Case Report

Unilateral linear punctate palmoplantar keratoderma

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ABSTRACT

Punctate palmoplantar keratoderma (Brauer-Buschke-Fischer syndrome) is a rare entity. Among punctate keratoderma, the linear presentation is much rarer, and exact incidence is not known. Unilateral linear punctate palmoplantar keratoderma is not yet reported in the literature. Here we report a case 12-year-old child presented with asymptomatic linear punctate plaque on the left sole and hand; histology revealed hyperkeratotic epidermis without columns of parakeratosis or cornoid lamella.

Key words: Linear and unilateral, punctate, palmoplantar keratoderma

INTRODUCTION

Palmoplantar keratodermas are heterogeneous group of disorders characterized by abnormal keratinization of palmar and plantar skin.[1] Among which punctate palmoplantar keratoderma (PPPK) is much rarer with an estimated prevalence rate of 1.17/100,000. It is usually manifested bilaterally as asymptomatic, tiny, hyperkeratotic punctate papules/plaque on the palmoplantar surface.[2] The exact etiology of this disorder is not known, but a dual influence of genetic and environmental factors may trigger the disease.[2] Here, we report a case of unilateral linear PPPK for the first time.

CASE REPORT

A 12-year-old male presented with asymptomatic linear thickening and yellowish discoloration of left sole and left palm since 7 years and 5 years, respectively. Lesions first appeared on the left heel as punctate small plaques that gradually extend in linear fashion up to the third and fourth toe over a period of 7 years. Two years later, similar kind of lesions appeared on the left hypo-thenar eminence which slowly progressed and extend to the base of left little finger. There were no similar lesions on any other part of the body, and the lesions were asymptomatic throughout the course of the disease.

Family members had no similar complaints, no history suggestive of any systemic involvement, prolonged drug intake, chronic weight loss, drug or food allergy, recurrent rhinitis, itching, and dry skin.

Clinical examination revealed a yellow-colored thick linear punctate plaque on the left sole extending from the central part of the heel up to the third and fourth toe with variable width ranging from 1 to 5 cm. The surface of the plaque had multiple punctums; on palpation lesion was firm, rough, and non-tender [Figure 1]. Thickening and yellowish discoloration was more over pressure areas. Similar type of plaques with punctums ranging from 2 × 3 cm to 1 × 2 cm were present on the left palm extending from hypothenar eminence to the base of little finger [Figure 1]. Right palm and right sole were normal.

Apart from these cutaneous clinical findings, rest

Access this article online	
Quick Response Code:	Website: www.ijdvl.com
	DOI: 10.4103/0378-6323.90953

How to cite this article: Sharma S, Barman KD, Garg VK, Jain S. Unilateral linear punctate palmoplantar keratoderma. Indian J Dermatol Venereol Leprol 2012;78:85-8.

Received: June, 2011. **Accepted:** July, 2011. **Source of Support:** Nil. **Conflict of Interest:** None declared.

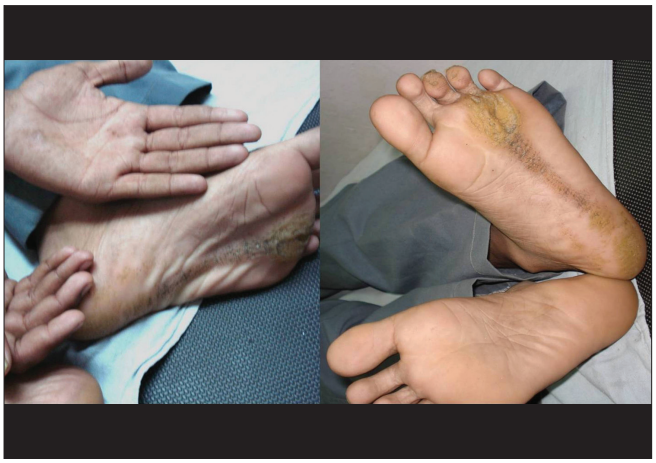

Figure 1: Localized plaques with punctums were present on the palmar surface of the left hand along with yellowish-colored thick linear punctate plaque on the left sole

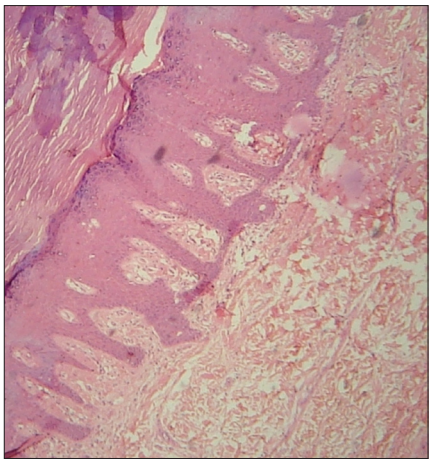

Figure 2: Histopathologic examination revealed hyperkeratotic epidermis without columns of parakeratosis or coronoid lamella (H and E, ×10)

of dermatological and systemic examinations were normal.

