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The relevance of recognizing clinical and morphologic features of pityriasis lichenoides: clinicopathological study of 29 cases

Jandrei Rogério Markus[1], Vânia Oliveira Carvalho[2], Monica Nunes Lima[3], Kerstin Taniguchi Abagge[2], Alexandre Nascimento[4], Betina Werner[4]

1 Division of Pediatrics, ITPAC-Porto Nacional, Tocantins, Brazil
2 Division of Pediatric Dermatology, Clinical Hospital of the Federal University of Paraná (UFPR), Curitiba, Brazil
3 Division of Statistics, Department of Pediatrics, Clinical Hospital of the Federal University of Paraná (UFPR), Curitiba, Brazil
4 Division of Medical Pathology, Federal University of Paraná (UFPR), Curitiba, Brazil

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Corresponding author: Vânia Oliveira de Carvalho, Rua Richard Strauss 62, Vista Alegre. Curitiba, CEP: 80820-110, Paraná, Brazil. Tel/Fax. +55 (41) 335-3477.

Email: rcarvalho50@hotmail.com

ABSTRACT

Background: Pityriasis lichenoides (PL) is a lymphoproliferative disease of unknown origin; its diagnosis is based on clinical characteristics and confirmed by histology.

Objectives: To describe clinical and histological features of PL in 29 pediatric patients.

Materials and Methods: Retrospective descriptive study of children (patients less than 15 years old) diagnosed with PL between 1986 and 2010 at a Reference Service in Pediatric Dermatology from South Brazil.

Results: Twenty-nine PL cases were found by chart review in 24 years. Mean age of diagnosis was 8 years (22 to 178 months) and a mean time of diagnosis was 13.8 months (1 to 120 months). Twenty cases (69%) were male. Seasonal correlation was found with colder months in 62% of cases ($p<0.01$). Clinical diagnosis was pityriasis lichenoides chronica (PLC) in 25 cases, and pityriasis lichenoides et varioliformis acuta (PLEVA) in four. Itching was the main reported symptom occurring in 13 (45%). Fourteen cases had been histologically evaluated. In six, microscopic findings were consistent with PLC, in four consistent with PLEVA, and four biopsies exhibited mixed characteristics of both forms. Concordance between clinical and histological diagnosis was seen in most cases.

Conclusion: PL occurs in children and young adults, more commonly in males, and during cold months. PLC was the more frequent clinicohistologic form, and necrotic lesions characterized PLEVA. Associating clinical and histological findings is important for differentiating between PLC and PLEVA diagnosis.

Introduction

Pityriasis lichenoides (PL) is one group of inflammatory skin diseases which includes pityriasis lichenoides et varioliformis acuta (PLEVA), ulceronecrotic febrile Mucha-Habermann disease (a subtype of PLEVA which presents fever with ulcerated and necrotic lesions and other systemic symptoms), and pityriasis lichenoides chronica (PLC) [1,2].

In PLEVA, lesions begin as redish-brown macules and papules which occur in successive crops and can be asymptomatic or itchy. They normally measure 2 to 3 mm and evolve quickly to crusted purpuric necrotic lesions. Resolution can result in varioliform scarring. PLC presents papules with characteristic signs of fine scaling, and resolution with postinflammatory hypopigmentation without scarring may occur [1].

In general, PL is more common in the first ten years of life, and it is believed that 19 to 38% of cases occur in the pediatric age group [1,3]. There are several hypotheses about its pathogenesis, including dermatitis mediated by immune complexes [3-7]. It has also been considered a lymphoproliferative disease triggered by antigen stimuli such as viruses or other infectious agents because clonality and prominent CD30 positivity has been demonstrated, especially in some histological variants [3,7-11].

The objective of this study is to describe the clinical and histological characteristics of PL in the pediatric age in a reference pediatric dermatology center in south Brazil, with the aim of correlating history, clinical and microscopic findings with the classification of disease type (PLEVA or PLC).

Method

A retrospective descriptive observational study evaluated clinical and histological data from patients less than 15 years old diagnosed with PL seen at a specialized pediatric dermatology division of a Public Hospital in south Brazil from 1986 to 2010. One pediatric dermatologist at this center evaluated all patients. The institution's Research Ethics Committee approved this study.

Medical records of PL patients were reviewed for the following variables: gender, age at start of signs or symptoms, history of infections in the six months before lesions, medication history, family history of similar lesions, disease duration, skin lesion distribution, treatments, associated symptoms, and pigment alterations and their duration after improvement.

Two researchers reviewed photographic documentation, when available, to confirm diagnosis and classify the disease into the two clinical forms—PLEVA or PLC. Those without photographic evidence were classified according to medical records. Cases were classified as PLC when there were residual hypochromic lesions, erythematous papules, and exfoliation with signs of fine scaling, and PLEVA when there was necrosis, hematic crusts, and the presence of atrophic scarring or associated fever.

