Pityriasis lichenoides in childhood

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Introduction and objectives

- 20% of cases of pityriasis lichenoides (PL) occur in children.
- PL has been regarded as a benign reactive process, but infrequent reports of PL evolving into cutaneous T-cell lymphoma (CTCL) and detection of T-cell clonality have led to hypothesize that PL is the benign end of a spectrum of lymphoproliferative diseases that includes lymphocytic malignancy.
- **Objectives:** 1) To characterize the epidemiological, clinical and genotypic features of PL in a series of children followed up in two third level referral hospitals. 2) To determine the evolution of PL and the risk of developing cutaneous T cell lymphoma (CTCL) in children with PL.

Methods

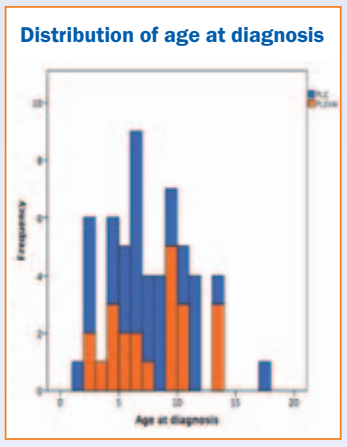
- Retrospective longitudinal study of children <18 with a histologically confirmed diagnosis of PL carried out at Hospital del Mar and Hospital Sant Joan de Déu (Barcelona) between 2006 and 2016.
- Clinical, histopathological features and treatments prescribed were collected. TCR gene rearrangements were performed in some patients. Monitoring data and risk of developing malignancy were also retrieved.

Results

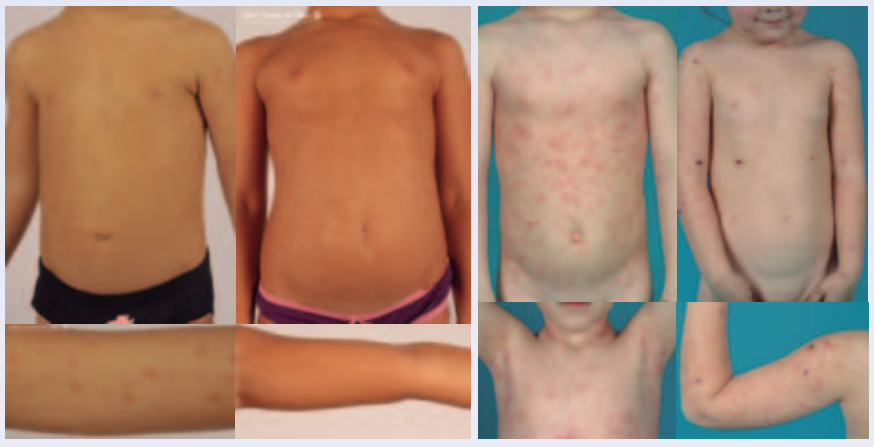
Clinical features

- 57 children (42 from Hospital Sant Joan de Déu and 15 from Hospital del Mar)
- 38 (67% boys) vs. 19 girls
- Median age at diagnosis 7.5 (1-17)
- Peaks of incidence at 7 and 9 years of age.
- 15% involvement of the face
- Pruritus 38%

Distribution of age at diagnosis

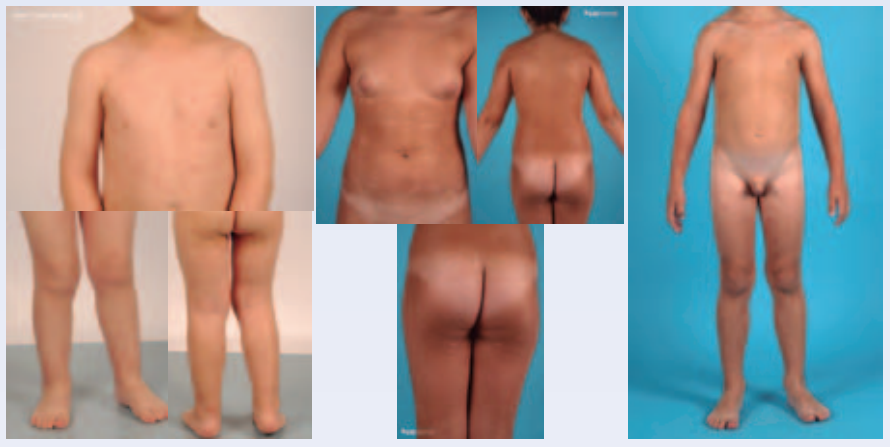


Clinical presentation



PLC 61% PLEVA 39%

Distribution pattern



Generalized (62%) Central (26%) Peripheral (12%)

