Case report

POROKERATOSIS HYPERTROPHICA ET DISSEMINATA

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ABSTRACT

We report on a 54-year-old man with an unusual variant of porokeratosis that has been confirmed histologically. Nearly the whole skin surface was involved. On the heels and on the low extremities there were lesions of porokeratosis Mibelli of hypertrophic type with bizarre polycyclic configuration. On the upper extremities and on the trunk the skin lesions resembled an exaggerated variant of disseminated actinic porokeratosis. The lesions on the face were typical of the common type of the latter. Patient's father and elder brother have the same lesions but less pronounced. A systemic etretinate therapy during a period or 5 months was relatively successful. Because of the unusual clinical pattern the possibility of a new variant of porokeratosis is discussed.

KEY WORDS

porokeratosis Mibelli, hypertrophic and disseminated variant, systemic, etretinate therapy

INTRODUCTION

Porokeratosis was first described by Mibelli in 1893 (1). It is an inherited disorder of the skin, characterized by solitary or multiple lesions with hypertrophic border and an atrophic centre, most commonly located on the extremities.

The cause of the disease is unknown. Reed and Leone in 1970 suggested that abnormal clones of keratinocytes are responsible for the development of porokeratotic lesions (2).

Porokeratosis is probably an autosomal dominant condition with variable penetrance, where the predisposition to it is inherited but the predisposition to the type of porokeratosic lesions may be not. What determines the type of morphologic expression of the disease is not yet clarified (2).

A wide variety of clinical manifestations include small ring-like lesions, hypertrophic verrucous lesions, then lesions of the superficial disseminated type, lesions in zosteriform distribution, lesions occurring over the buccal mucosa and finally linear lesions that resemble linear verrucous epidermal nevus (3,4,5,6,). Development of squamous cell carcinoma or of Bowen's disease within features of porokeratosis had been reported in patients with solitary, disse-