

MYCOLOGICAL EXAMINATION IN PACHYONYCHIA CONGENITA

A. Kansky, M. Dolenc-Voljč, P.E. Bowden and M. Belič

ABSTRACT

Introduction. Pachyonychia congenita (PC) is a hereditary disorder of keratinization characterized by extremely thickened nails, follicular hyperkeratosis, insular palmoplantar keratoderma and thickened, whitish lingual and buccal mucosa sometimes involving also gingivae. It was established during the last few years that this disorder is due to mutations of keratin genes. The additional role of yeasts in the pathogenesis of PC was sometimes questioned, however no systemic study could be traced in the literature.

Methods. Mycologic investigations of nails and insular keratoses were carried out in eight Slovenian patients with PC-1: direct microscopy and cultivation on Sabouraud's dextrose agar. In all these patients the keratin mutations had been defined.

Results. *Candida species (Cand sp)* were isolated from fingernails in two and *Aspergillus sp* in another patient. From toenails *Cand sp* was isolated in 2 and *Rodotorula sp* in another patient. From the keratinous material from the soles *Trichophyton mentagrophytes* was isolated in two patients and *Cand sp* in another.

Conclusion. Detection of *Cand sp*, *Aspergillus sp* or *Rodotorula sp*, spores in just a few patients is considered to be an accompanying phenomenon.

KEY WORDS

mycological examination, nails, pachyonychia congenita, PC-1, yeasts

INTRODUCTION

Pachyonychia congenita (PC) is a well-defined entity within the broad group of hereditary palmoplantar keratodermas (HPPK). According to McKusick (1) two broad phenotypic variants of PC are recog-

nized: type Jadassohn-Lewandowsky or PC-1 (MIM 167200) and type Jackson-Lawler or PC-2 (MIM 167210). PC-1 is characterized by a thickened, brownish nail plate with a rough surface, insular hyperkeratoses of palms and soles, follicular hyperkeratosis affecting

