinal tract, and nervous system. The diagnosis of idiopathic HES requires the exclusion of all primary and secondary HE as well as lymphocytic-variant hypereosinophilic syndrome. The previously used criterion was peripheral blood eosinophils > 1.5×10^9/L for more than 6 months, accompanied by tissue damage. However, this was only for retrospective analysis of this category of diseases rather than for diagnosis. The current diagnostic criteria proposed by the World Health Organization in 2024 is peripheral blood eosinophils >

Received: 11 September 2025
Accepted: 18 December 2025
Published: 20 December 2025

1.5×10^9/L for 2 to 4 weeks continuously.[1] Recent insights into the pathogenesis of HES have highlighted the crucial role of the Janus kinase-signal transducer and activator of transcription (JAK-STAT) signaling pathway. Several cytokines pivotal for eosinophil proliferation, survival, and activation—including IL-5, IL-3, and granulocyte-macrophage colony-stimulating factor (GM-CSF)—signal through this pathway.[2,3] This provides a strong rationale for investigating JAK inhibitors as a therapeutic strategy in HES. Although the efficacy of other JAK inhibitors such as ruxolitinib and baricitinib in diseases associated with eosinophilia has been reported, the experience with novel and more selective drugs, especially for cases with predominant skin symptoms and refractory conditions, remains extremely limited.[4,5] In this report, we present the case of a woman diagnosed with idiopathic HES who was successfully treated with a combination of abrocitinib and Methylprednisolone. This breakthrough provides a promising therapeutic alternative for future management of HES.

Case Presentation

A 58-year-old female presented with a 10-year history of generalized erythema, papules, and severe pruritus, progressing to flushing and desquamation. The pathological biopsy conducted at the time of admission showed hyperkeratosis, hypokeratosis, mild acanthosis, spongiotic edema, scattered lymphocytic inflammatory cell infiltration around superficial dermal vessels, and sparse eosinophils (Figure 1A). Initial diagnoses included erythroderma and atopic dermatitis. Despite extensive treatments (antihistamines, tripterygium glycoside tablets, compound glycyrrhizin injection, sodium