With these clinical findings, we had made the probable diagnosis of linear PPPK.

Patient routine investigation, stool for occult blood, chest X-ray, X-ray of left hand and feet, and ECG were within normal limit.

A 4-mm punch biopsy specimen from the left heel was sent to the department of pathology Maulana Azad Medical College, New Delhi, for histopathological examination, which revealed hyperkeratotic epidermis without any columns of parakeratosis or cornoid lamella [Figure 2]. With these clinical findings and histopathological features, the final diagnosis was made linear PPPK.

DISCUSSION

Palmoplantar keratodermas are heterogeneous group of disorders characterized by abnormal keratinization of palmar and plantar skin.[1] The palmoplantar keratodermas (PPKs) can initially be divided based on whether they are inherited or acquired. The inherited PPKs are further classified into three distinct types based on clinical pattern of epidermal involvement. The first is diffuse PPK, the second is focal PPK, and the third is punctate keratoderma.[3]

Incidence of various etiological factors causing hyperkeratosis of palms and soles in Indian patients is not known. Palmoplantar keratoderma is found to be more common in males (64.63%), with highest incidence in the age group of 11–20 years (32.92%). Occupation-wise manual workers such as laborers, farmers, and mechanical workers contributed to 48.16%, students 33.15%, and housewives 18.69%.[4]

Exacerbation of hyperkeratosis was seen in winter (41.46%), summer (20.73%), and autumn (19.51%).[4]

The exact etiology of this disorder is not known, but a dual influence of genetic and environmental factors may trigger the disease.[2] It has been described in few large autosomal dominant pedigrees. A genomewide scan was performed on an extended autosomal dominant pedigree, and linkage to chromosome 15q22-q24 was identified.[5] Martinez *et al*.[5] reported two new families with the same phenotype, which confirmed the mapping of the locus for punctate PPK to a 9.98-cm interval, flanked by markers D15S534 and D15S818.

Clinically, PPPK present as an asymptomatic, tiny, hyperkeratotic punctate papules on the palmoplantar surface. In PPPK, involvement is usually bilateral with no case unilateral linear PPPK reported in the literature.[2] Nail abnormalities in the form of longitudinal ridging, onychorrhexis, onychoschizia, trachyonychia, and notching can be seen.[6] Individual with PPPK lack associated systemic features; however, spastic paralysis, ankylosing spondylitis, facial sebaceous hyperplasia, and atopy have been reported.[2] An association with gastrointestinal and pulmonary malignancy is possible.[2,7,8] Because the affected individuals appear to be at increased risk of

Table 1: Differential diagnosis of punctate palmoplantar keratoderma

	Clinical features	Histopathology
Punctate PPK[1,2]	1. Asymptomatic 2. Age at onset is variable, between 10 and 70 years 3. Tiny, hyperkeratotic papules on the palmoplantar surface 4. Yellow waxy plaques of focal palmoplantar keratoderma 5. Nail changes may be seen 6. Family history may be present	1. Hyperkeratosis 2. Orthokeratosis 3. Columnar parakeratosis
Punctate porokeratosis[9]	1. Asymptomatic 2. Age at onset is variable, between 15 and 60 years 3. Multiple punctate seed-like keratoses and/or pits are present on the palm and sole 4. Occasionally occurs in a linear configuration	1. Column of parakeratotic keratinocytes (lamellar parakeratosis) (coronoid lamella)
PEODDN[10,11]	1. Asymptomatic 2. Mostly present from birth 3. Resembles comedo naevus, comprising small keratotic papules with a central plugged pit in a linear distribution, usually on palm and soles	1. Dilated comedo-like epidermal invaginations filled with parakeratotic plugs 2. Keratinocytes with vacuolated cytoplasm and pyknotic nuclei
Arsenical keratosis[12]	1. Small areas of hyperkeratosis (resembling corns) present on the fingers, backs of the hand, and more proximal parts of extremities 1. Induration, inflammation, and ulceration occurs when the lesion becomes malignant	1. A range of changes may be seen from a benign looking hyperplasia or dysplasia to frank Bowen's disease
Striate PPK[13,14]	1. Onset in infancy or first few years of life 2. Linear pattern of skin thickening on palm>sole 3. 1.Nail changes may be seen 4. Associated with wooly hair and cardiomyopathy 5. Family history may be present	1. Acanthosis 2. Hypergranulosis 3. Hyperkeratosis
Naegeli-Franceschetti-Jadassohn syndrome[15]	1. Autosomal dominant 2. Age of onset 1–5 years 3. Reticular pigmentation on neck, chest, and abdomen 4. Diffuse punctate palmoplantar keratoderma ± linear pattern 5. Hypohidrosis 6. Severely affected teeth 7. Absence of dermatoglyphics	1. Hyperpigmentation of the basal layer 2. Dermal melanophages 3. Reduction in sweat glands 4. Colloid-amyloid bodies in the papillary dermis
Punctate accentuated keratosis[16] (dermatopathia pigmentosa reticularis)	1. Age of onset birth or early childhood 2. Clinical triad of reticulate hyperpigmentation, mild non-scarring alopecia, and mild onychodystrophy 3. Punctate or diffuse palmoplantar hyperkeratosis	1. Interface dermatitis

