ORIGINAL ARTICLE

Pityriasis Lichenoides in Thai Children: A 10-Years Review of Clinical and Treatment Outcome

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Background: Pityriasis lichenoides (PL) represents a unique group of inflammatory dermatologic conditions. Distinct skin manifestations typically lead to diagnosis, which is confirmed through histopathology in most cases. Treatment for this group of conditions varies. Disease progression is chronic in most cases, though it is generally benign.

Objective: To investigate epidemiology, clinical subtypes, treatment, and disease progression of PL in children.

Materials and Methods: A retrospective data collection from the medical records of patients diagnosed with PL at the National Children's Health Institute during a ten-year period, between January 1, 2012, and December 31, 2022, was accomplished.

Results: In the present study, there were 43 patients, and the male-to-female ratio was 1.3:1. The most common age groups at onset of PL was 4 to 7 years old. Ten cases (23.3%) of pityriasis lichenoides et varioliformis acuta (PLEVA) and 33 cases (76.7%) of pityriasis lichenoides chronica (PLC) were identified. All cases were confirmed by skin biopsy. There were two cases of febrile ulceronecrotic Mucha-Habermann disease (FUMHD). Systemic treatment included erythromycin in 95.3%, prednisolone in 9.3%, and methotrexate in 9.3%. The duration of illness of PLC ranged from 2 to 54 months, with an average of 7±4 months, whereas in PLEVA's disease duration ranged from 1 to 20 months, with an average of 4±2 months ($p<0.05$). Dyspigmentation was predominantly observed in PLC, whereas varioliform scarring was more common in PLEVA ($p<0.05$). Among all patients diagnosed with PL, one patient (2.3%) developed mycosis fungoides as cutaneous T-cell lymphoma, during follow-up.

Conclusion: PL is typically a benign and chronic condition, though it may be life-threatening in rare cases. Diagnosis is primarily based on clinical presentation and histopathological examination of skin biopsies, although it can be challenging and subject to delays. Prognosis is generally favorable; however, ongoing monitoring is essential.

Keywords: Pityriasis lichenoides; Pityriasis lichenoides et varioliformis acuta (PLEVA); Pityriasis lichenoides chronica (PLC); Febrile ulceronecrotic Mucha-Habermann disease (FUMHD)

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Pityriasis lichenoides (PL) is an uncommon dermatologic condition. It affects children and adults, with the incidence of one in 2,000 people per year, with almost 20% of cases occurring in children[1-3]. Due to it is uncommonness, they are usually misdiagnosed as arthropod bite reactions, other viral exanthems such as varicella infection, Gianotti-Crosti syndrome, erythema multiforme, pityriasis rosea, guttate psoriasis, vasculitis, and secondary syphilis. Although the exact etiology is unknown, some believe that it is a T-cell-mediated reaction that may be precipitated by an acute infection particularly *Toxoplasma gondii*, *Mycoplasma*, *Staphylococcus*, Epstein-Barr virus, *Cytomegalovirus*, and parvovirus B19[3].

PL presents in two distinct forms, Pityriasis lichenoides et varioliformis acuta (PLEVA) and pityriasis lichenoides chronica (PLC). Both forms can coexist in the same patient[4]. PLC is more commonly observed in children and young adults, typically presenting as scattered red papules and plaques with centrally adherent scales. It generally follows a more indolent clinical course compared to PLEVA, developing slowly and often exhibiting a waxing-and-waning pattern over several years. In most cases, PLC leaves behind hyperpigmented or hypopigmented macules (Figure 1). In contrast, PLEVA lesions typically present with an abrupt onset of recurrent crops of small, erythematous pseudovesicles of 2 to