Treatment

PLC

- First-line therapy: topical corticosteroids (54%)> oral erythromycin (40%)
- Second-line therapy (outbreak or non-response): topical corticosteroids (10 patients)> oral erythromycin (2 patients) > phototherapy (1 case)

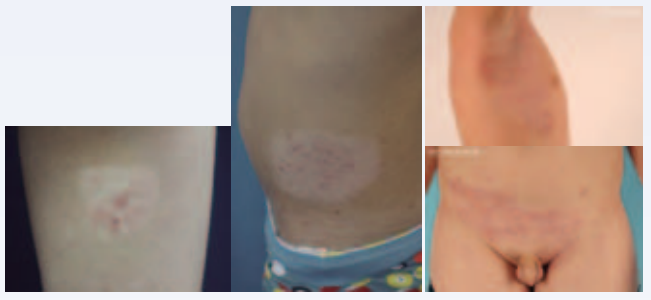
PLEVA

- First-line therapy: oral erythromycin (81%) > topical corticosteroids (18%)
- Second-line therapy (outbreak or non-response): phototherapy (7 patients) > topical corticosteroids (3 patients) > methotrexate (1 patient)

Evolution and progression to CTCL

- Median follow-up: 15 months (0,3-118 months)
- 26 losses to follow-up within the first 4 months
- 3 cases (5.2%) developed mycoses fungoides (MF) (see table and pictures).
- Risk of malignancy
  5 years: 1.8 % (IC 95% 0.3-9.4%)
  10 years: 5.6 (IC 95% 1.9-15.1%)

	Sex	Age PL	Type PL	Age MF	Location	TCR rearrangement	MF treatment
1	M	11	PLC	12	Thigh	Monoclonal TCRγ	Topical corticosteroids Photodynamic therapy Almost completed response
2	M	5	PLEVA evolution to PLC	12	Trunk and lower extremities	Oligoclonal TCRγ and TCRβ	Methotrexate Topical corticosteroids Lost to follow-up
3	F	5	PLC	9	Abdomen and axilla	Monoclonal TCRγ	Topical corticosteroids Phototherapy Lost to follow-up



Case 1 Case 2 Case 3

TCR rearrangement

- Performed in 9 patients: in 5 PLC and 1 PLEVA, TCR rearrangement was no clonal. In cases that progressed to MF, lesions typical of PL were no clonal however, in lesions suggestive of MF, rearrangement was clonal.

Discussion

- Epidemiological and clinical features were coincident with previously reported data[1,2].
- Treatments prescribed were apparently accompanied in most patients by an improvement and eventual disappearance of the lesions of 2 to 4 months. Nevertheless, it was impossible to evaluate the efficacy of the treatments because of the natural tendency of the disease to spontaneously remit.
- 3 cases evolved to CTCL (MF). Patients developed persistent large hypopigmented plaques with erythematous papules or with poilokiloderma and atrophic changes. Mean duration of the disease prior to the diagnosis of CTCL was 5 years, similar to previous data[3].
- Although most patients follow a benign course, particular attention should be given to changes in the morphology of the lesions (hypopigmentation, poikiloderma or atrophy), especially when these lesions are persistent and rule out a progression to CTCL.

1. Romaní J, Puig L, Fernández-Figueras MT, de Moragas JM. Pityriasis lichenoides in children: clinicopathologic review of 22 patients. Pediatr Dermatol. 1998;15(1):1-6.
2. Ersoy-Evans S, Greco MF, Mancini AJ, Subasi N, Paller AS. Pityriasis lichenoides in childhood: a retrospective review of 124 patients. J Am Acad Dermatol. 2007;56(2):205-10.
3. 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;175(4):807.

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