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Case Report

Dual cutaneous lymphoid dyscrasias in a pediatric patient, with progression to mycosis fungoides: case report

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ABSTRACT

This case report explores the progression of cutaneous lymphoid dyscrasia to a lymphoma in a 13-year-old boy with pityriasis lichenoides chronica and lichenoid pigmented purpuric dermatitis, the latter evolving into mycosis fungoides. We highlight some of the crucial clinical and histological characteristics to differentiate between these entities. Early identification of this progression is critical not only for preventing further lymphoma development but also for initiating prompt therapy.

Keywords: Dyscrasia, Cutaneous lymphoma, Mycosis fungoides, Progression, Purpuric pigmented dermatitis, Pediatric

INTRODUCTION

Cutaneous T-cell lymphoid dyscrasias are defined by the monoclonal or oligoclonal proliferation of T cells without fulfilling the diagnostic criteria for cutaneous lymphomas. In certain cases, they can occur several years before cutaneous T-cell lymphomas onset, with mycosis fungoides (MF) being the most prevalent.[1] This case report presents a pediatric patient with a 10-year follow-up period, who underwent two different types of lymphoid dyscrasias: *pityriasis lichenoides chronica* (PLC) and lichenoid pigmented purpuric dermatitis (LPPD). The latter exhibited a progression that culminated in MF.

CASE REPORT

A 13-year-old male presented with a bilateral disseminated dermatosis, affecting the lower limbs. It was characterized by erythematous-scaly papular plaques on the thighs (Figure 1a). Additionally, there was a localized dermatosis on the distal right lower limb characterized by a well-defined, slightly scaly, purple plaque (Figure 1b), which had been present for 10 and 8 years, respectively. These conditions remained asymptomatic.

With suspicion of PLC, LPPD, and MF, two biopsies were performed, one from a small erythematous-scaly plaque on the thigh and the other from the purpuric lesion on the distal lower limb. The first biopsy revealed focal parakeratosis, vacuolization of the basal layer, suprabasal necrotic keratinocytes, extravasation of erythrocytes, and a perivascular infiltrate of slightly enlarged lymphocytes, findings consistent with *pityriasis lichenoides* (Figure 2a). The second biopsy showed a band-like infiltrate composed of slightly enlarged lymphocytes, lymphocyte exocytosis, erythrocyte extravasation, and abundant hemosiderin, consistent with LPPD (Figure 2b).

The patient had a history of presenting multiple erythematous-scaly plaques on the trunk and all four limbs at the age of 3, with biopsies reporting PLC. At that time, the patient was lost to follow-up.

With the established diagnosis of LPPD and PLC, narrow band ultraviolet B (NB-UVB) phototherapy was initiated. After 18 months (71 sessions of phototherapy), a significant improvement in PLC lesions was observed. However, the LPPD appeared more pigmented, prompting the decision to pause treatment for a month and perform a

new biopsy. The subsequent histopathological study described a band-like infiltrate of large and atypical lymphocytes (Figure 3). Immunostaining revealed a CD4/CD8 ratio of 4:1, a weakly positive CD7, and reported erythrocyte extravasation. A diagnosis of plaque-phase MF was established. Treatment was resumed with NB-UVB phototherapy, calcineurin inhibitors, and topical retinoids for another 2 years, resulting in significant clearance and improvement. Unfortunately, patient follow-up was lost due to the COVID-19 pandemic.

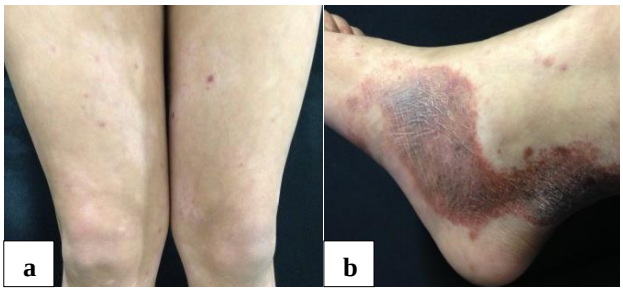

Figure 1: (a) Pytiriasis lichenoides chronica on the thighs, and (b) lichenoid pigmented purpuric dermatitis on the right limb.