Patients who had undergone skin biopsies had their slides reviewed and the following histological parameters evaluated. In the epidermis: acanthosis, spongiosis, lymphocyte exocytosis, parakeratosis, necrotic keratinocytes and scale crust. And in the dermis: papillary edema hemorrhage, perivascular lymphocytic infiltrate and atypical lymphocytes. These criteria were evaluated for the presence or absence in each case, and when present as discrete, moderate, or accentuated in an attempt to differentiate between PLC and PLEVA. Cases that presented a moderate or accentuated epidermal or dermal inflammatory component; and or frequent necrotic keratinocytes; and or scale crust were classified as the acute form (PLEVA). Cases with more subtle histological alterations or with their absence were classified as the chronic form (PLC). Cases where parameters from both groups were found were classified as the mixed form.

The summary measurements used in the descriptive statistics were mean, standard deviation, median, minimum and maximum values, and frequencies depending on type of variable studied. Differences between ages were studied using the Mann-Whitney test considering a significance level of 5%. Data were analyzed with the aid of software package Statistica 7.1 (Statsoft).

Results

Twenty-nine PL cases were evaluated; median age was 96 months (22 to 178 months). Age at the start of symptoms had peaks at 8 years, followed by 12 and 14 years. Mean time between start of symptoms and diagnosis was 1.15 years (1 month to 10 years).

Twenty patients were male (69%). In the comparison between gender and age at start of symptoms, median age at the start in males was 8.3 years (1 to 14.1 years) and in females 8.8 years (1.6 to 14.8 years; p=0.5). Disease start predominated in winter and autumn months (Figure 1).

Cutaneous alterations were present in all patients at examination. The most frequently observed lesions were papules (n=27), hypopigmentation and scaling (n=19) and

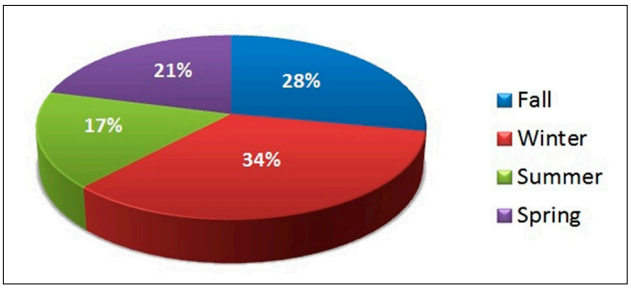

Figure 1. Distribution of patients according to season of the year that symptoms started. [Copyright: ©2013 Markus et al.]

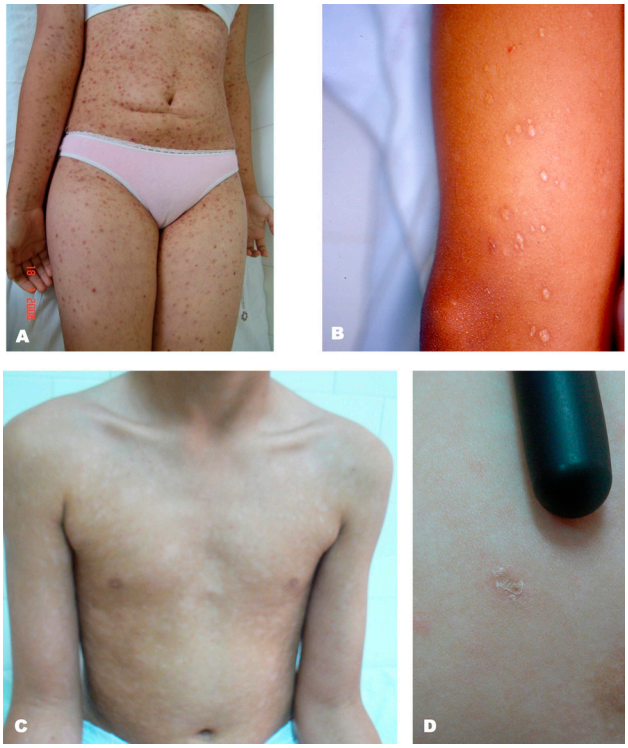

Figure 2. (A) PLEVA—2 to 3 mm reddish-brown purpuric papules. (B) Varioliform scarring. (C) PLC—erythematous papules with residual macules of postinflammatory hypopigmentation. (D) Papule with a fine scaling that detaching from the periphery to the center (PLC). [Copyright: ©2013 Markus et al.]

fine scaling (n=13). There was also erythema (n=12), crusts (n=12), hyperpigmentation (n=5), necrosis (n=4), and purpuric lesions (n=3). Classification by lesion distribution was of a central form, where lesions were found on the trunk and proximal limbs in 14 (48%), followed by diffuse (generalized lesions) in 13 (45%), and peripheral, where only the extremities presented lesions, in 2 (7%) cases.

As for symptoms, 15 (52%) patients were asymptomatic, 13 (45%) had itching, and one fever. Clinical classification revealed 4 PLEVA (Figure 2A and 2B) and 25 PLC (Figure 2C and 2D).

