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PEDIATRIC PITYRIASIS LICHENOIDES CHRONICA: A CASE REPORT

Dermatology

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ABSTRACT

Pityriasis Lichenoides encompasses a continuum from Pityriasis Lichenoides et Varioliformis Acuta (PLEVA) to Pityriasis Lichenoides Chronica (PLC). A wide variety of clinical conditions need to be differentiated from the chronic form of the disease. Though the exact etiology is unknown, there is no specific treatment and the duration of the disease unpredictable and it is thought to be self-limiting. A pediatric case of PLC with remitting and relapsing course is presented for its rarity.

KEYWORDS

INTRODUCTION

Pityriasis Lichenoides (PL) is an uncommon, acquired spectrum of skin conditions that could progress to cutaneous lymphoma or an ulceronecrotic form. The spectrum of PL encompasses a continuum that includes an acute form known as Pityriasis Lichenoides et Varioliformis Acuta (PLEVA), and a chronic form known as Pityriasis Lichenoides Chronica (PLC). A febrile ulcero-necrotic form of PLEVA is also known[1].

Reports of immunoglobulin and complement deposition in vessels prompted this condition to be labelled as a vasculitis[2]. But absence of fibrinoid deposits in the blood vessel wall doesn't lend credence to a vasculitic theory[3]. Other pathophysiologic theories suggest a (1) cell-mediated mechanism, based on a cytotoxic/suppressor phenotype T-lymphocytic infiltrate, diminished epidermal Langerhans cells and reduced Cd4/ CD8 ratio; (2) self-limited self-healing lymphoproliferative disease[4], based on findings of CD30 (Ki-1) cells; (3) T-cell dyscrasia[5], based on the presence of intraepithelial atypical lymphocytes, phenotypic abnormalities, and TCR-gamma rearrangements. Rarely, PLC could transform into cutaneous lymphoma[6]. Though the exact etiology is unknown, antibodies against Epstein Barr virus, toxoplasma and HIV have been demonstrated in some studies[7,8,9].

There is no standard and specific treatment of PL. The disease is thought to be self-limiting though the duration of the disease cannot be predicted.

We report a pediatric case of PLC with remitting and relapsing course.

CASE REPORT

A nine year old boy developed gradually progressive red rash over the trunk and limbs over a period of two years. The eruptions were painless, dry, flaky and slightly itchy in nature. There were no constitutional or systemic features. There was no history of drug intake or food allergies prior to onset of his symptoms. Dermatological examination revealed multiple, small, discrete, erythematous popular, polymorphic eruptions, some topped with a thin 'frosted glass' or 'micaceous' scale, scattered over the front and back of the trunk as well as over the extremities with no specific predilection for the extensors (Fig 1). The mucosae, palms, soles and scalp were normal. Hematological and biochemical parameters were normal. Histopathological examination with H&E stain revealed mildly acanthotic epidermis and thick confluent mounds of parakeratotic cells interposed between basket weave to orthokeratotic layers. Mild perivascular and sparse lymphocytic infiltrate was present in papillary to upper reticular dermis (Fig 2). Interface pathology in the form of lymphocytes filling the dermal papillae and abutting the basal layer which, in turn, showed vacuolization and occasional apoptotic keratinocyte. Foci of mild lymphocytic exocytosis into epidermis as well as sparse extravasated erythrocytes in papillary dermis were also seen (Fig 3). A diagnosis of PLC was made. He was started on a one week course of oral erythromycin and tapering dose of low dose corticosteroids for 2 weeks. He responded well with regression of lesions but the lesions gradually recurred on stopping treatment. Subsequently, he was started on NB-UVB @ 200mJ thrice weekly for three months to which he responded well. The lesions resolved well with residual post-inflammatory hypopigmented macules (Fig 4).

DISCUSSION

PL is seen in the first three decades of life, while in children it occurs within 3-15 years of age. The chronic form (PLC) is more common in children[10]. PLEVA and PLC as clinical forms are believed to be the edges of a continuum. PLEVA is an eruption of multiple, small, erythematous papules, vesicles, pustules, hemorrhagic crust-covered papules, and shallow ulcers that heal in weeks to months with pox-like scars. It has a predilection for the anterior trunk, flexural surfaces and proximal extremities. PLC is characterised by gradually developing, small, red-brown maculopapules with fine centrally attached shiny scales. The lesions are scattered on the trunk and proximal extremities and resolve with hypo/hyperpigmentation in the absence of scarring. This rash may resolve in months or wax and wane for years.