Table 2: Distinctive differentiating features in between striate PPK, punctate PPK, and present case (LPPPK)

Striate PPK[13,14]	Punctate PPK[1,2]	Present case (LPPPK)
Focal hereditary PPK	Focal hereditary PPK	Focal hereditary PPK
Onset in infancy or first few years of life	Age at onset is variable, between 10 and 70 years	12 years
Bilateral Linear bands of hyperkeratosis (Hyperkeratotic plaques) present on the volar fingers, palms and soles Linear presentation is common	Bilateral Tiny, hyperkeratotic papules on the palmoplantar surface Coalesces to form yellow waxy plaques of focal palmoplantar keratoderma Linear presentation is unusual	Unilateral Hyperkeratotic papules coalesces mainly over pressure point areas of sole to form yellowish waxy plaques Linear lesions on palm and sole
Elbow and knee involved invariably, hand may show callous formation	Elbow and knee involvement not present, no callous formation in hand	Elbow and knee involvement not present, no callous formation in hand
Variable nail changes may be seen	Variable nail changes may be seen	No nail changes
Associated with wooly hair and cardiomyopathy	Association with gastrointestinal and pulmonary malignancy is possible	No associated abnormality
Histopathological features consist of hyperkeratosis, hypergranulosis, acanthosis	Histopathological features consist of hyperkeratosis, orthokeratosis, columnar parakeratosis	Histopathology showed hyperkeratosis, orthokeratosis no acanthosis and hypergranulosis

PPPK: Punctate palmoplantar keratoderma, PPK: Palmoplantar keratodermas

developing malignant conditions, a regular follow-up with thorough history taking and clinical examination is mandatory in cases of hereditary punctate PPK.[8]

Histopathological features of PPPK are hyperkeratotic epidermis without any columns of parakeratosis or elastorrhexis. Electron microscopy shows enlarged nucleoli with abundant tonofilaments in the basal cells and keratohyalin-like granules confined to the upper part of the stratum spinosum.[2]

In this case, we also find the similar histological findings. Due to non-availability, we cannot perform the electron microscopic examination of this case.

Our patient is also presented with an asymptomatic, tiny, hyperkeratotic punctate papules on the palmoplantar surface with unilateral left sole and left palm involvement without any associated diseases and we had made the differential diagnosis of linear PPPK,[1,2] linear punctate porokeratosis,[9] porokeratotic eccrine ostial and dermal duct naevus (PEODDN),[10,11] arsenical keratosis,[12] striate PPK,[13,14] Naegeli-Franceschetti-Jadassohn syndrome,[15] and punctate accentuated keratosis.[16]

Above differential had been ruled out by their classical clinical presentation and histopathological features [Tables 1 and 2].[1,2,9-16]

Treatment of keratodermas is generally symptomatic like saltwater soaks, paring, topical keratolytics to specific therapies such as systemic retinoids, psoralens and ultraviolet A (PUVA), and re-PUVA.[2,8] When specific therapy does not works, then patient can be considered for reconstructive surgery with total excision of the hyperkeratotic skin followed by grafting.[2]

Lienemann *et al.* reported 95% improvement following 2-week course of intravenous 5-FU at 250 mg/m^2/day.[17]

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