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PROGRESSIVE SYMMETRIC ERYTHROKERATODERMA ASSOCIATED WITH SYMMETRIC SYNDACTYLISM

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A case of progressive symmetric erythrokeratoderma (PSEK) associated with symmetric syndactylism is being reported. The interesting feature being the first time description of any association with PSEK.

Key Words : Progressive symmetric erythrokeratoderma, Syndactylism

Introduction

Progressive symmetric erythrokeratoderma (PSEK) is a rare inherited cornification disorder first described by Darier in 1911[1] and is characterized by symmetric erythematous hyperkeratotic plaques. Fewer than 30 cases have been reported[2,3] and to our knowledge, no association of this condition with other disorders has been reported so far. This is the first description of PSEK associated with syndactylism.

Case Report

A 5-year-old boy was evaluated recently in our unit because of asymptomatic cutaneous lesions of 4 months duration. On examination he showed erythematous, palmoplantar keratoderma that was sharply marginated over the wrists and ankles, with erythematous hyperkeratotic plaques distributed on the back of the hands and feet, elbows, knees, tendo Achillis, malleoli and buttocks. The lesions were well demarcated and exhibited remarkable symmetry (Figs. 1 and 2). In addition syndactyly of both feet involving the two outer toes (Fig. 1) and keratosis pilaris lesions over legs and lower back (Fig. 2) were also noted. The abdomen, thorax and face were unaffected. Rest of the physical examination was unremarkable.

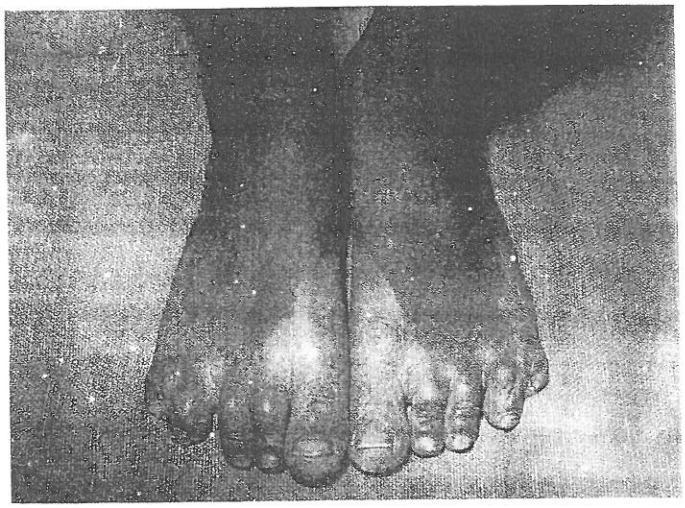

Fig. 1. Characteristic PSEK lesions with symmetric syndactylism.

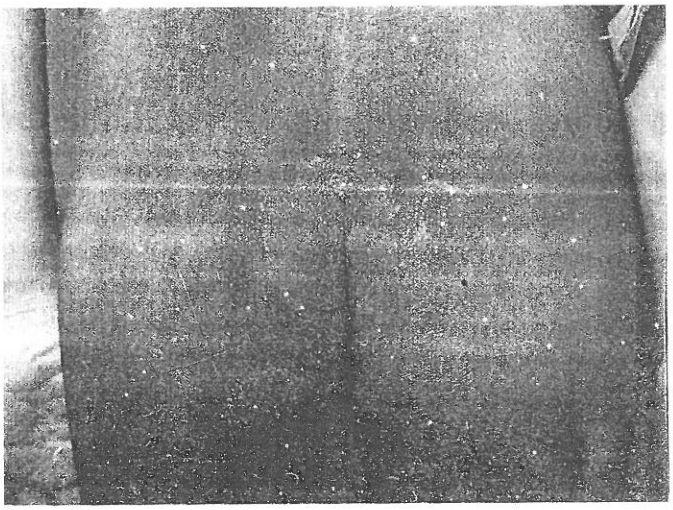

Fig. 2. Lesions of PSEK and keratosis pilaris on buttocks.

There was no family history of similar skin eruptions and/or congenital defects, and the patient's parents were not related.

All of the laboratory examination were

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within normal limits. X-rays of the feet showed no synostosis. Histologic examination of skin biopsy specimen from erythematous hyperkeratotic plaque revealed changes compatible with a diagnosis of PSEK.

Comments

This case is doubly interesting not only in its rarity but also in its first time association with other disorders. Furthermore the associated syndactylism also shows a striking degree of symmetry akin to PSEK lesions. Finally, the triple association of PSEK, symmetric syndactylism and keratosis pilaris in the present case is just coincidental or actually features of a new syndrome?

References

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