A case of PLC should be differentiated from guttate psoriasis, lichen planus, lymphomatoid papulosis, pitysiasis rosea, viral exanthema and drug eruption. Extensor predominant plaques topped with loosely adherent silvery white scales and positive Auspitz sign characterize psoriasis. Inverse Christmas tree patterned erythematous rash with peripheral rim of centrally detached scales characterizes pityriasis rosea. Lymphomatoid papulosis is a T-cell lymphoproliferative disorder of the skin, characterized by polymorphic pruritic papules with histology of malignant lymphoma[11].

Management of PL includes antibiotics (erythromycin, tetracyclins), immune suppressants (methotrexate, cyclosporine), anti-inflammatory drugs (corticosteroids, acitretin), phototherapy (PUVA, NB-UVB), tacrolimus.[12,13,14]

PL in children is more likely to run an unremitting course, with greater lesional distribution, more dyspigmentation and a poorer response to conventional treatment modalities[15].

CONCLUSION

PLC is a chronic form of a rare disease PL. It should be considered as a differential diagnosis in maculopapular eruptions in pediatric age group owing to its chronic course, unpredictable response to treatment as well as the rare progression to lymphoma.

Illustrations

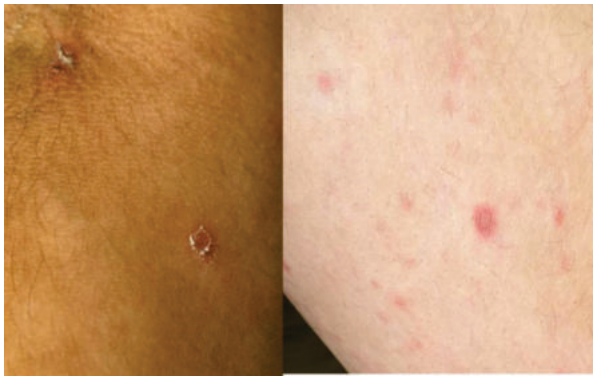

Figure 1. Erythematous papulosquamous lesions of PLC on lower

limb; the left panel revealing typical scale

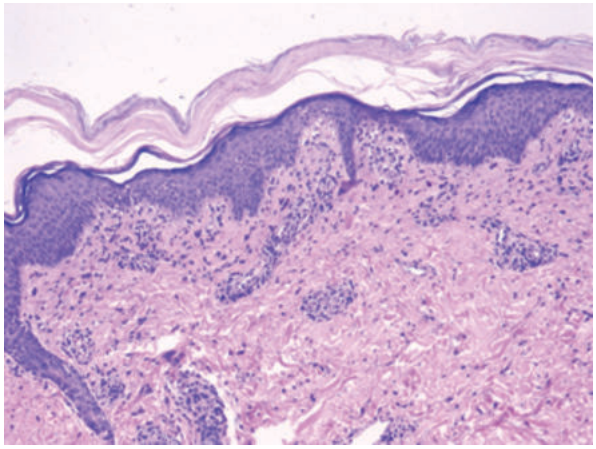

Figure 2: Histopathology (H&E X10)

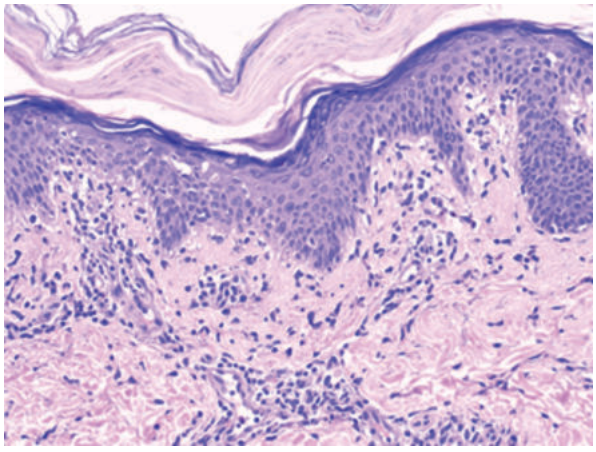

Figure 3: Histopathology (H&E X20)

Figure 4: Multiple ill-defined post-inflammatory hypopigmented macules on both forearms and back of trunk

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