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Atopic dermatitis and mycosis fungoides in a child: an overlooked association

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Atopic dermatitis and mycosis fungoides in a child: an overlooked association.

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Manuscript text words: 641

Number of references: 5

Figures: 1

Funding sources: None

Conflict of interest: None declared

Key Words: Mycosis Fungoides, Atopic Dermatitis, Children, Pathology, Pathogenesis.

All authors read and approved the final version of the manuscript

MAIN TEXT

A 13-year-old Caucasian boy presented a 6-month history of recalcitrant hypopigmented patches/plaques on both antecubital folds and the anterior aspects of the left forearm (Fig. 1a-b). The patient presented mild skin xerosis and had atopic dermatitis (AD) in his first years of life. He had negative personal and family history of allergic rhinitis and asthma. The pediatrician prescribed a 4-week course of topical steroids (methylprednisolone aceponate 0.1% cream SID) followed by emollients. Four months later, residual hypopigmentation was observed on the flexor aspects, whereas coalescing flat-topped plaques with marginal erythema, alopecia, and scratch marks progressed on the extensor side of the left upper limb. The serological tests were within normal ranges, including complete blood and immunoglobulin counts (including IgE levels), serum protein electrophoresis, erythrocyte sedimentation rate, reactive c protein, renal and hepatic markers, antinuclear (ANA) and transglutaminase (tTG-IgA) antibodies.

A 5mm-punch biopsy showed spongiosis and a perifollicular or intrafollicular infiltration of atypical T cells with a CD3+,CD4+,CD8- immunophenotype (Fig. 1c-d).

A diagnosis of hypopigmented folliculotropic mycosis fungoides (MF) stage IA mSWAT 3 was established. The patient received 24 psoralen plus ultraviolet A (PUVA) sessions, starting with the dose of 0.5 J/cm², which was incremented each session of 0.5 J/cm² up to 6.5 J/cm². Complete clearance of skin plaques was maintained at 6-month follow-up.

MF accounts for 65% of all primary cutaneous lymphoma in the pediatric age and reaches a prevalence of about 0.5 cases per million people¹. The mean age at diagnosis is 10 years, and the male: female ratio is 1.1:1¹. Pediatric MF accounts for 5% of the total cases, and is characterized by a higher prevalence of non-classic variants, including, in order of decreasing frequency, the hypopigmented, folliculotropic, poikilodermatous and hyperpigmented forms, or a combination of these¹.

Most pediatric cases fall into early stages (IA, IB or IIA) and progression to the tumoral stage (IIB) is a rare event compared to the adult form³. In adults diagnosed as early-phase MF, the cumulative

percentage of progression to advanced stages was 21.5%, according to a recent multicentric study based on 1422 patients³. Pediatric MF prognosis does not correlate with the age of onset, and appears better than the adult form¹. A 77% response rate have been reported in pediatric cases within 1-year from treatment initiation, although the occurrence of relapse is common¹.

Compared to adult MF, pediatric MF is more often characterized by a subset of cytotoxic T-helper lymphocytes (CD3+, CD8+, CD4-). However, the most common neoplastic infiltrate of MF shows a T - helper differentiation (CD3+, CD4+, CD8-)¹.

The diagnostic delay ranges between 2-5 years, as misdiagnoses are frequent: 1/3 of pediatric MFs are initially treated as eczema^{1,2}.

The hypopigmented MF encompasses over 50% of pediatric cases¹ and could mimic AD both on clinical and pathological examinations¹.

Cases of MF occurring in atopic patients have been documented in the literature².

Even though evidence coming from the field of veterinary medicine evidenced that dogs with AD are 12 times at risk of developing MF, this association was confuted in human medicine by a large retrospective study on 157 patients affected by cutaneous T-cell lymphoma². Despite the absence of a causal relation between MF and AD², immature Langerhans cells (iLC) may promote a tolerogenic micro-environment in AD⁴. The cross-talk between malignant T-cell and iLC is crucial in MF pathogenesis, since it may trigger a possible clonal neoplastic T-cell selection⁵.

In conclusion, MF is the most common cutaneous T-cell lymphoma of the pediatric age, especially in its hypopigmented and folliculotropic variants. Due to the low prevalence of this condition in children, the diagnosis is often overlooked. The risk of not diagnosing MF becomes higher when the clinical presentation affects the most typical anatomical sites of atopic dermatitis. Although the association between MF and atopic dermatitis remains controversial, the detection of alopecic or hypopigmented plaques/patches unresponsive to standard therapy must always raise the suspicion of MF.

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Figure Legends

Figure 1: A-B: Clinical presentation of coalescing dome-shaped hypopigmented plaques with erythematous rims distributed on the left arm(A), and hypopigmented patch located on the right antecubital fold (B); the clinical pictures were obtained after the completion of a 4-week course of topical steroids and a 12 week-course of emollients. C: Giemsa stain showed spongiosis with a mononuclear infiltrate consisting of small lymphocytes, with convoluted nuclei with a peri-adnexal distribution. D: Diffused CD4+ lymphocytes could be detected at immunohistochemistry.

Authors' contribution

Dr Andrea Sechi had full access to all of the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

Study concept and design: Dr Andrea Sechi

Acquisition, analysis, and interpretation of data: Dr. Clara Bertuzzi and Prof. Annalisa Patrizi

Drafting of the manuscript: Dr. Alba Gugliemo

Critical revision of the manuscript for important intellectual content: Dr. Alessandro Pileri

Administrative, technical, or material support: Dr. Iria Neri

Study supervision: Dr. Alessandro Pileri