Pityriasis Lichenoides in Children

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We studied 24 cases (13 boys and 11 girls) of childhood pityriasis lichenoides to determine clinical differences between childhood form and adult form.

Nine patients complained of pruritus and 2 patients complained of fever. Facial involvements were noted in 6 cases, palm-sole involvements in 4 cases. The majority of our patients were treated with erythromycin or tetracycline and a few patients with UVB phototherapy. After treatment 15 cases were cleared of the lesions in which 8 cases were pityriasis lichenoides et varioliformis acuta (PLEVA) and 7 cases were pityriasis lichenoides chronica (PLC), and their average durations of the disease were 5.9 months (range, 2-14 months) for PLEVA and 12.3 months (range, 2-32 months) for PLC. It seemed that there were more subjective symptoms and more atypical distributions of skin lesions in childhood form, and clinical course was shorter in childhood form.

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Pityriasis lichenoides (PL) is a self-limited, asymptomatic skin disorder of unknown origin. The disease occurs mainly in adolescents and young adults but can affect children and has a slight male predilection. The disease is usually divided into an acute form called pityriasis lichenoides et varioliformis acuta (PLEVA) also known as Mucha-Habermann disease, and a chronic form called pityriasis lichenoides chronica (PLC).

The acute form is characterized by crops of round, reddish brown macules and papules that evolve into vesicular and necrotic lesions; residual varioliform scarring with skin discoloration (hypo- or hyperpigmentation) may occur. The acute eruption may be generalized but is usually confined to the trunk and the proximal aspects of the extremities. This form is said to run a course of a few weeks to several months[2].

The chronic form features reddish brown papules with adherent central scales that tend to separate spontaneously. It clears without scarring, showing only transient skin discolorations. The eruption involves the trunk and the proximal aspects of the extremities, but the course of the disease may be as long as a few years[2].

There are few reports of childhood form of PL in which the clinical aspects, therapeutic responses, and detailed clinical courses are mentioned in comparison with adult form of PL. The aims of this study are to know the prevalence rate of PL in children and the clinical course or duration of disease after treatment, and to find clinical characteristics of childhood PL as compared with adult form of PL.

PATIENTS AND METHODS

All patients under 15 years old diagnosed as PLC or PLEVA in the Department of Dermatology of Seoul National University Hospital during 10 years between 1980 and 1990 were examined. A total of 24 patients were included in the study. All cases were diagnosed by history, clinical appearance, and histopathologic findings. The majority of these patients were treated with 4-to 10-week courses of oral antibiotics (erythromycin or

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tetracycline) and a few patients whose lesions resisted to the antibiotic therapy with 4-to 6-week courses of UVB phototherapy. Some patients were treated with antihistamine, DDS, and prednisolone. Clearing after treatment was defined as the disappearance of scale and erythema of all skin lesions.

Thirty nine adult patients diagnosed as PLC or PLEVA between 1986 and 1990 were also included in the study to make our comparison with pediatric patients more reliable because our Pedatric Department of Dermatology has been officially separated from main hospital since 1986. These adult patients received treatments similar to those of pediatric patients.

The clinical findings including subjective symptoms, locations of the lesions, clinical courses, and therapeutic responses were analyzed regardless of their types of disease, that is, PLEVA or PLC to compare these clinical characteristics with those of adult form of PL.

RESULTS

Between 1986 and 1990 the numbers of childhood PL were 16 among the total 7499 patients and those of adult PL were 39 among the total 20723 patients. Therefore the prevalence rate of childhood PL was estimated to be 0.21%. This prevalence rate was quite similar to that of adult, 0.19% (Table 1).

When 24 childhood PL patients were subdivided into 2 forms, 9 patients were considered to have PLEVA and 15 to have PLC. 13 patients were boys and 11 girls and the sex ratio was almost same. Age at first diagnosis was between 1 year and 14 years (mean 8.3 years).

When 39 adult PL patients were subdivided into 2 forms, 16 patients were considered to have PLE-

Table 1. Estimated prevalence rate of PL

Year		No. of Pt. Childhood PL	Total Pt.	Adult PL	Total Pt.
1986	PLC	1		6	
	PLEVA	2		1	
1987	PLC	1		3	
	PLEVA	2		4	
1988	PLC	2		2	
	PLEVA	1		7	
1989	PLC	1		6	
	PLEVA	2		6	
1990	PLC	1		2	
	PLEVA	3		2	
Total		16 (16/7499=0.21%)	7499	39 (39/20723=0.19%)	20723

Table 2. Subjective symptoms in childhood and adult form of PL

Symptoms	No. of childhood Pt(%)	No. of adult Pt(%)
Pruritus	9(37.5)	12(30.8)
Fever	2(8.3)	—
None	13(54.2)	27(69.2)
Total	24	39

Fig. 1. Erythematous crusted macules and papules on the face and palms in a childhood PLEVA case.