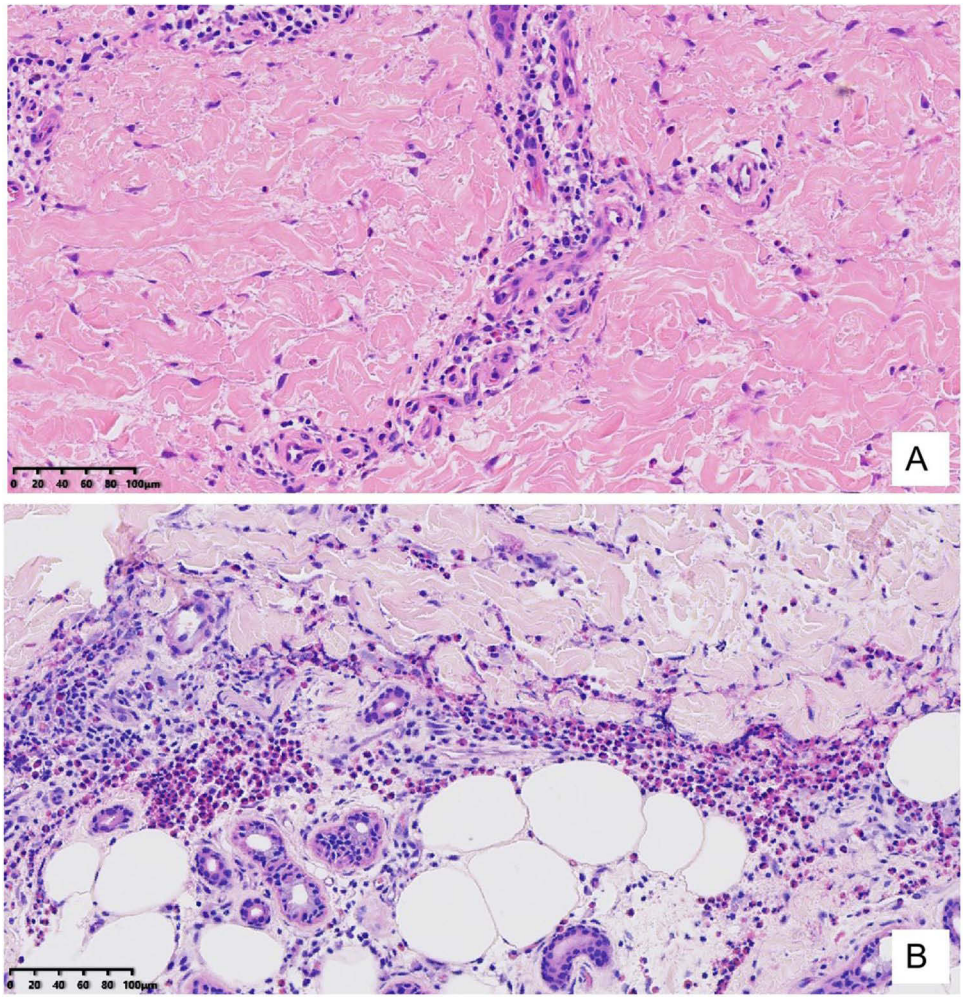

Figure 1 (**A**) On December 25, 2023: scattered lymphocytic inflammatory cell infiltration around superficial dermal vessels, and sparse eosinophils. (H&E stain, original magnification ×200). (**B**) a spongiotic edematous type of deep and superficial perivascular dermatitis with eosinophils with eosinophliginal magnification ×200).

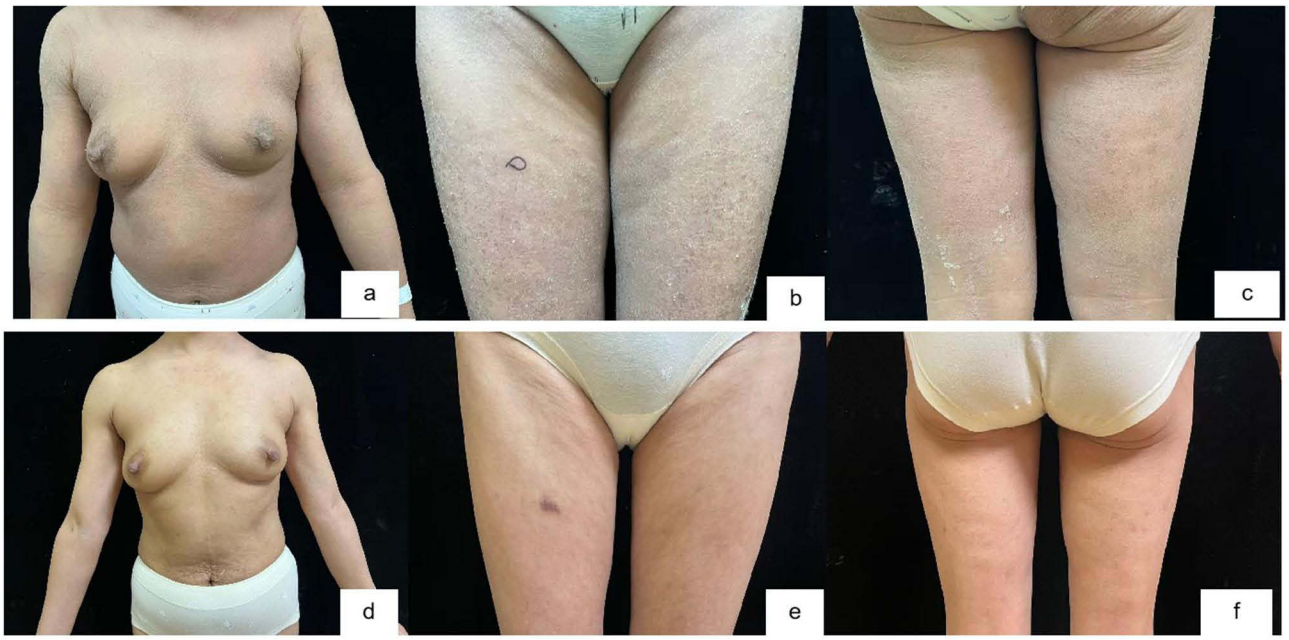

Figure 2 (**a–c**) At admission: diffuse erythema covering the entire body with numerous small, grayish-white flaky scales on the surface of the erythematous areas; no thin film formation or punctate bleeding was observed after scaling. (**d–f**) Two months later: the whole body rash subsided.