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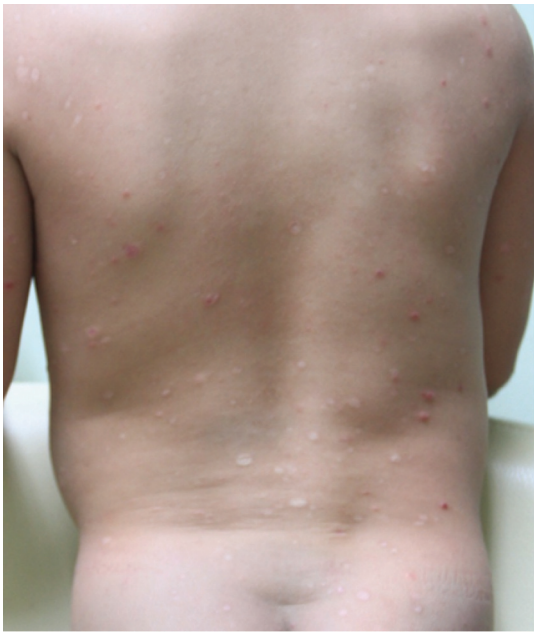

Figure 1. PLC lesions as scattered red papules and plaques with centrally adherent scales and hypopigmented macules on the trunk.

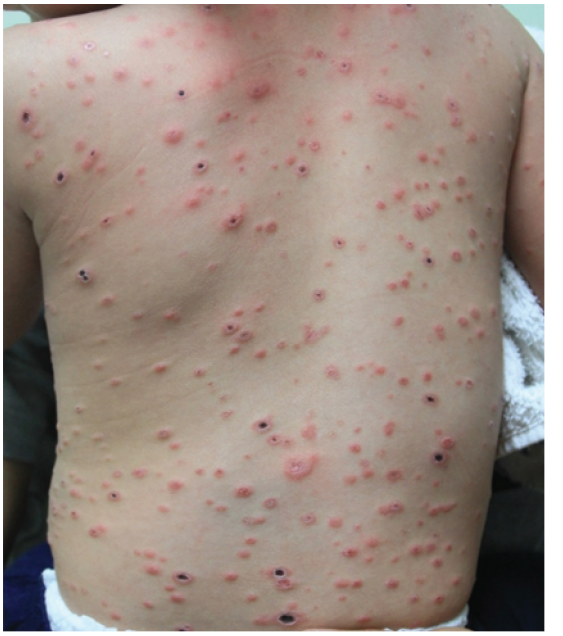

Figure 2. PLEVA lesion as crops of small, erythematous pseudovesicles with central areas of necrosis on the trunk.

3 mm in diameter, with central areas of necrosis. These lesions may leave scars, including chickenpox-like or varioliform scars (Figure 2). Febrile ulceronecrotic Mucha-Habermann disease (FUMHD), also known as PLEVA fulminans, is a severe subtype of PLEVA associated with fever. In FUMHD, patients develop cutaneous lesions resembling those of PLEVA, which progress rapidly into large ulceronecrotic lesions, accompanied by high fever and other systemic symptoms. This form can be life-threatening[5,6].

Currently, there are no standard guidelines for the treatment of PL. First-line treatment usually includes topical corticosteroids and oral antibiotics. Erythromycin is widely used as an anti-inflammatory in children and adults because it is effective, well tolerated, and accessible[7]. Immunosuppressants and phototherapy are also effective treatments for PL[8]. In patients with extensive lesions or those who do not respond to oral antibiotics, phototherapy, particularly narrow-band UVB and PUVA, is preferred[9].

In cases that are refractory to initial therapy or present with severe symptoms, consideration should be given to immunosuppressive agents such as methotrexate, cyclosporine, or systemic steroids, which have the potential to mitigate disease progression[10,11]. However, caution is warranted in the use of these agents due to their significant potential for adverse effects.

Material and Methods

The present study was a retrospective study aimed to investigate and compile data on patients diagnosed with PL based on medical records at Queen Sirikit National Institute of Child Health between January 1, 2012, and December 31, 2022. One forty-three cases were included in the analysis.

