

Palmoplantar Keratoderma: Rare Case Report

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Abstract

Palmoplantar keratodermas (PPK) are group of cornification disorders characterized by epidermal hyperkeratotic lesions involving the palms and soles. A 50years old healthy male, presented with history of multiple punctate hyperkeratotic papules since last 5 years.

Keywords: Palmoplantar keratoderma • Punctate • Hyperkeratotic papules

Abbreviations: PPK: Palmoplantar keratodermas • PUVA: Psoralen plus Ultraviolet A • PPPK: Punctate Palmoplantar keratodermas • USG: Ultrasound Sonography • VRDL: Venereal Disease Research Laboratory Test • ELISA: Enzyme-Linked Immunosorbent Assay

Introduction

Palmoplantar keratoderma (PPK), clinically and genetically comprises of heterogenous group of disorders characterised by hyperkeratosis of palms and soles [1]. It can be hereditary or acquired. Hereditary PPK can be further divided into three major categories: diffuse, focal, and punctate PPK (PPPK) [2]. These diseases can be distinguished from each other on the basis of pattern of inheritance, onset, distribution, morphology, severity, histopathological findings, additional dermatological findings and systemic manifestations [3]. The prevalence of PPPK was estimated to be 1.17/100,000 [4]. The exact etiology is little understood. It is believed that both genetic and environmental factors play some roles. PPPK has an autosomal dominant pattern of inheritance and linked with two loci on chromosomes 15q22 15q24 and 8q24.13–8q24.21 [5].

PPPK have three distinct types:

Type 1 -Punctate Palmoplantar Keratoderma

Type 2 - Porokeratotic type

Type 3 - Focal Acrohyperkeratosis [6]

Male patients are affected more [7].

Case Report

A 50-year-old male presented with multiple, diffuse hyperkeratotic papules on palms and soles since 5 years. They were gradually increasing in number spontaneously and pain increased while walking. The patient was a known case of diabetes mellitus. No family history of any such lesion or any systemic complaints. Routine investigations, blood biochemistry, USG abdomen and x-ray chest were inconclusive. VDRL and ELISA for HIV were negative. On cutaneous examination multiple, punctate hyperkeratotic papules of a near uniform size of 0.3 to 0.5 cm diameter were seen on plantar aspect of both feet including the lateral borders. Palms had few hyperkeratotic papules.

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Mucosal surfaces were not involved. Biopsy sample was received. On histopathological examination of biopsy revealed massive hyperkeratosis over sharply limited area with depression of malpighian layer below general level of epidermis. There was increase in the thickness of granular layer. The dermis was free of inflammation. Compilation of clinical and laboratory data helped to conclude the diagnosis of Palmoplantar Keratoderma-Punctate type.



Figure 1: Shows hyperkeratotic papules on Feet.



Figure 2: Shows hyperkeratotic papules on Hands.

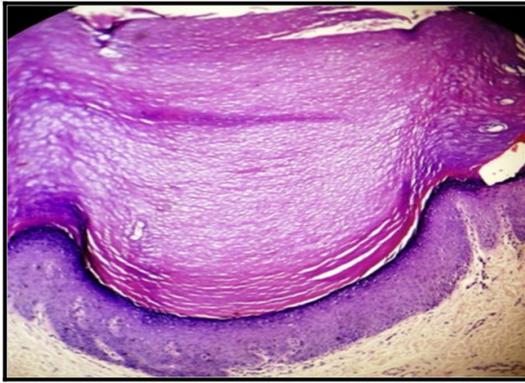


Figure 3: Hyperkeratosis causing depression of malpighian layer.

Discussion

Its pathophysiology is still unclear, however Giehl et al. reported in 2012, for the first time the exact pathophysiological mechanism of the disease: two heterozygous nonsense mutation of the AAGAB gene located on chromosome 15 were found in all affected individuals in the study [8]. Acquired PPKs may arise due to changes in a person's health or environment. This usually occurs later in life and may be due to many causes, such as drugs, malnutrition, chemicals, systemic diseases, cancer, and infection. Differentiating features of type 1, 2 and 3. Type 1-Hyperkeratotic papules-Columns of orthohyperkeratosis, absence of epidermal dyskeratosis or hydropic change. Type 2-Acuminata (spiny) keratosis-Cornoid lamella. Type 3-Oval or polygonal papules a) Focal acral hyperkeratosis-Absence of elastorrhexis b) Acrokeratoelastoidosis- Presence of elastorrhexis [9]. Some cases have been associated with nail abnormalities such as longitudinal ridging, onychoschizia, onychorrhexis, trachyonychia, and notching. Although systemic involvement is uncommon, patients with PPPK may have an increased risk of developing malignant conditions of gastrointestinal, lung, and breast as well as Hodgkin's lymphoma.

PPPK1 is usually an isolated finding. Rarely, it may be associated with psoriasis, lentigo simplex, and guttate hypo-pigmentation. Guttate hypo-pigmentation and PPPK1 are parts of the constellation of clinical features of Darier disease and epidermolysis bullosa simplex with mottled pigmentation [10].

Differential diagnosis of PPPK1 include verruca vulgaris, callosities, porokeratosis, aquagenic palmar keratoderma, hereditary papulotranslucent acrokeratoderma, punctate keratosis of palmar creases, and arsenical keratosis [11]. PPPK lesions do not resolve spontaneously, and treatment is generally symptomatic. Topical keratolytics, systemic retinoids, liquid nitrogen, PUVA (psoralen plus ultraviolet A), re-PUVA, and pairing have been used with variable success. The case is being presented in view of its rarity and no family history except for the patient having diabetes mellitus.

Conclusion

Although it is a clinical rarity, a spot diagnosis can usually be made, provided one is familiar with the condition thereby unnecessary diagnostic procedures can be avoided.

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