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Eosinophilic dermatosis in a patient with chronic lymphocytic leukemia: A case report

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Abstract

An intensely pruritic dermatosis has been described in patients with hematologic diseases that is reminiscent of insect bites [1]. Initially it was referred to as exaggerated insect bite reaction and was classically associated with chronic lymphocytic leukemia (CLL). Nowadays, the term eosinophilic dermatosis associated with hematologic malignancy (EDHM) is used to emphasize that it can be related to other hematologic disorders [2,3]. This report describes a case of EDHM that represented a diagnostic and therapeutic challenge.

Keywords

Chronic lymphocytic leukemia; Hematologic malignancy; Pruritic dermatosis.

Case Report

A 55-year-old man presented with a 2-week history of a pruritic dermatosis. His past medical history was relevant for a third relapse of CLL (RAI I, Binet B), for which he was being treated with FCR (fludarabine, cyclophosphamide, and rituximab). During his follow-up, tuberculous lymphadenitis due to *Mycobacterium bovis* was documented, so chemotherapy was withheld, and treatment for tuberculosis was started.

Upon examination, tense erythematous bullae were observed on the scalp and legs, and crusted papules were present on the arms. Treatment was started with topical antibiotics, antihistamines and topical steroids, without improvement. One month later, new lesions on the extremities had appeared, and the pruritus persisted. A skin biopsy showed sub epidermal blistering associated with superficial and deep lymphocytic nodular dermatitis with countless eosinophils. Negative results were obtained from parasite stool test and HIV serology. Finally, a diagnosis of EDHM was made.

After 3 months of anti-tuberculosis treatment, FCR was restarted due to the development of new lymphadenopathies secondary to CLL. After the re-start of chemotherapy, the patient reported improvement in pruritus and no new lesions. After completing 6 cycles of FCR, here mained without skin lesions and pruritus. He lost his follow-up in our institution due to the COVID-19 pandemic and unfortunately expired 3 years after the onset of EDHM.

Discussion

A hallmark of EDHM is pruritus, and this entity should be suspected when patients refer intense pruritus that does not respond to conventional treatment. Itching significantly interferes with patients' quality of life, and it is often the chief complaint. The onset of EDHM can precede the diagnosis of a hematologic disease [2]. Upon its recurrence, pruritus should also be regarded as a cutaneous sign of treatment failure and malignancy relapse. Clinicians who care for individuals with CLL and other hematologic malignancies must be aware of this entity since it can be a herald of the disease's course.

The diagnosis of EDHM is challenging and many times reached retrospectively. There is no gold standard test to confirm the diagnosis and distinguishing from other differentials may be impossible. A skin biopsy can support the diagnosis of EDHM but findings are not pathognomonic and can be found in other differentials. Another tool that can aid in establishing a diagnosis is the proposed criteria by Byrd et al. (Table 1).

Table 1: EDHM diagnostic criteria.

1)	Pruritic papules, nodules, and/or vesiculobullous eruptions that were resistant to conservative management
2)	Histopathologically confirmed eosinophil-rich dermal lymphohistiocytic infiltration at the superficial and deep dermis
3)	Exclusion of other causes of tissue eosinophilia
4)	A pre-existing diagnosis of a hematological malignancy

The pruritus can be improved with topical high-potency steroids and systemic antihistamines. However, our case shows that some will be refractory to this medications and warrant directed treatment to the malignancy in order to reach full resolution [2-4]. An alternative is the use of systemic steroids, however, as shown in our case, comorbidities, infectious processes, and undesired side-effects can bar their use.

Conclusion

This report seeks to highlight the importance of recognizing EDHM as a cause of intractable pruritus with important quality of life effects, as well as a cutaneous sign that can lead to a diagnosis, or relapse, of hematologic malignancy. The diagnosis is challenging because no pathognomonic finding exists and clinicopathological correlation in the appropriate clinical setting must be reached. In cases refractory to conventional treatment, EDHM might only respond to malignancy-directed therapy.

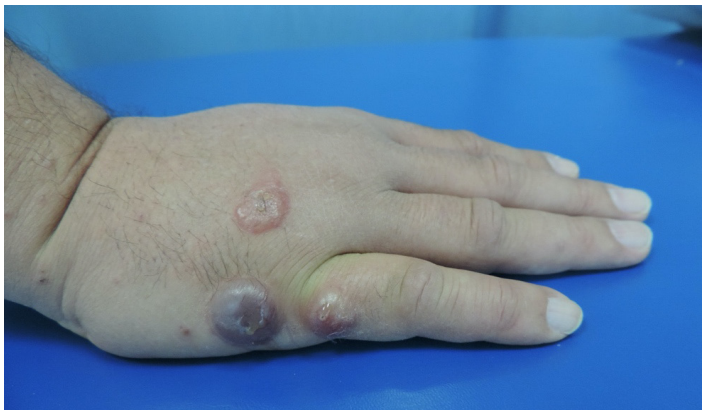

Figure 1: Detailed view of the hand with eroded bullae and serous hemorrhagic blister.

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