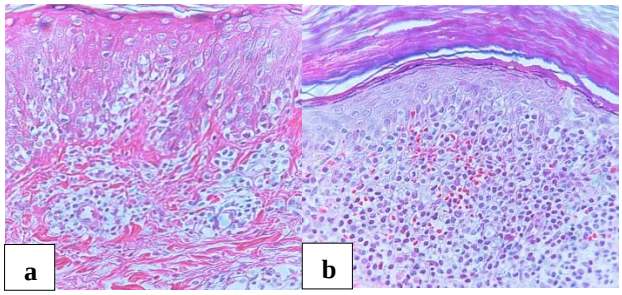

Figure 2: (a) Histopathological features of pytiriasis lichenoides chronica showing vacuolization of the basal layer, suprabasal necrotic keratinocytes, extravasation of erythrocytes and a perivascular infiltrate of slightly enlarged lymphocytes; and (b) histopathological features of liquenoid pigmented purpuric dermatitis showing slightly enlarged lymphocytes, lymphocyte exocytosis, erythrocyte extravasation, and abundant hemosiderin.

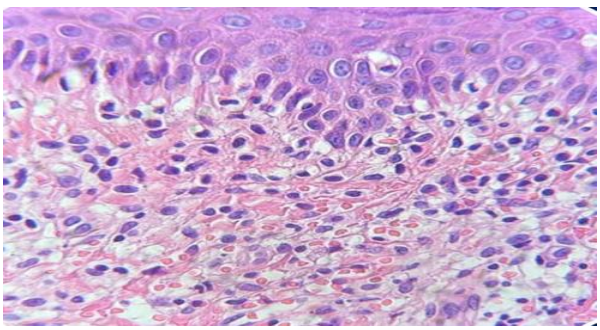

Figure 3: Histopathological features of mycosis fungoides showing a large and atypical lymphocytes infiltrate in a band-like pattern.

DISCUSSION

The PLC is a variant of cutaneous T-cell lymphoid dyscrasias, characterized by papulosquamous lesions, and is mainly diagnosed in pediatric and young adult patients. Its inclination to transition to MF has been well documented, especially in those with immunophenotypic profiles characterized by reduced CD7 and CD62.[2]

On the other hand, pigmented purpuric dermatitis (PPD) constitutes a group of various patterns of the same illness with a similar histopathology, it is commonly a benign skin disease associated to autoimmune diseases, dyslipidemias, drugs, venous insufficiency, among others. There are reports and studies about its association with MF. While some experts categorize them in certain cases as precursor lymphoid dyscrasias of cutaneous lymphomas, others suggest that they could be considered an atypical variant of MF.[3-5]

In clinical cases showing suggestive signs of PPD with a tendency to evolve towards MF, a broader than usual cutaneous dissemination is observed, with larger lesions that tend to coalesce. These lesions are also lichenoid, accompanied by itching, and persist for at least one year. In the context of pediatric patients with chronic progressive dermatosis, MF should be taken into consideration as a potential differential diagnosis.[4,6,7]

From a histopathological perspective, the presence of Pautrier's clusters, cerebriform lymphocytes, lymphocytic infiltrate with epidermotropism, and the loss of expression of mature T-cell markers, such as CD7 and CD62L, are characteristics related to MF. However, it is possible to confuse PPD with MF due to the similarity of some histological aspects, such as epidermotropism and lymphocytes at the dermoepidermal junction.[1,4,6] To clarify the diagnosis, immunohistochemical techniques demonstrating a predominance of CD8+ infiltrate and genetic rearrangement studies revealing polyclonality can be performed. Together, the analyses provide a higher degree of certainty regarding the nature of benign cases of PPD not associated with progression to a cutaneous lymphoma.[4,6]

With confirmed diagnoses of PLC and PPD, and the worsening of the latter one, close monitoring was part of the management protocol in this patient, aiming to detect early evolution to a lymphoma. Although formal management guidelines are still lacking for lymphoid cutaneous dyscrasias, treatments for early stages of cutaneous lymphomas, such as topical steroids, topical calcineurin inhibitors, sun exposure and ultraviolet phototherapy type A and B are some of the potential therapies.[1]

CONCLUSION

The patient's follow-up becomes extremely important with unusual presentations of PPD. It is critical to diagnose this

and other lymphoid dyscrasias with the proper patient care and histopathological studies, as soon as possible in order to be aware of the development of further lymphomas. Currently, there are some reported cases of patients with either PLC or PPD evolving into cutaneous T-cell lymphomas; however, we were unable to locate any data pertaining to the co-occurrence of PLC and PPD in the same individual with progression to MF.

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