Diagnostic and Therapeutic Challenges of Cutaneous Lymphoid Hyperplasia in the Facial Region: A Case Report

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ABSTRACT Cutaneous lymphoid hyperplasia (CLH) is a benign skin disorder that clinically and histopathologically mimics cutaneous lymphoma, making it a diagnostic challenge. CLH can exhibit a predominance of B cells, T cells, or a combination of both. This report illustrates a case of a 27-year-old male patient with an erythematous plaque on his cheek, initially histopathologically described as lymphoid hyperplasia of B-immunophenotype. The lesion recurred years later, necessitating a thorough diagnostic investigation.
Clonality analysis coupled with imaging led to the exclusion of cutaneous lymphoma, therefore confirming B-cell CLH as the final diagnosis. Treatment options were limited by the esthetic demands of the facial region, as well as the patient's age, considering the risks of radiotherapy in younger patients.
Furthermore, the possibility of progression to cutaneous lymphoma was considered, highlighting the importance of long-term patient follow-up.

KEY WORDS: Esthetics; Lymphoproliferative Disorders; Pseudolymphoma; Skin Neoplasms

INTRODUCTION

Cutaneous lymphoid hyperplasia (CLH) is a benign skin disorder that clinically and histopathologically resembles cutaneous lymphoma (1). Common synonyms for CLH used in the literature include cutaneous pseudolymphoma, lymphocytoma cutis, reactive lymphoid hyperplasia, lymphadenosis benigna cutis, and Spiegler-Fendt sarcoid (2). CLH can exhibit a predominance of B cells, T cells, or a combination of both. Recent classifications categorize pseudolymphomas as nodular pseudolymphomas, pseudolymphomas as stimulators of Mycosis fungoides, intravascular pseudolymphomas and other pseudolymphomas (3). The condition usually affects adults but can develop at any age. B-cell CLH is more frequent and more commonly seen in women when compared to T-cell CLH (4).

Epidemiological data on these conditions remain limited, and no familial cases have been reported (4). B-cell CLH typically presents as a solitary, painless, and slowly growing erythematous nodule or plaque (3). While most B-cell CLH cases are idiopathic, some may be associated with factors such as Borrelia Burgdorferi infection, arthropod bites, tattoos, vaccines, piercings, or drugs (5). We present a case of a CLH on

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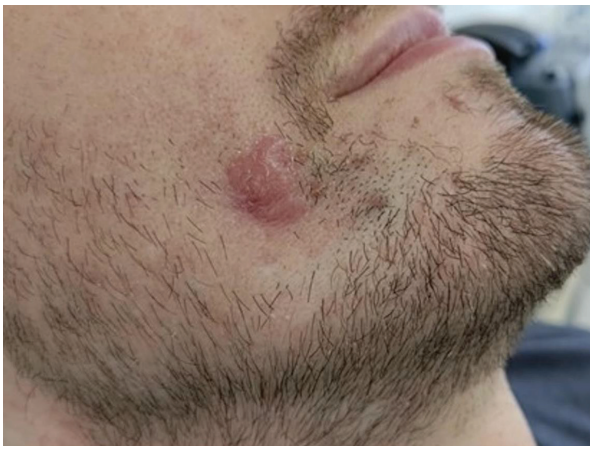

Figure 1. CLH on the right cheek.

the cheek that highlights the diagnostic and therapeutic challenges, particularly considering the aesthetic considerations related to this delicate localization.

CASE REPORT

A 27-year-old male presented with a solitary erythematous papule in his cheek in the mandibular region that progressed to a 12 mm plaque. [Figure 1, Figure 2] A biopsy with the subsequent histopathological analysis pointed to a lymphoid hyperplasia of B-immunophenotype. [Figure 3] Four years later, the plaque reappeared near the previous excision scar, necessitating additional diagnostics to exclude the possibility of cutaneous lymphoma. An excisional biopsy was performed, which once again indicated lymphoid hyperplasia of B-immunophenotype. Clonality analysis of the sample, along with an MSCT of the thorax, abdomen, and pelvis, were performed. Clonality analysis revealed a polyclonal B-cell population, and the MSCT results were unremarkable. The conclusive diagnosis of CLH and the exclusion of cutaneous lymphoma could only be confidently established after conducting these procedures. Considering that cutaneous lymphoma was ruled out, further diagnostic evaluation has been carried out. The serology for Borrelia burgdorferi was negative, and the patient denied any history of arthropod bites or other common etiological factors, making this most likely an idiopathic case. The lesion reccurred in the previous location once again, initially responding well to topical corticosteroids and achieving total regression. However, it recurred and proved refractory to both topical and intralesional corticosteroid treatments. Cryotherapy was administered through multiple cycles, resulting in partial regression with subsequent recurrences following each treatment. The patient is currently being treated with alternating cycles of cryotherapy and intralesional corticosteroid application.

