Eosinophilic Dermatosis of Hematologic Malignancy Presenting as Suspected Drug Eruption in a Patient with Chronic Lymphocytic Leukemia

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Abstract

Eosinophilic Dermatosis of Hematologic Malignancy (EDHM) is a rare cutaneous reaction associated with hematologic malignancies, including Chronic Lymphocytic Leukemia (CLL). The lesions clinically and histologically resemble insect bites; however, in most cases the patients strongly deny any history of such bites. Treatment modalities vary, though systemic steroids are most commonly used. Because EDHM can be associated with a more aggressive malignancy course, careful monitoring of patients with this eruption is essential. We present a severely pruritic, rapid onset cutaneous eruption initially thought to be a drug or bug bite reaction but ultimately presented as EDHM.

Keywords: Eosinophilic dermatosis of hematologic malignancy • Chronic lymphocytic leukemia • Pruritic

Introduction

Eosinophilic Dermatosis of Hematologic Malignancy (EDHM) is an uncommon cutaneous reaction that is associated with hematologic malignancies. Of all hematological malignancies, it has most commonly been associated with Chronic Lymphocytic Leukemia (CLL) [1]. Though many treatment modalities have been reported, EDHM often responds poorly to treatment. We present a patient with CLL who was diagnosed with EDHM and treated with steroids.

Case Study

A 60-year-old female with a past medical history of Chronic Lymphocytic Leukemia (CLL) who recently started Ibrutinib presented to the emergency department with a four-day history of a severely pruritic, eruptive

full-body rash. The lesions started on her bilateral legs and then spread to the rest of her body. The patient was started on an antitussive and oral prednisone for upper respiratory tract symptoms 7 days prior to ED presentation.

Physical examination revealed smooth papules and plaques scattered along the extremities, chest, abdomen, back, and face. The papules and plaques were pink, edematous, and dome-shaped. Many contained a central vesicle, while others exhibited central crusting (Figures 1 and 2).

The clinical differential diagnosis included drug eruption, arthropod assault, Sweet syndrome, and bulbous pemphigoid.

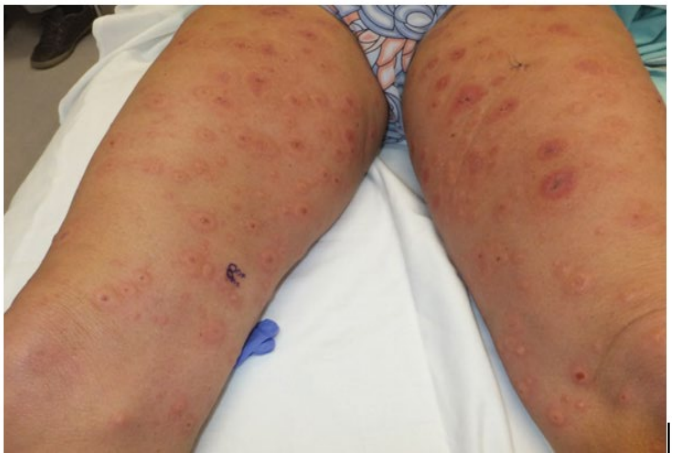

Figure 1. Pruritic papules and plaques scattered along the patient's lower extremities.

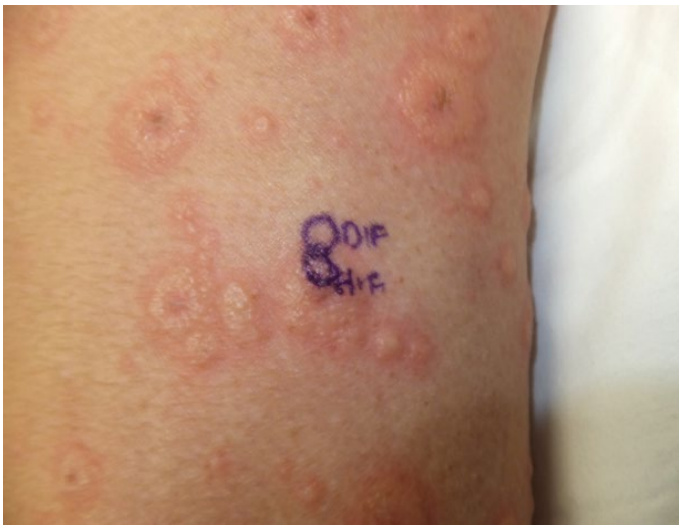

Figure 2. Close-up view of lesions showing central vesicles and crusting.