thiosulfate injection, and topical glucocorticoids), her condition worsened, with persistently elevated eosinophils (peak: 8.65×10^9/L; 52.6% of total WBC) and abnormal liver function. Physical examination revealed diffuse erythema with scaling and lower limb edema (Figure 2a–c). Laboratory findings included leukocytosis with severe hypereosinophilia, hypoalbuminemia, elevated lactate dehydrogenase, coagulopathy, leukocyturia (+3500 cells/μL) and hematuria (+++/HP). Stool occult blood, urine microalbumin, ANCA, ANA, anti-dsDNA, renal function, IgE, troponin, and a TB-specific T-cell assay, were within normal limits. Imaging studies (abdominal ultrasound, chest CT) and an electrocardiogram also showed no abnormalities. The previously reported hematuria resolved on repeat testing and was deemed transient, likely related to menstrual contamination. Skin biopsies demonstrated spongiotic edema and eosinophil-rich perivasculitis (Figure 1B). Bone marrow biopsy confirmed active eosinophilic hyperplasia (32.5% eosinophils), A comprehensive 56-gene fusion panel screening for mutations including *FIP1L1-PDGFRA*PDGFRB, FGFR1, JAK2, ETV6-PDGFRB, and BCR-ABL1 and serological testing for nine parasites (including *Schistosoma*, *Clonorchis*, *Paragonimus*, *Toxoplasma gondii*, Cysticercus, *Echinococcus*, *Trichinella spiralis*, Sparganum mansoni, and *Angiostrongylus cantonensis*) were negative. Finally, the patient denied any neurological or significant gastrointestinal symptoms upon detailed interview and examination. A diagnosis of HES was established. The initiation of combination therapy with intravenous methylprednisolone (40 mg/day, ≈0.8 mg/kg/day) and oral abrocitinib (100 mg/day) prompted a rapid clinical response. Within one week, eosinophil counts fell significantly to 1.24×10^9/L (7%), liver enzymes normalized, and skin lesions showed substantial resolution. On this basis, a structured corticosteroid taper was implemented. The regimen was successfully de-escalated from methylprednisolone to triamcinolone, progressing from 32 mg/day (Jan 17–30) to 16 mg/day (Jan 31-Feb 14) and then to 12 mg/day (Feb 15-Mar 1). By the follow-up on March 2, The patient was transitioned to abrocitinib monotherapy (100 mg/day), following the discontinuation of prednisone. At 2-month follow-up, complete remission of rash was sustained (Figure 2d–f), and thus did not return for subsequent visits.

Discussion

This case illustrates the successful and rapid management of a refractory, long-standing cutaneous lesions in idiopathic hypereosinophilic syndrome (HES) with predominant cutaneous involvement using a combination of systemic glucocorticoids and the Janus kinase 1 (JAK1)-selective inhibitor, abrocitinib. The patient, who had failed a decade of conventional therapies, achieved reduction of eosinophils, resolution of abnormal liver enzymes, and complete clearing

of her debilitating skin lesions within a remarkably short timeframe. This outcome underscores the potential of JAK inhibition as a highly effective and potential for steroid-sparing in the treatment algorithm for HES.

