Clinical and histological differences between pediatric pityriasis lichenoides and hypopigmented mycosis fungoides.

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FOCUS AREA: Challenging Cases of Cutaneous Lymphomas **Abstract Number #174**

Introduction

Pityriasis lichenoides (PL) and cutaneous T-cell lymphoma (CTCL) are rare lymphocytic disorders in the pediatric population. Their clinical presentation is polymorphic and similar to other common hypopigmented dermatoses, such as pityriasis alba and post-inflammatory dyschromia.

Pityriasis lichenoides commonly occurs in the first decade of life, presenting in two forms with clinical and histological overlap: pityriasis lichenoides et varioliformis acuta (PLEVA), which manifests with papulo-vesicles, necrotic or ulcerative acute eruptions, and atrophic scars[1]; and a severe variant, febrile ulceronecrotic with systemic involvement known as Mucha-Habermann disease[2], along with pityriasis lichenoides chronica (PLC), an indolent rash consisting of recurrent crops of small lichenoid scaly papules and macules[1].

Hypopigmented mycosis fungoides (HMF) is the most common form of CTCL in children, characterized by hypopigmented macules and patches in sun-protected areas, with a mean age of diagnosis around 10 years and an excellent prognosis. Histologically, it typically presents with atypical lymphocytes demonstrating epidermotropism and a CD8+ T-suppressor phenotype. It is important to note that while clonal T-cell receptor gene rearrangements are common in cases of adult MF patients, data in pediatric cases remains controversial[3,4, 5].

Case Report

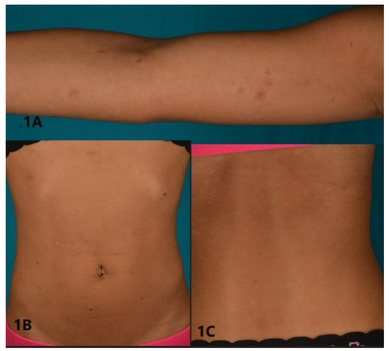

Figure 1: Pityriasis lichenoides chronica: A e B erythematous papules with a micaceous scale on the arm and abdomen C hypopigmented macules in the torso

14-year-old girl who presented with erythematous papules topped with a micaceous scale and multiple hypopigmented macules. She had no systemic symptoms. Histopathological examination revealed a lichenoid infiltration along with focal extravasation of erythrocytes, and no lymphoid atypia was observed. She was diagnosed with PLC. She has been using a weekly dose of 5 mg of methotrexate for 3 years, resulting in the remission of lesions; however, they recur when treatment is suspended.

Case Report

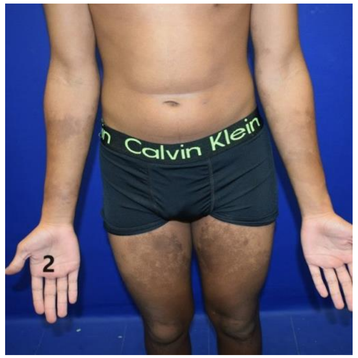

Figure 2: Hypopigmented mycosis fungoides: hypopigmented macules on the arms and thighs

14-year-old boy who presented with hypopigmented macules and patches on his arms, thighs, and gluteum. He did not exhibit lymphadenopathy or systemic symptoms. Histopathological examination revealed atypical lymphocyte infiltration with immunoexpression of CD3, CD5, and CD8, and hypoexpression of CD7, consistent with hypopigmented mycosis fungoides (HMF). He has been treated with narrow-band UVB and is nearly clear of lesions.

Discussion

PLC and PLECA represent lymphocytic dyscrasias and may share overlapping features with HMF. PLC usually persists longer than PLEVA, and persistent pigment alteration is more common in children than adults, particularly hypopigmentation. Sunlight exposure, phototherapy, topical corticoids, antibiotic therapy, and methotrexate are therapeutic options[6].

Atypical forms of PL have a recurrence of activity for a long period (>3 years), a monoclonal population of T lymphocytes, nuclear atypia, aberrant phenotypes such as CD30 expression, double positivity for C4 and CD8, loss of CD2, expression of CD56, and TCR-gamma gene rearrangement. This presentation carries an increased risk for the development of MF, even though no isolated changes are typical of MF progression. A small number of patients (5.3%) with PL have been reported to develop MF after a prolonged clinical course ranging from 3 to 11 years, with such occurrences rarely reported in children[6,7,8].

Conclusion

Although it is a challenging task, comprehensive clinical data, along with histopathological and phenotypical features, contribute to a confident diagnosis of PL and MF.

References: 1. Lam J, Pope E. Pediatric pityriasis lichenoides and cutaneous T-cell lymphoma. Curr Opin Pediatr. 2007 Aug;19(4):441–5. 2. Cumming MM, Salathiel AS, Paino MA, Delort S, Roselino AM. Doença de Mucha-Habermann (variante febril úlcero-necrótica) com acometimento mucoso exuberante: relato de caso [Febrile ulceronecrotic Mucha-Habermann disease with exuberant mucosal involvement: case report]. An Bras Dermatol. 2009 Nov-Dec;84(6):655-8. 3. Wu JH, Cohen BA, Sweren RJ. Mycosis fungoides in pediatric patients: Clinical features, diagnostic challenges, and advances in therapeutic management. Pediatr Dermatol. 2020 Jan;37(1):18-28. 4. Jung JM, Lim DJ, Won CH, Chang SE, Lee MW, Lee WJ. Mycosis Fungoides in Children and Adolescents: A Systematic Review. JAMA Dermatol. 2021;157(4):431–438. 5. Kothari R, Szepietowski JC, Bagot M, Sandhu S, Patil A, Grabbe S, Goldust M. Mycosis fungoides in pediatric population: comprehensive review on epidemiology, clinical presentation, and management. Int J Dermatol. 2022 Dec;61(12):1458-1466. 6. Borra T, Custrin A, Saggini A, Fink-Puches R, Cota C, Vermi W, Facchetti F, Cerroni L. Pityriasis Lichenoides, Atypical Pityriasis Lichenoides, and Related Conditions: A Study of 66 Cases. Am J Surg Pathol. 2018 Aug;42(8):1101-1112. 7. Sibbald C, Pope E. Systematic review of cases of cutaneous T-cell lymphoma transformation in pityriasis lichenoides and small plaque parapsoriasis. Br J Dermatol. 2016 Oct;175(4):807-9. 8. Zang JB, Coates SJ, Huang J, Vanderheid EC, Cohen BA. Pityriasis lichenoides: Long-term follow-up study. Pediatr Dermatol. 2018 Mar;35(2):213-219.