The authors reviewed demographic data including age, gender, type, disease onset, duration, treatment modalities. Descriptive statistics represented as percentages, mean, standard deviation, and range were used. Differences between groups for continuous variables were analyzed using independent sample t-tests. Comparative analysis of categorical data entailed the utilization of either the chi-square test or Fisher's exact test. Data was analyzed with IBM SPSS Statistics, version 23.0 (IBM Corp., Armonk, NY, USA) with the p-value at a significant level of 0.05.

The present study was approved by the Research Ethics Review Committee of the Queen Sirikit National Institute of Child Health, QSNICH IRB, approval REC.055/2567.

Results

In the present study, 43 patients diagnosed with PL were included, comprising 25 males and 18 females, with a male-to-female ratio of 1.9:1. The most common age group at onset was 4 to 7 years. The mean age at symptom onset was 66 months,

Table 1. Clinical data of pityriasis lichenoides patients (n=43)

Clinical subtype	PLC (n=33)	PLEVA (n=10)	p-value
Age (months); mean (range)	77 (10 to 152)	65 (12 to 121)	0.219
Symptom; n (%)			
Pruritus	14 (42.2)	3 (33.3)	0.480
Fever	1(3.0)	3 (33.3)	0.030
Hepatomegaly	0 (0.0)	2 (20.0)	0.050
Dyspigmentation; n (%)	33 (100)	6 (60.0)	0.001
Varioliform scar; n (%)	0 (0.0)	2 (20.0)	0.050
Disease duration (months); mean (range)	7 (2 to 54)	4 (1 to 2)	0.001

PCL=pityriasis lichenoides chronica; PLEVA=pityriasis lichenoides et varioliformis acuta

with a range of 10 to 152 months. The majority of patients (85.4%) had no significant medical history. Six patients (13.9%) had comorbidities, including one case of allergic rhinitis, two cases of obesity, and three cases of Kawasaki disease. Ten cases (23.3%) of PLEVA and 33 cases (76.7%) of PLC were identified, all of which were confirmed by biopsy. One patient initially diagnosed with PLC later evolved into PLEVA. Disease distribution was diffuse in all cases, affecting both PLEVA and PLC patients.

Among PLC patients, the mean age at onset was 77±35 months, with a range of 10 to 152 months. The average duration from symptom onset to diagnosis was 3±2 months, with a range of 0 to 32 months. Associated symptoms included 14 patients with pruritus (18%) and one fever (1.3%). Pigmentary changes, or hypopigmentation, were observed in 33 patients (42.8%) (Figure 3), with no cases of scarring. Thirty-two patients (96%) were treated with topical steroids, and 32 patients (96%) received oral erythromycin. Two patients (6%) were treated with prednisolone and methotrexate.

Among PLEVA patients, the mean age at onset was 65±48 months, with a range of 12 to 121 months. The average duration from symptom onset to diagnosis was 1.5±1 month with a range of 0 to 9 months. Associated symptoms included three patients with pruritus (30%), three fever (30%), and two hepatomegaly (20%). Six cases (60%), showed pigmentary changes, and two cases (20%) developed varioliform scars. Nine patients (90%) were treated with topical steroids, nine patients (90%) received oral erythromycin, and two patients (20%) were treated with systemic corticosteroids and methotrexate (Table 1).

The authors also reported two cases of FUMHD, a severe yet rare variant of PLEVA. A 10-year-old boy presented with a 10-day history of abrupt onset, rapidly spreading erythematous

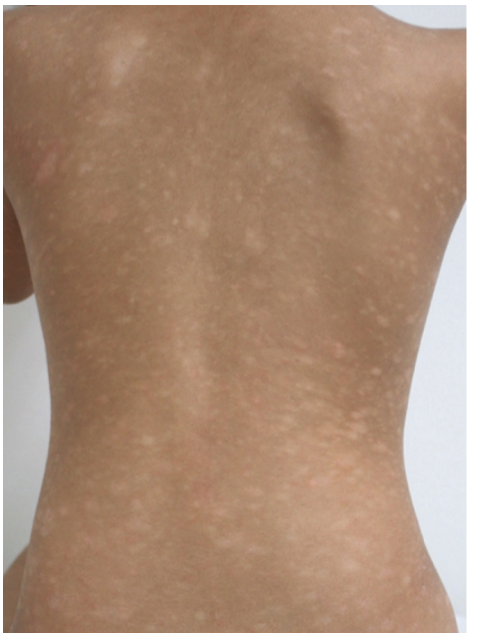

Figure 3. Generalized hypopigmented macules found on PLC patient.