Histological sections demonstrated a sub epidermal blister containing lymphocytes, eosinophils and neutrophils, as well as mild perivascular and interstitial lymphocytic infiltrate with eosinophils within the dermis. Direct immunofluorescent antibody localization demonstrated negative immunoreactivity for immunoglobulins IgG, IgA, IgM and complement C3.

The patient strongly denied exposure to bugs and the clinical differential was settled on EDHM *vs* drug eruption, given that she had recently started ibrutinib. The patient was treated with a prednisone taper, topical corticosteroids, and antihistamines. Two weeks later there was significant improvement in the skin lesions, with almost complete relief from symptoms. She continued to have complete resolution of her cutaneous eruption despite continuing ibrutinib therapy. When other potential causes were excluded, the patient was given the diagnosis of EDHM.

Discussion

Chronic Lymphocytic leukemia (CLL) is a monoclonal disorder characterized by progressive accumulation of dysfunctional lymphocytes. EDHM is a rare cutaneous reaction associated with

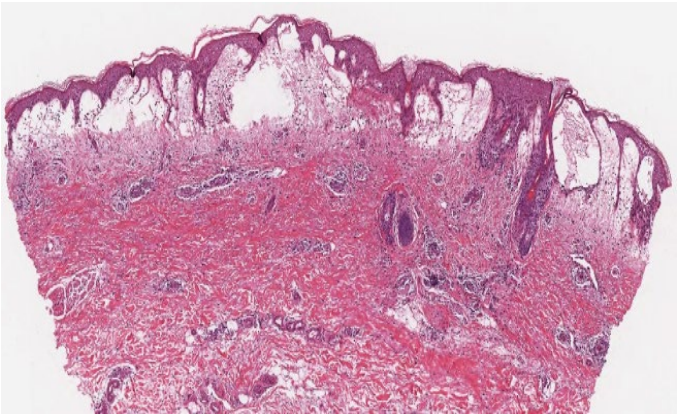

Figure 3. A biopsy specimen revealed a sub epidermal blister containing lymphocytes, eosinophils, and neutrophils, as well as mild perivascular and interstitial lymphocytic infiltrate with eosinophils within the dermis (H&E, 60x).

many hematologic malignancies, though most commonly seen with CLL [1]. The eosinophilic eruption most likely manifests through a Th2 chemokine response, most likely due to interleukin-5, a key cytokine in eosinophil recruitment [2]. EDHM is also referred to as insect bite-like reaction due to clinically and histopathologically resembling insect bites, despite patients denying a history of being bitten. Histological sections can also be confused with a drug eruption, as seen in this case.

EDHM can present with pronounced symptoms of pruritus and tenderness, as seen in this patient. A retrospective cohort study of 37 patients diagnosed with EDHM found only 25% of cases presented with lesions on the face [3] (Figure 3).

Various treatment strategies for EDHM have been used, utilizing antibiotics, steroids, antihistamines, dapsone, phototherapy, radiation, interferon alpha, and intravenous immunoglobulin. Cycles of chemotherapy have also been reported to improve EDHM symptoms [2]. This patient was successfully treated with a prednisone taper, topical triamcinolone 0.1% ointment, and antihistamines. EDHM may be associated with a more aggressive CLL-disease course, as it has been reported with Richter transformation and other fatal complications of CLL [3,4].

Conclusion

We report a case of EDHM in a 60-year-old patient with a past medical history of CLL. This patient presented clinically with widespread papules and plaques that were severely pruritic. The patient was treated with a prednisone taper, topical corticosteroids, and antihistamines, with significant improvement in skin lesions and almost completes relief from symptoms within 2 weeks of treatment. The diagnosis of EDHM was given after other potential causes were excluded, such as drug eruption and arthropod bite reaction. Careful monitoring of these patients is essential because EDHM can be associated with a more aggressive malignancy course.

References

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