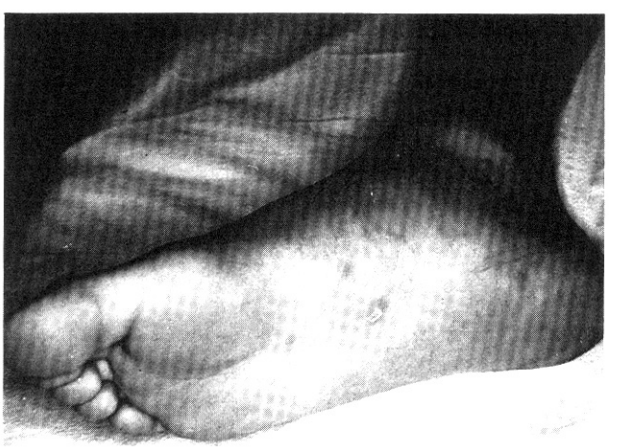

Fig. 2. Erythematous scaly macules and papules on the right sole in a childhood PLC case.

Table 3. Distributions other than trunk and extremity

Site of lesions	No. of childhood Pt(%)	No. of adult Pt(%)
Face	6(25.0)	3(7.7)
Palm, sole	4(16.7)	3(7.7)
Scalp	1(4.2)	1(2.6)
Mucous membrane	0	0
Total	24	39

Table 4. Duration of disease of childhood PL after treatment

No. of patients	Duration of disease	
Cleared	15	
PLEVA	8	5.9 months(2-14)
PLC	7	12.3 months(2-32)
Persistent or recurrent	9	
PLEVA	1	51 months
PLC	8	32.6 months(19-58)

VA and 23 to have PLC. 21 patients were males and 18 females and the sex ratio was almost same.

While 9 childhood patients complained of pruritus and 2 fever, 12 adult patients complained of pruritus (Table 2). In childhood patients facial involvements were noted in 6 cases, palm-sole involvements in 4 cases (Fig. 1, 2) and scalp involvement was noted in 1 case, but in adult patients facial involvements were noted in 3 cases, palm-sole involvements in 3 cases and scalp involvement was noted in 1 case (Table 3).

In childhood patients 15 cases were cleared of the lesion after treatment in which 8 cases were PLEVA and 7 cases were PLC. The average duration of disease in the 8 PLEVA cases was 5.9 months (range, 2-14 months) and that in the 7 PLC cases 12.3 months (range, 2-32 months), respectively. Only 1 case was PLEVA among 9 patients whose lesions persisted or were recurrent (Table 4).

DISCUSSION

In 1894, Neisser and Jadassohn independently described pityriasis lichenoides. Both authors included in their report cases of what would now be considered both acute and chronic variants of the disease. Juliusberg redescribed chronic pityriasis lichenoides in 1899 and designated it a separate entity, pityriasis lichenoides chronica[3].

In 1902, Brocq proposed the term parapsoriasis for the first time and intended it to denote a group of separate diseases described previously. Certain features in common of that group were unknown etiology, chronicity, failure to respond to therapy, and lack of symptoms, particularly of pruritus. He included PL under parapsoriasis. In 1916, Mucha redescribed acute pityriasis lichenoides as an entity separate from both chronic pityriasis lichenoides and from other disease, and Habermann, in 1925, gave this entity the name pityriasis lichenoides et varioliformis acuta[3].

Ever since this division of PL into two entities remains a controversial issue, but PL was distinguished, on clinical grounds, as a separate entity from the other parapsoriasis and removed from the group by other authors afterwards. So most authors today consider PL separately from the other parapsoriasis, and now in general it seems more appropriate to consider PL as a single entity with acute and chronic variants[3].

