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Dermatology

A CASE REPORT OF FAMILIAL PUNCTATE PALMOPLANTAR KERATODERMA.

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ABSTRACT Palmo-plantar keratodermas (PPKD) are a group of hereditary and acquired disorders, characterized by excessive epidermal thickening of the skin of palms and soles. These can be isolated and non-syndromic or can be a part of a complex of symptoms i.e., syndromic.

Here, we report a case of punctate palmoplantar keratoderma in four generations of a family.

KEYWORDS : Palmoplantar keratoderma [PPKD] , Autosomal dominant, keratinization, keratin.

INTRODUCTION

Palmoplantar keratodermas [PPKD] are disorders of keratinization characterized by the deposition of excessive keratin in the horny layers of palms and soles. They can be either hereditary or acquired. Clinically, PPKDs are of four types: Diffuse, focal, punctuate, and striate. Punctate PPKD is an autosomal dominant keratoderma, clinically characterized by small rounded papular lesions on the palms and soles that tend to coalesce over pressure points. Also called Keratosis punctata palmoplantaris type Buschke–Fischer–Brauer and Palmoplantar keratoderma punctata type 1 (PPKP1].

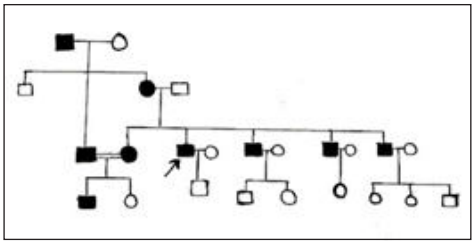
The disorder has been mapped to two chromosomal regions 15q22 and 8q24.13–8q24.21. It occurs in early adolescence but also up to the sixth decade of life. Pinpoint keratotic papules, initially translucent and with a depression in the center but later opaque and warty, on the palms and soles. Severity is variable among family members. Lesions are more florid in manual workers. There is no involvement of the dorsa of the hands or legs, nor the knees or elbows.

Histology shows Orthohyperkeratosis with compact acanthosis and hypergranulosis with a depression in the center of the lesion.

CASE REPORT

A 65-year-old male patient presented with as ymptomatic lesions on palms and soles for approximately 40 years. The lesions started at 13-15 years of age in the patient. Similar lesions of variable severity are noted in other family members [all 4 siblings, mother and maternal uncle, maternal grandfather, and nephew]. The patient and All siblings are involved in farming.

On histopathological examination (3 mm punch biopsy specimen taken from sole of patient) , orthohyperkeratosis, hypergranulosis , acanthosis were noted. Depression was present in the centre.



Pedigree Chart Of Family

On cutaneous examination, both palms and soles showed multiple discrete skin-colored to brown hyperkeratotic papules of 1-5 mm sizes and some show central depression [pits]. soles have more extensive involvement with more lesions on weight- bearing areas. The Dorsum of hands and feet were normal. No lesions in other areas of the body. Nails of fingers and toes, hair, teeth, mucosae were normal. No systemic abnormalities were noted.

FIGURES

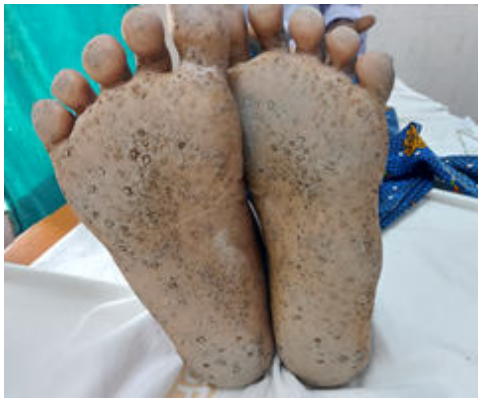


Fig-1: Soles Of Patient

FIG-2: Palms Of Patient

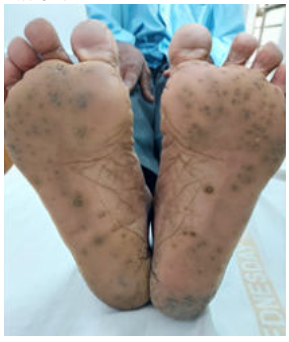


Fig 3: SOLES OF YOUNGER SIBLING

DISCUSSION

Punctate keratoderma also called keratoma dissipatum; keratoderma punctate; papulosa; disseminated clavus (Davies-Colley 1879) is one of the clinical variants of PPKD. Acquired forms of punctate PPKD may be associated with Lynch type II malignancies, chronic arsenicosis, angiosarcoma of the liver, bronchial adenocarcinoma. The hereditary types can be classified into 3 types:

Type 1 (Brauer-Buschke-Fischer keratoderma): Autosomal dominant condition with onset in the latter half of the second decade, possible association with malignant conditions.

Type 2 (porokeratosis punctata palmaris et plantaris): Numerous, tiny, pruritic keratotic spines resembling the spines of a music box. On histology, columnar parakeratosis is seen simulating coronoid lamella.

Type 3 (acrokeratoelastoidosis lichenoides): Polygonal or crateriform discrete papules on the lateral aspect of palms and soles.

Brauer-Buschke-Fischer PPKD may be associated with Lynch type II malignancies, liver, bronchus, squamous cell carcinoma of the chest wall, ethmoidal carcinoma, Hodgkin's lymphoma, malignancies of the colon, pancreas, kidney, breast, prostatic carcinoma, and atypical fibroxanthoma.

Differential diagnoses include focal keratoderma associated with malignancies such as breast and colonic adenocarcinoma, Autosomal dominant punctate porokeratosis (also referred to as PPKP2) , acrokeratoelastoidosis (referred to as PPKP3).

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