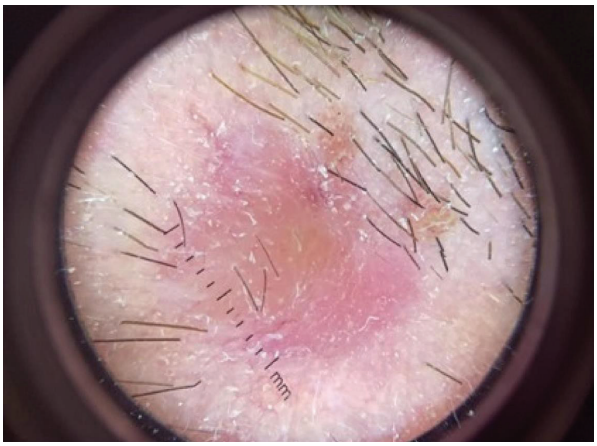

Figure 2. Dermoscopic findings of CLH.

DISCUSSION

CLH is a benign lymphoproliferative disease that clinically closely resembles cutaneous lymphoma and shows similar histopathological characteristics, which can pose a diagnostic challenge. CLH and low-grade cutaneous lymphoma are at two ends of a spectrum, which is why clinico-pathological correlation, sometimes followed by clonality analysis, is crucial for the correct diagnosis, with the finding of a polyclonal lymphocyte population favoring the diagnosis of CLH. However, cases of patients who have initially been diagnosed with CLH later being found to have cutaneous B-cell lymphoma have been described in the literature (6, 7, 8). It remains uncertain whether

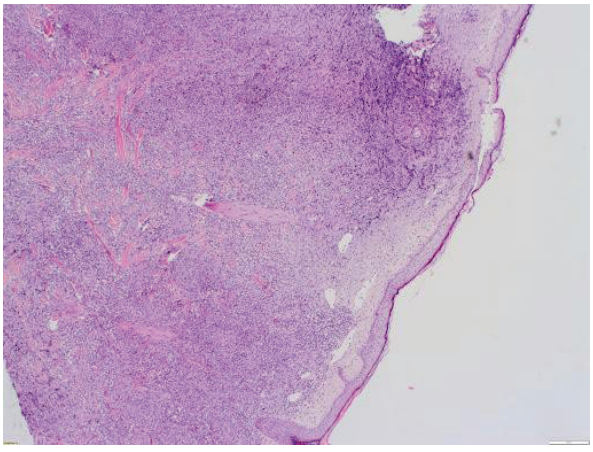

Figure 3. Histopathological finding. Throughout the thickness of the dermis, there is an extensive infiltrate, partly nodular and partly diffuse, consisting of small, medium-sized, and large lymphocytes. Several lymphatic follicles with germinal centers of normal appearance and an expanded marginal zone are observed. The lymphocytes do not exhibit cytological atypia.

disease progression from CLH to cutaneous lymphoma occurred or whether the patients had low-grade cutaneous lymphoma that had been misdiagnosed as CLH. Therefore, existing diagnostic criteria are not precise enough for all patients, which is why closely monitoring patients to timely detect the possible progression is vital, even if cutaneous lymphoma is ruled out initially. The question of the frequency of follow-up exams needs to be further discussed.

Additionally, given that this is a case of a young patient presenting with CLH in the facial region, esthetic demands are very important when looking at therapeutic options. Due to the benign nature of CLH, the initial approach involved the least aggressive therapeutic options, such as cryotherapy and corticosteroids, both topical and intralesional, which led only to a minor improvement. Considering esthetic outcomes, a third excision would be suboptimal. On the other hand, radiotherapy has proven useful in the therapy of CLH, yielding satisfactory outcomes (9). However, given the location of the lesion and the patient's age, potential complications have to be taken into account. Firstly, there is a risk of radiation-induced skin fibrosis, which might worsen the cosmetic result (10). Furthermore, the fibrosis could complicate any subsequent excisions if they are needed. Secondly, the risk of radiation-induced malignancy, even though it is low, presents too great of a downside for this type of therapy given the patient's age and the expected lifespan (11). To improve the management of CLH, a larger patient cohort is essential.

Case reports such as this one play a crucial role in highlighting the challenges of treating CLH in this challenging anatomical location. Conclusion

This case illustrates the complexities associated with the management of CLH, in particular when it occurs in a delicate facial region. Given that the line between CLH and cutaneous lymphoma can be difficult to delineate, a thorough diagnostic approach that includes clonality analysis should be coupled with long-term follow-up. From a therapeutic standpoint, it is important to weigh the esthetic considerations with the risks tied to more aggressive therapeutic options such as radiotherapy, especially when treating younger patients.

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Disclosure of interest

The authors declare that they have no competing interest.

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