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Case Report

Chronic papulosquamous skin lesions in a 9-year-old boy

一名9歲男童患有慢性丘疹與鱗狀病損

WK Tang 鄧偉祺

Pityriasis lichenoides chronica belongs to a group of disease collectively called pityriasis lichenoides. This disease generally runs a benign but protracted course. In this report a 9-year-old boy who has this condition for one year was described. Pityriasis lichenoides with emphasis on its chronic type will be briefly reviewed.

慢性苔蘚樣糠疹是屬於一組統稱苔蘚樣糠疹的疾病。其臨床過程良性但反覆不癒。本病例報告報導一9歲男童患此症一年。苔蘚樣糠疹尤其慢性型將予簡單回顧。

Keywords: Cutaneous lymphoma, papulosquamous lesion, pityriasis lichenoides, pityriasis lichenoides chronica

關鍵詞：皮膚淋巴瘤、丘疹性鱗屑疹、苔蘚樣糠疹、慢性苔蘚樣糠疹

Introduction

Pityriasis lichenoides chronica (PLC), pityriasis lichenoides et varioliformis acuta (PLEVA), febrile ulceronecrotic Mucha-Habermann disease are collectively under the group of disease called pityriasis lichenoides (PL). It is generally believed that they belong to the same disease spectrum and mainly different in the clinical course and severity of their cutaneous manifestations. Although majority of the cases run a benign clinical course, malignant transformation had rarely reported. It is also important to bare this condition in mind in the differential diagnosis of other papulosquamous conditions.

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Case report

TM was a 9 years old boy. He enjoyed good past health until 8 years old when he developed generalised itchy brownish rash on the body. Apart from slightly worse in the winter and hot weather, the lesions were rather stable in size and extent throughout the past year. He has no systemic upset all along.

He had no family members shared similar skin problem.

Physical examination showed multiple well-defined discrete brownish-red papulosquamous patches and plaques, without necrosis randomly distributed on the trunk and limbs (Figures 1 & 2). The face, palms and soles were relatively spared. Mild ichthyosis was noted. There was no excoriation mark. No abnormality was found on the buccal mucosa, the teeth and nails. There was no lymphadenopathy or organomegaley detected.

A skin biopsy was performed on the right leg, which showed mild hyperkeratosis, focal parakeratosis, and mild acanthosis. Lymphocytic exocytosis with mild spongiosis and a few scattered necrotic keratinocytes are seen in the epidermis. Vague vacuolar changes in the basal epidermis are present. Moderately dense perivascular lymphohistiocytic infiltrate with focal extravasated red cells are found in the upper reticular dermis. The whole histological picture is compatible with pityriasis lichenoides chronica.

The patient was treated symptomatically with emollients and topical steroid [Flucinolone 0.0125% (Synalar) & Mometasone furoate (Elomet)] with fair response.

Discussion

Pityriasis lichenoides chronica (PLC), pityriasis lichenoides et varioliformis acuta (PLEVA), febrile ulceronecrotic Mucha-Habermann disease are collectively under the group of disease called pityriasis lichenoides (PL).

Pityriasis lichenoides was first described by Neisser and Jadassohn in separate reports in 1894.[1,2] From previous large case series studies,[3-6] it was found that in general population (age >18 years old), the average age of onset was 29-year-old with prevalence, and peaked in the third decade, and there was no strong predisposition in either

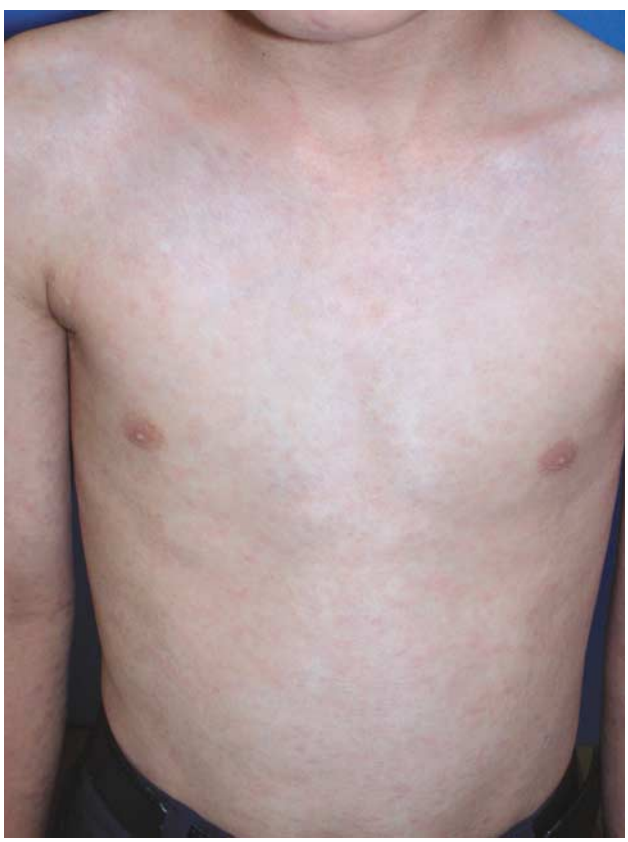

Figure 1. Papulosquamous lesions on the trunk.