macules, papules, and vesicles that gradually evolved into centrally ulceronecrotic and crusted lesions. The eruption initially appeared on the trunk, with subsequent extension to the extremities. The patient reported mild pain and pruritus at the affected sites and exhibited symptoms of general malaise. He was initially diagnosed with a bacterial skin infection at a local hospital and treated with oral clarithromycin and ciprofloxacin. However, the eruption continued to disseminate, with a maximum body temperature of 38.9°C. There was no personal or family history of dermatologic diseases. Physical examination revealed abundant erythematous macules, papules, papulovesicles, and papulopustules with central hemorrhagic necrosis. Some of the hemorrhagic necrotic centers were covered with thick crusts, while

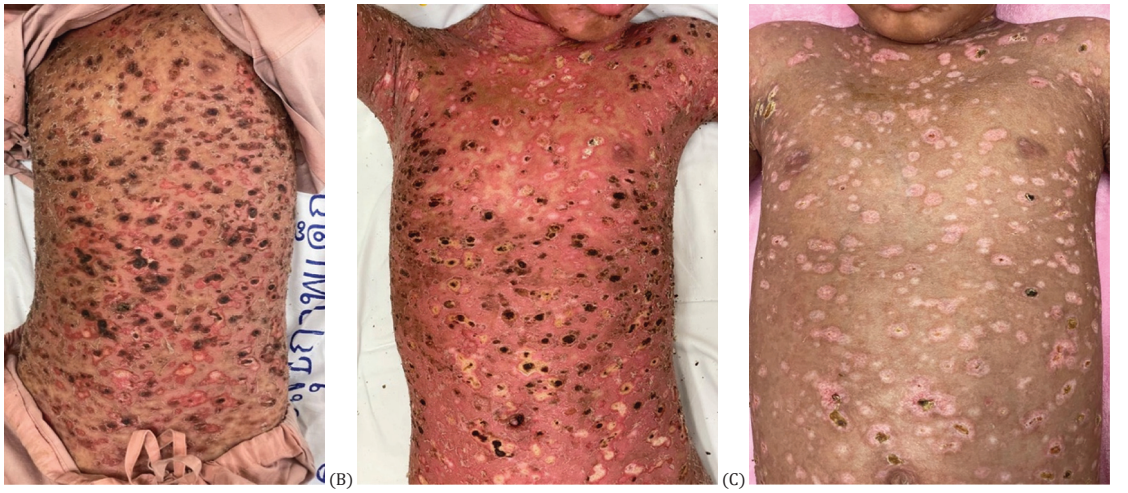

Figure 4. (A) Generalized ulceronecrotic papules and plaques on first day of admission. (B) On hospital day 10, most lesions on the patient's trunk were crusted with erythroderma. (C) on hospital day 16, most lesions evolved into atrophic scars, and left residual erosions after the crusts peeled off.