Histological evaluation was possible in 14 patients; six (43%) presented histology consistent with the chronic form (PLC), four (28.5%) with the acute form (PLEVA), and four (28.5%) presented histology with characteristics of both forms and were classified as mixed. Comparing histology and clinical diagnoses showed that the six cases with PLC histology also had the same clinical diagnosis. The four mixed histology cases clinically presented as PLC. In the four acute histology (PLEVA) cases, one was clinically classified as PLC (Figure 3A and 3B).

Follow-up was lost in 11 cases. Of the 18 followed up, improvement was seen in 15, of which 11 had recevied erythromycin or tetracycline and four were treated with moisturizing cream until returning after biopsy, when they presented disease remission without the need for specific treatment.

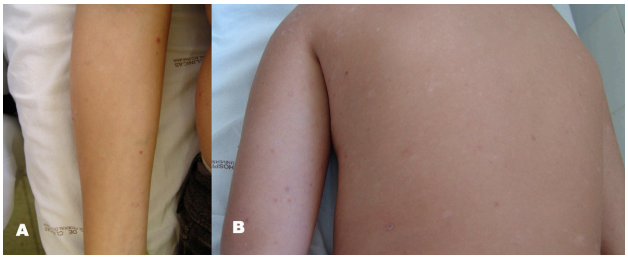

Figure 3. (A) Erythematous papules with a fine scaling without necrosis. (B) Hypochromic macules on the trunk. [Copyright: ©2013 Markus et al.]

Mean evolution time to improvement in these 15 patients was seven months (1 to 96 months). Three cases still had lesions at last evaluation and their mean follow-up time was 15 months.

Discussion

Pityriasis lichenoides is an uncommon skin disease with few studies on its clinical and histological presentation, especially in the pediatric age.

Mean age at the start was 8 years, with peaks at 8, 12, and 14 years. This varies depending on author between 5 and 9 years [8,12]. It predominated in males (69%), agreeing with literature where values range from 53% to 60% [8,12,13].

This dermatosis is believed to be seasonal, predominantly in autumn and winter, as seen in 62% of cases in this study; Ersoy-Evans et al. found this in 65% while Wahie et al. did not observe seasonal variation [8,13]. Despite the study having the limitation of being retrospective, the question exists whether there really are fewer cases in the warm months, or whether exposure to the sun could be an improvement factor for symptoms and consequently fewer patients seek for medical attention in these months. Another possibility is that viral infections, which are prevalent during cold months, could be triggers [7,14].

Most patients presented with papules in the first evaluation, followed by residual hypopigmentation and fine scaling. These clinical findings are compatible with PLC. Necrotic lesions, which are feature of PLEVA, were observed in 4 cases, which is less than in the literature [1,4].

In lesion distribution, the majority of lesions were located in the central body region. Literature reports a predominance of the diffuse form (skin lesions spread in the entire body) in 70% of cases [8]. It must be stressed that classification by lesion distribution is observer dependent, considering the highest lesion concentration, allowing a patient with the central form to have some peripheral lesions. This brings into question the validity of using topographical classification for PL.

The commonest symptom was itching, but most patients were asymptomatic, differently to Ersoy-Evans et al., who reported that most patients had pruritus. Symptom evaluation could have been underestimated due to the nature of the retro-

spective study; another possibility is the question of a subjective symptom, causing variations between different populations. Fever was reported in one patient with PLEVA diagnosis [8].

Follow-up is important to determine clinical improvement and adherence to prescribed treatment. Follow-up was abandoned in 38% of patients; this also occurred in vitiligo patients in an earlier study at the same service [15], probably reflecting a characteristic peculiar to the type of population seen in this center. In most cases where evaluative follow-up was possible, there was clinical improvement and erythromycin or tetracycline treatment was effective, therefore this is the treatment of choice due to its anti-inflammatory effect and immunological control [8,12].

As in the clinical study, histology showed a predominance of parameters considered more characteristic of the chronic form. Microscopic findings described in PL are controversial. Some studies show a predominance of PLEVA, in 57.3% of patients [8], others show PLC in 72% [12]. This probably reflects the diverse range of clinical presentations of the disease, which can characteristically exhibit lesions in different stages of evolution. There was one case where clinical and histological findings disagreed; this probably occurred due to the choice of lesion submitted for biopsy, which presented more accentuated erythema and scaling. The microscopic aspect actually showed the characteristics of the evolution phase of the lesion that was analyzed and not necessarily the disease of the patient as a whole. This reinforces the need to associate clinical and histological parameters in classifying the disease, it being advisable to not base PL classification just on microscopic aspects.

Conclusion

Pityriasis lichenoides in the pediatric age group begins around 8 years of age and is more frequent in boys. PLC was prevalent and a delay in diagnosis was seen reinforcing the need for knowing the clinical characteristics of this dermatosis. Disease seasonality could have implications in physiopathology, but needs evaluation with a larger number of cases for confirmation. Cutaneous biopsy can be very valuable in definitive diagnosis, however morphological findings depend directly on the evolutionary phase of the lesion chosen for analysis.

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