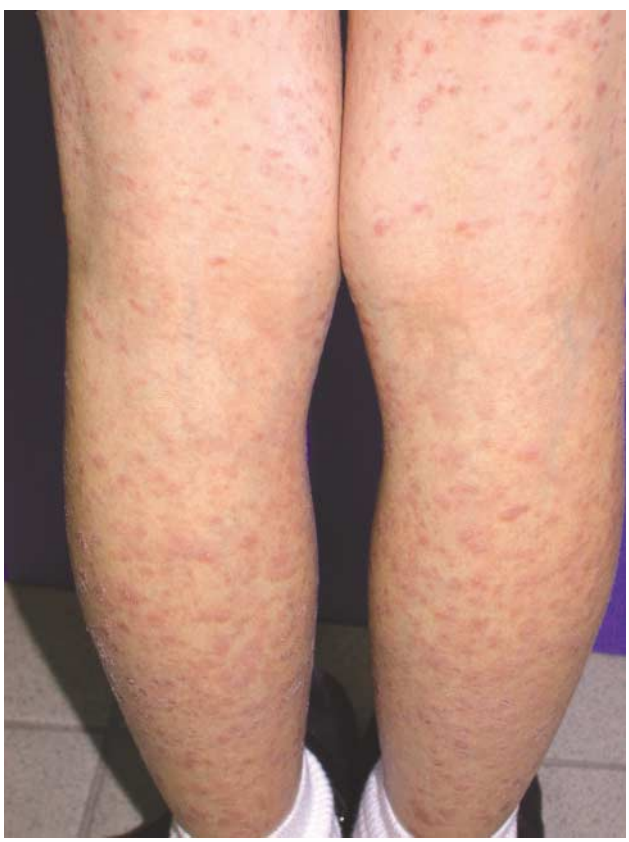

Figure 2. Papulosquamous lesions on the limbs.

sex. While in paediatric group, the disease was peaked at 5 and 10 years of age with male predominance. In the febrile ulceronecrotic subtype, the mean age of onset was 30.6 years, and was peaked in the second decade of life, and was also male predominant.

It is generally accepted that PLEVA and PLC represented two ends of a continuous spectrum. Features of both acute and chronic lesions may occur in the same person. The descriptive terms *acute* and *chronic* are referring to the characteristics of the individual lesions and not the course of the disease.

The disease is characterised by recurrent crops of erythematous papules with a centrally adherent mica-like scale that could be easily detached to reveal a shiny, pinkish-brown surface. They occur on the trunk and *proximal portions* of the extremities. Unlike PLEVA, the papules flatten spontaneously and regress over a period of weeks. It often leaves behind a hyper-hypopigmented macule.

Each lesion could last for several weeks, and there are often exacerbations and remissions. The entire course could take several years. In general, those with diffuse distribution of lesions have the shortest average course (11 months), whereas those with peripheral distribution have the longest average course (33 months); and the central distribution one is intermediate.

Dark-skinned individuals could primarily present as generalised hypopigmented macules that lack scale. So a high index of suspicions is needed in interpretation of this disease.

Cutaneous lesion with papulosquamous morphology is commonly found in patients with chronic eczema. However, the lesions in chronic eczema are usually less well-defined and itch is a very common associated symptom. Pityriasis rosea could also present with papulosquamous lesions. However about half of the patients have characteristic herald patches, and female is more commonly affected. Moreover, our patient's skin lesions persisted much long than the usual course in pityriasis rosea, which usually resolved in two months. Our patient did not volunteer any history of drug exposure before the onset of the skin rash. And the persistence of the skin rash over a year refuted the possibility of drug eruption. On the other hand, mucosal or nail involvement, and Wickham's striae on the lesions were expected in lichen planus, but they were absence in our index patient. Other differential diagnoses which could not be easily differentiated clinically included guttate psoriasis, pityriasis lichenoides (chronic type), lymphomatoid papulosis (LyP) and secondary syphilis. Gianotti-Crosti disease (papular acrodermatitis of childhood) is an uncommon disease with papulosquamous skin lesions, but in which the lesions tend to be more edematous and were acrally distributed.

Histopathologically, there is a gradual change between PLEVA and PLC. In general, more extensive epidermal involvement with keratinocytes necrosis and dense lymphohistocytic infiltration is found in PLEVA (Table 1).

