Unusual Connection: Are there links between pachydermatous eosinophilic dermatitis and granulomatous slack skin?

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FOCUS AREA: Challenging Cases of Cutaneous Lymphomas **Abstract Number #163**

Introduction

Adding a layer of complexity to the vast and intricate spectrum of cutaneous lymphomas, this report explores an association not yet described in the literature. Granulomatous slack skin is a well-established variant of mycosis fungoides, but pachydermatous eosinophilic dermatitis, first reported in 1996, remains largely unexplored with few documented cases. Individually, each condition is intriguing due to its rarity, but a connection between these entities would be even more significant. The combination of these two unusual manifestations challenges conventional understanding: could these two entities be linked?

Case Report

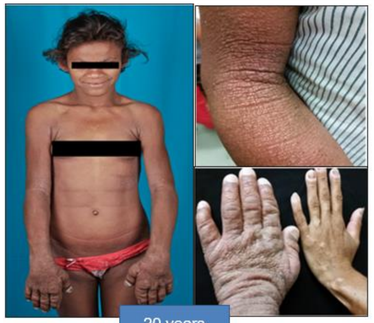

Figure 1: Intense lichenification involving 100% of the body surface area and enlargement of the extremities.

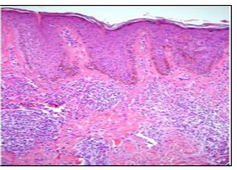

Figure 2: H&E: epidermal hyperplasia, papillary dermal fibrosis, and an inflammatory infiltrate rich in eosinophils

We describe the case of a woman who presented at the age of 20 with a ten-year history of progressive swelling of the extremities and generalized skin thickening (mainly in the hands, feet, and genital region), accompanied by intense itching. She denied a history of atopy, hospitalizations, or recurrent infectious episodes. No digital clubbing or significant dental alterations were noted. Due to pachydermia and acromegalic changes, the following investigations were carried out:

a) **serial histopathological examinations** revealed epidermal hyperplasia, papillary dermal fibrosis, and an inflammatory infiltrate rich in eosinophils;
b) **laboratory tests**: marked peripheral eosinophilia, IgE > 2500 kU/L, serum protein electrophoresis unremarkable; thyroid function, inflammatory markers, and growth hormone levels were normal;
c) **imaging studies**: X-rays of the hands and feet showed soft tissue enlargement without periostosis; cranial MRI, sella turcica X-ray, and total abdominal ultrasound were normal.

Case Report

Figure 3: reduction in skin infiltration and extremity enlargement after dapsone treatment. Slack skin was better visualized in the groin folds.

After excluding pachydermoperiostosis, acromegaly, and hypereosinophilic syndrome, the diagnosis of pachydermatous eosinophilic dermatitis was considered.

Treatment with varying doses of prednisone, antihistamines for itching, and phototherapy was initiated, but the patient responded best to dapsone, with reduction in extremity volume and improvement in itching.

However, after approximately 5 years of follow-up, the patient developed clinical features of slack skin in the bilateral inguinal region. Serial biopsies were performed, and the histopathological examinations revealed a dense subepidermal lymphocytic infiltrate, with frequent eosinophils and irregularly distributed plasma cells, associated with dermal hypervascularization, papillary dermal fibrosis and epidermal hyperplasia, along with a reduction or absence of elastic fibers. There were some atypical epidermotropic lymphocytes, which were better visualized with CD8 immunohistochemical staining.

These lymphocytes showed a CD2, CD3, CD5, CD7, CD8 phenotype. T cell receptor clonality is not avaliable in our hospital. Laboratory tests (including protein electrophoresis) showed no abnormalities, and PET-CT revealed enhanced uptake in bilateral inguinal regions, along with mildly hypermetabolic bilateral inguinal lymph nodes.

Discussion

Although a definitive diagnosis of granulomatous slack skin could not be established, it could not be ruled out. The typical clinical presentation and the dense lymphocytic infiltrate are highly suspicious for the disease. We believe that assessing T cell receptor clonality would be instrumental in defining the diagnosis. The patient has not shown improvement or progression since then and continues to be monitored.

Pachydermatous eosinophilic dermatitis is a syndrome characterized by skin thickening associated with peripheral eosinophilia and elevated serum IgE levels. Starting from an extremely rare condition, we now add inguinal slack skin to the clinical picture. The question arises whether there is an intersection between these two conditions or if they are distinct diseases coexisting in the same patient.

References: **1)** Jacyk WK, Simson IW, Slater DN, Leiferman KM. Pachydermatous eosinophilic dermatitis. Br J Dermatol 1996; 134 (3): 469-74. **2)** Salomon J, Bialynicki-Birula R, Wozniak Z, Baran E. Pachydermatous eosinophilic dermatitis. Acta Dermatovenerol Croat 2011; 19(1): 31-5. **3)** de Gracauw E, Beltraminelli H, Simon HU, Simon D. Eosinophilia in Dermatologic Disorders. Immunol Allergy Clin North Am. 2015 Aug;35(3):545-60. **4)** Guerini MB, Barbato MT, Sá NB, Nunes DH, Zeni PR. Pachydermoperiostosis: the complete form of the syndrome. .An Bras Dermatol. 2011;86(3):582-4. **5)** Navarro-Triviño FJ. Pruritus in Dermatology: Part 1 - General Concepts and Pruritogens. Actas Dermosifiliogr. 2023 Jun;114(6):512-522.