The efficacy of this combination therapy can be attributed to the complementary mechanisms of action targeting the core pathophysiology of HES. Glucocorticoids exert broad anti-inflammatory and immunosuppressive effects, rapidly reducing eosinophil proliferation and activation. However, as evidenced by this case, their long-term efficacy is often limited by dependency and significant side effects. Abrocitinib, on the other hand, provides a more targeted approach. A substantial body of evidence implicates the JAK-STAT signaling pathway as central to the production, survival, and activation of eosinophils, primarily mediated by cytokines such as IL-5, IL-4, and IL-13.[3,6,7] By selectively inhibiting JAK1, abrocitinib potently blocks the signaling of these key cytokines, thereby interrupting the upstream drivers of eosinophilic inflammation and Th2-mediated immune response that likely underpinned this patient's persistent dermatitis and pruritus.

Our findings are consistent with the growing body of literature supporting the role of JAK inhibitors in HES. Previous reports have demonstrated success with the JAK1/2 inhibitor ruxolitinib in both corticosteroid-resistant and -dependent HES. However, this case adds several novel and important insights. First, to our knowledge, this is the first detailed report of the successful use of abrocitinib, a JAK1-selective inhibitor primarily approved for atopic dermatitis, in the management of HES. Its high selectivity may offer a favorable benefit-risk profile compared to broader-spectrum JAK inhibitors. Second, the speed and magnitude of the response were striking, with significant improvement in both hematological and cutaneous parameters within one week. This rapid onset of action suggests that JAK inhibition can be particularly valuable for achieving quick disease control. Finally, the use of abrocitinib facilitated a dramatic reduction in glucocorticoid exposure, directly addressing a major unmet need in the long-term management of HES—minimizing steroid-related morbidity.

The diagnostic journey in this case underscores the complexity of HES. The initial pathological finding of "subacute dermatitis consistent with eczema" highlights how HES can masquerade as more common benign dermatoses. The subsequent biopsy revealing eosinophil-rich perivasculitis was a critical clue, prompting a systemic workup that ultimately confirmed the hematological nature of the disease through bone marrow studies. The exclusion of parasitic infections and other secondary causes was essential to solidify the HES diagnosis. Our case also highlights certain diagnostic challenges. We acknowledge limitations such as the clinical diagnosis of acute interstitial nephritis without biopsy confirmation or eosinophiluria data, and the fact that L-HES was not ruled out by specific T-cell immunophenotyping. These gaps, however, point directly to future directions. The compelling efficacy of JAK inhibitors in L-HES, as shown by Faguer,[8] suggests that in similar cases with atypical or refractory courses, re-evaluation with these advanced diagnostics could unlock pivotal targeted treatment options, moving beyond empirical therapy.

We acknowledge the limitations inherent in a single case report. The findings require validation in larger, prospective cohorts and randomized controlled trials to firmly establish the efficacy and safety of abrocitinib in HES. The follow-up period of two months, while demonstrating excellent initial response, is insufficient to evaluate the long-term durability of remission and the potential for adverse events associated with chronic JAK inhibition. Furthermore, the simultaneous initiation of methylprednisolone and abrocitinib makes it challenging to delineate the individual contribution of each agent to the observed outcome, though the prior failure of steroid monotherapy suggests a pivotal role for abrocitinib.

Conclusion

In conclusion, this case provides compelling evidence that abrocitinib, in combination with glucocorticoids, can serve as a highly effective and rapidly acting therapeutic option for refractory HES with cutaneous manifestations. It offers a promising steroid-sparing strategy, potentially mitigating the long-term complications of corticosteroid therapy. Future studies should focus on identifying biomarkers that predict response to JAK inhibition, determining the optimal timing and sequencing of therapy, and evaluating the long-term outcomes of JAK1-selective inhibitors in the management of HES and other eosinophil-driven disorders.

Abbreviations

HES, Hypereosinophilic syndrome; JAK1, Janus kinase 1.

Ethics Statement

The patient provided written informed consent for publication of this report and accompanying images. The Ethics Committee of Jiangxi Provincial Dermatology Hospital, has approved the publication of the case details.

Acknowledgments

Bei Liao, Xinze Li and Qianjie Qiu are the co-first authors for this paper. These authors contributed equally to this work.

Funding

There is no funding to report.

Disclosure

The authors report no conflicts of interest in this work.

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