Both PLEVA and PLC lesions have shown that T cells predominate in both dermal and epidermal inflammatory infiltrates, with CD8+ and CD4+ cells predominate in PLEVA and PLC respectively.

Aetiology of PLC is still unknown. As clustering of cases and familial outbreaks had been observed, so infectious causes such as Toxoplasma gondii,[7] Epstein-Barr virus,[8] cytomegalovirus, HIV,[9] and varicella zoster virus had been suggested.[10]

Immune-complex mediated vasculitis was first suggested by Black and Marks. But they failed to find any evidence of immunoglobulin or complement deposition in their biopsies.[3] Until

Table 1. Histopathological change between PLEVA and PLC

Sites of involvement	PLEVA	PLC
Epidermis	-	-
1. Parakeratosis, dyskeratosis, spongiosis, acanthosis	Focal & confluent	Focal
2. Vacuolisation of basal cells	With necrotic keratinocytes	Minimal
3. Focal epidermal necrosis	Prominent	Few
Dermis	-	-
Perivascular lymphohistiocytic infiltrate	Dense, wedge-shaped and extend deep into reticular dermis	Mild superficial, only focally obscures the DEJ
Vascular change	Extravasation of erythrocytes, invasion of vessel walls	Few/absent

1972, circulating immune complexes, and IgM were found in DEJ or vessel wall in the skin biopsies by Clayton.[11] However, these results were not reproducible in other studies.

Wood et al in 1987 postulated that PLEVA and PLC were a primary T-cell lymphoproliferative disorders.[12] And a subset of infiltrating cells might be the primary target, with epidermal destruction representing a secondary event. The clinical course of the disease might just reflect the host immune response to abnormal cells (neoplastic T cells). In this hypothesis, a small clonal subset of CD8+ cells infiltrate the skin (below the detection threshold of PCR) to give rise to PLC. With more influx of clonal CD8+ cells, the clinical manifestations of PLEVA become evident. At this point, a T-cell clone can be detected. In PLEVA, the host immune response is able to prevent the development of a cutaneous lymphoma; otherwise, the clone may acquire enough genetic alterations to overwhelm the body's immune system. LyP (lymphomatoid papulosis, a relatively more malignant condition) might be the result of a less effective host immune response.

Malignant transformation of PLC, though occurred rarely, had been reported. In English literature, the first of such case was published in 1986 by Bleehan and Slater.[13] Fortson et al reported the first cutaneous T-cell lymphoma (CTCL) from a child with PLVEA in 1990.[14] Sporadic cases had been reported thereafter with patients' age ranged from eight-months-old to 62-year-old.[15,16] Panizzon et al's cases series was the most interesting.[17] Although all of his patients had PLC and lymphoma, one of them developed Hodgkin disease three years prior to the onset of PLC. And in another patient, lymphoma occurred in the lung. The author argued that PLC might be considered as a paraneoplastic syndrome rather than genuine transformation.

All members of the parapsoriasis group have been shown dominant T-cell receptor clonality.[18] And the presence of T-cell clonality might favour the possibility of malignant transformation. Recently, a small cases study consisting of six PLC patients, T-cell clonality (by using primers Vγ1-8/Jγ1-2 and primers Vγ9/Jγ1-2 for T-cell receptor gene) had been detected in three patients.[19] One of them developed CTCL and another had LyP later. However, the significance of this study was doubt as the number of the patients involved was small, and the T-cell receptor gene rearrangement in PLC lesions was not consistent

with the CTCL lesions. And in the patient with LyP, even no such rearrangement was found in the LyP lesions.

Treatment of PLC is largely anecdotal or based on uncontrolled case series. Topical corticosteroids are the first line of treatment. However, no studies have specifically compared the efficacy of it with that of either placebo or other treatments.

Systemic agents include antibiotics [Tetracycline (2 g/day) and erythromycin (600-800 mg/day)], methotrexate (7.5-20 mg/wk), cyclosporine, dapsone, and pentoxifylline had been tried in the order of frequency.

PUVA had been used with success in both PLEVA and PLC. The total amount of energy used varied from 10-370.5 J/cm^2. However, recurrences after cessation of therapy or after long periods of remission may occur.

Recently, promising results had been demonstrated in UVB. Complete remission (BUVB, nUVB) could be reached up to 93.1%.[20] Disease free period was upto 58 and 38 months with BUVB and nUVB respectively. However, in this study, most patients being treated were of type I and II skin. Longer treatment duration and/or higher energy may be needed in darker skin type individuals.

In conclusion, current evidences suggested that PLEVA and PLC may represent different stages of evolution of a single entity. Clonality does not seem to correlate with clinical outcome, and only rare cases have been associated with malignant lymphoma. There is no standard treatment for PLEVA and PLC. As PLC is a self-remitting disease, symptomatic treatment is all that is needed in most cases.

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