others had developed erosions and ulcers (Figure 4A). Enlarged lymph nodes were palpated in both axillae. Laboratory findings at admission showed leukocytosis at 9.6 ×10⁹/L, with 71% neutrophils. Repeated blood cultures were sterile, while skin swab cultures were positive for *Staphylococcus epidermidis*. Skin biopsy revealed focal epidermal parakeratosis, irregular acanthosis, focal necrosis, vacuolar degeneration of basal cells, and lymphohistiocyte infiltration in the dermis. FUMHD was diagnosed based on the rapid progression of the condition, the development of ulceronecrotic lesions, and the histopathological findings. The patient was treated with oral erythromycin, intravenous methylprednisolone at 1.5 mg/kg daily, cloxacillin, and ceftazidime. Regardless of the prompt treatment, the patient's condition deteriorated, and new lesions continued to develop together with generalized erythroderma (Figure 4B). His temperature suddenly spiked to 40.2°C on hospital day 10. Consequently, the dose of methylprednisolone was increased to 2.3 mg/kg/day, and methotrexate was initiated at 0.3 mg/kg weekly. Simultaneously, antibiotics were switched to meropenem and vancomycin. Following the initiation of methotrexate, the patient's cutaneous condition stabilized. By hospital day 26, most lesions on the trunk had evolved into atrophic scars, and lesions on the extremities either crusted or left residual erosions after the crusts peeled off (Figure 4C). The patient was discharged on oral prednisolone at 15 mg daily, which was gradually tapered over four weeks. Methotrexate at 5 mg weekly, was continued for a total duration of four months, with a gradual tapering of the dose. There were no new eruptions or signs of relapse during the 6-month follow-up period.

Among the 43 patients with PL, 40 patients (93%) experienced no treatment-related complications. One patient (2.3%) developed nausea and vomiting after receiving oral erythromycin. Additionally, steroid-induced acne was diagnosed in two patients (4.6%) following topical steroid treatment. In the follow-up of treatment outcomes, the duration of illness in PLC ranged from 2 to 54 months, with an average of seven months. In contrast, the duration of illness in PLEVA ranged from 1 to 24 months, with an average of four months. Among all patients diagnosed with PL, one patient (2.3%) developed mycosis fungoides as cutaneous T-cell lymphoma, during follow-up, while no patients developed lymphomatoid papulosis.

Discussion

PL is a chronic dermatological condition of uncertain etiology. It is relatively rare, affecting both children and adults, with a higher prevalence in the pediatric population. Although certain clinical findings are characteristic of PL, its diagnosis can be challenging and is often delayed.

Onset has been reported as early as eight months of age, with symptoms persisting up to 15 years. Peaks incidence is typically observed between 2 and 3 years, and 5 and 7 years of age[12]. Some studies suggest an increased prevalence in older age groups, particularly at 5- and 10-year-old[2]. In the present study, the most common age group at onset was 4- to

7-year-old. The mean age at onset of symptoms was 66 months, with a range of 10 to 152 months.

Forty-three patients were identified with PL, comprising 23 males and 18 females, yielding a male-to-female ratio of 1.3:1. This slight predominance of males is consistent with the findings of Ersoy-Evan et al.[12] and Koh et al.[13], who reported male-to-female ratios of 1.3:1 and 1.5:1, respectively. PLC was more prevalent than PLEVA, with 78% and 22% of cases, respectively. One patient initially diagnosed with PLC subsequently evolved into PLEVA. These findings support the concept that PL, encompassing both PLEVA and PLC, represents a continuous spectrum disease with variable morphological manifestations[12,14].

In the cohort under study, the majority of patients, comprising 35 individuals (85.3%) had no significant medical history. In contrast, six patients (14.6%) had documented medical conditions. Specifically, one patient had a history of allergic rhinitis, two patients were diagnosed with obesity, and three patients had a prior history of Kawasaki disease. The duration from symptom onset to diagnosis was shorter in patients with PLEVA compared to those with PLC, averaging one point five and three months, respectively. This finding aligns with the study by Zang et al. (2020), which demonstrated that the time from symptom onset to diagnosis was shorter in PLEVA patients than in PLC patients[15]. This discrepancy may be attributed to the characteristic rash presentation in PLEVA, which is often more severe and associated with a higher prevalence of additional symptoms. As a result, patients with PLC may seek medical evaluation later than those with PLEVA.

In the present study, clinical manifestations of PL revealed pruritus in both the PLC and PLEVA groups. Additionally, systemic symptoms such as fever and hepatomegaly were significantly more common in PLEVA patients compared to PLC patients ($p<0.05$). These findings are consistent with those of Ersoy-Evans et al.[12], who reported a higher prevalence of systemic symptoms in PLEVA patients compared to PLC patients at 13% versus 4.3%, respectively. The authors also observed a higher prevalence of hypopigmentation or dyspigmentation in the PLC subtype ($p<0.05$). However, varioliform scars were found only in the PLEVA subtype.