PL is composed of generalized erythematous or brown, often hemorrhagic, scaly papules and small macules which either persist for many months or recur in periodic exacerbations. Lesions are predominantly present over the trunk and flexural surfaces of the extremities. The chronic form may be distinguished from the acute form by the lack of acute, necrotic varioliform lesions. Except for the clinical appearance, however, there is no characteristic diagnostic criteria for either the acute or chronic form[1, 3].

PL is usually described as a disease of young adults and there are few reports of childhood PL especially in comparison with adult form. Marks et al[4] reviewed 128 cases of PL and said that 20% of the patients were less than 10 years old. Longley et al[5] reported that PLEVA was similar both clinically and histopathologically in children and adults, although lesions of the scalp and mucous membranes appeared to be more rare in children and said that PLEVA was underdignosed in children. In our study the prevalence rates of both childhood PL and adult form were quite similar, so our finding supported that there may be as many childhood cases as adult cases.

PL is described as a disease that is usually

asymptomatic or even to the extent of completely asymptomatic[1]. Marks et al[4] said that the disorder was only troublesome cosmetically, although some patients admitted that it was mildly itchy. But in our pediatric group 9 patients (37.5%) complained of pruritus and 2(8.3%) fever while in the adult group 12 patients (30.8%) complained of pruritus. Therefore, there seemed to be more subjective symptoms in childhood form.

Trunk and proximal extremity are generally described as the predilection sites for adult form of PL[1, 3]. But Ingram[6] documented lesions on the scalp or mucous membranes in up to 9% of patients, in whom both childhood and adult PL were included. In our pediatric group, in addition to the usual distributions of the skin lesions facial involvements were noted in 6 patients (25%), palm-sole involvements in 4 patients (16.7%) while in the adult group facial involvements were noted in 3 patients (7.7%), palm-sole involvements in 3 patients (7.7%). There seemed to be more atypical locations in childhood form than in adult form.

Longley et al[5] said that lesions of the scalp and mucous membranes appeared to be more rare in children, but both groups in our study showed only 1 case of scalp involvement and no mucosal involvement.

Truhan et al[7] reported that erythromycin was effective in that 11(73%) of 15 childhood PL treated with erythromycin had a remission usually within 2 months. Rasmussen[8], however, noted no consistently beneficial response in 8 children with PL treated with erythromycin for a minimum of 6 weeks. Fourteen of our childhood patients were treated with erythromycin alone, and 10 patients among these were cleared. In our cases erythromycin alone was a quite effective method of treatment. The case treated with prednisolone in this study was "febrile ulceronecrotic PLEVA" which is a rare form of PLEVA.

Shavin et al[9] described that the terms "acute" and "chronic" refer to the morphological appearance of individual lesions rather than to the course of the disease, and said that the actual clinical course of either PLC or PLEVA is quite long due to their recurrent nature.

A few cases of PLEVA have been reported to evolve into PLC[1, 10], and one case of persistent childhood PLEVA in our study might be actually PLC that progressed clinically from PLEVA, but histopathologically we could not confirm.

Marks et al[4] described the duration of rash in cured patients which ranged from 3 months to over 5 years. In our pediatric group the average duration of disease for PLEVA after treatment was 5.9 months and that for PLC was 12.3 months, respectively. PLC and PLEVA did not seem to respond to treatment, especially, with antibiotic. There may be some possibility of recurrences among cleared patients if these patients are followed up for longer period, but until now these patients were followed up for more than a year after clearing.

In our adult group 1 patient refused treatment and 8 patients were not followed up. Therefore, the comparison of clinical courses between 2 groups was quite difficult but in 30 adult patients clinical courses were evaluated. 11 cases were cleared of the lesion after treatment in which 7 cases were PLEVA and 4 were PLC. The average duration of disease in the 7 PLEVA cases was 5.9 months (range, 1-16 months) and that in the 4 PLC cases 13.8 months (range, 13-15 months), respectively. In 19 patients whose lesions persisted or were recurrent, the average duration of disease in the 9 PLEVA cases was 26 months (range, 14-38 months) and that in the 10 PLC cases 44 months (range, 25-60 months), respectively. It is our impression that the clinical course may be a little shorter in childhood PL than in adult form.

In our study various laboratory works had not been done routinely in all patients, and we could not try to elucidate probable etiological factors or evaluate the associated systemic diseases but the general health did not appear to be affected by the disease itself except for the rare case of "febrile ulceronecrotic PLEVA".

We think that more attention must be paid to detect childhood PL and be able to specify various clinical characteristics of childhood PL in comparison with adult form.

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