Currently, there are no clear standard guidelines for the treatment of PL. However, several research studies have found that oral antibiotics, particularly erythromycin, can reduce disease progression. It is proposed that the efficacy of erythromycin is due to its anti-inflammatory effect[16,17]. In the present study, patients diagnosed with PLEVA and PLC were treated similarly, predominantly with topical steroids and oral erythromycin. Additionally, one case (3%) of PLC subgroup experienced uncontrolled symptoms while on erythromycin and topical steroids, necessitating the use of prednisolone and methotrexate for symptom management. Oral erythromycin was generally well tolerated. Gastrointestinal upset was rarely reported, and no significant adverse effects were observed, in the present study, only one case (2.3%) with mild GI side effect of nausea and vomiting[1,12,16,18]. Therefore, they are commonly recommended as first-line therapy.

The authors also reported a rare and severe variant of PLEVA, FUMHD, also known as PLEVA fulminans. This condition presents with typical PLEVA lesions but rapidly progresses to a large ulceronecrotic appearance, accompanied by high fever and systemic symptoms, which can be life-threatening. The patient was successfully treated with a combination of methylprednisolone and methotrexate. Although no consensus has been reached regarding the first-line therapy for FUMHD due to its rarity, a wide variety of treatment modalities have been suggested, including systemic steroids, methotrexate, cyclosporine, dapsone, and intravenous immunoglobulin (IVIG). A literature review has shown that methotrexate has been used in 15 pediatric FUMHD cases, with successful outcomes in 13 patients when combined with systemic steroids[19-21]. However, as seen in the current case, where initial systemic steroid therapy failed, methotrexate demonstrated excellent results.

The duration of illness in patients with PL ranged from 1 to 54 months, with a mean duration of 8.9 months. Specifically, the average duration of illness was significantly shorter in PLEVA patients compared to PLC patients at four months versus seven months, respectively. These findings are consistent with those of Ersoy-Evan et al.[12], who reported shorter disease durations in PLEVA patients compared to PLC patients at 18 months versus 20 months, respectively, although the durations in their study were longer than those observed in the current study.

PL typically follows a benign course, with spontaneous resolution being common. However, some reports have linked PLEVA and PLC to the development of cutaneous T-cell lymphoma upon long-term follow-up, ranging from four to seven years[22-25]. In the present study, no cases of transformation into lymphomatoid papulosis were

observed, but one patient diagnosed with PLC upon treatment and one-year follow-up subsequently exhibited increased widespread white patches on the legs and trunk. Upon repeated biopsy, this patient was diagnosed with mycosis fungoides. The relationship between PL and cutaneous T-cell lymphoma remains inconclusive and controversial. Forston et al.[23] reported two pediatric cases initially diagnosed with PLEVA at ages two and seven years, which later progressed to mycosis fungoides after several years, ten and five years, respectively. Additionally, Tomasini et al.[25] reported a 17-year-old female patient diagnosed with mycosis fungoides, who had a history of PLC at age 11. However, a study by Ersoy-Evans et al.[12], which compiled data from 124 pediatric patients with PL over a 10-year period, did not identify any cases of transformation into cutaneous lymphoma or lymphomatoid papulosis.

Conclusion

Awareness of dermatological manifestation of PL, especially in severe subtype fulminant variant of PLEVA, is crucial for early diagnosis, thorough evaluations, and appropriate treatment selection for these patients.

What is already known about this topic?

PL is an uncommon dermatologic condition in children. Disease progression is usually chronic, but they are also benign.

What does this study add?

Awareness of dermatologic manifestation of PL, especially in severe subtype fulminant variant of PLEVA, is crucial for early diagnosis, thorough evaluations, and appropriate treatment selection for these patients.

Conflicts of interest

The authors declare no conflict of interest.

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