Case Report

Spiny Keratoderma: The Gritty Tale

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Abstract

Spiny keratoderma is a rare disease; first described by Brown as “punctuate keratoderma.” It is characterized by asymptomatic keratotic pinpoint papules over palms and soles, resembling the old fashioned music box spine. The spiny spicules are often misdiagnosed and has a very subtle histopathological difference from punctate porokeratosis. With only less than 40 cases reported worldwide, we report our case as the very few mentioned in Indian literature.

Keywords: Gritty palm, music box dermatoses, spiny keratoderma

Introduction

Spiny keratoderma is a rare disease, first described by Brown in 1971[1] as “punctuate keratoderma.” It is characterized by asymptomatic keratotic pinpoint papules over palms and soles, resembling the old-fashioned music box spine. The disease can either be familial or acquired which is associated with systemic diseases and malignancies.

Case Report

A 74-year-old male farmer presented with asymptomatic keratotic lesions over both his palms and soles since the age of 14 years. These lesions progressed gradually. There was neither history of any comorbidities including malignancy nor history of exposure to arsenic at work or drinking of water from the well. There was no previous treatment with ayurvedic or homeopathic medicines for any ailments. There were no similar complaints in the family members.

Cutaneous examination revealed symmetrically distributed well-defined 1–2 mm hyperpigmented, hyperkeratotic, firm, pinpoint spiny papules present over both palms and soles and lateral surface of the digits [Figures 1 and 2]. The dorsal surface of both hands and feet was spared. There were no other cutaneous, mucosal, hair, and nail abnormalities. A differential diagnosis of punctate porokeratosis, spiny keratoderma, and arsenic keratosis was considered.

A punch biopsy was performed which showed a compact, narrow column of parakeratosis with underlying hypo to agranulosis. The stratum corneum was orthokeratotic elsewhere. There were no dyskeratotic cells or basal vacuolization noted in the epidermis below the parakeratotic column [Figures 3-5]. The dermis was unremarkable. Step sections also did not reveal the dyskeratosis and degeneration of the basal layer of epidermis. In view of these histological findings, the diagnosis of “spiny keratoderma” was finalized.

A battery of investigations were done for systemic evaluation. Complete blood count with peripheral smear, liver function test, renal function test, erythrocyte sedimentation rate, chest radiography, and ultrasonography of the abdomen and pelvis were within normal limits.

Discussion

Spiny keratoderma is a rare keratoderma, with less than 40 cases reported worldwide since it was first described by Brown in 1971 as punctate keratoderma. The term “spiny keratoderma” was first coined by Osman et al. in 1992. These spiny lesions have had different names in the past. Herman called it “punctuate porokeratotic keratoderma” as he noted sweat ducts in between the parakeratotic columns.[2] Lestringnant and Berge named it “porokeratosis punctata palmare et plantaris.”[3] The other

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names given were “filiform hyperkeratosis,” “multiple minute palmar-plantar digitate hyperkeratosis,” “punctuate keratosis of the palms and soles,” and “music spine box keratoderma.”

It is either hereditary or acquired. The hereditary form shows autosomal dominant inheritance and appears between the age of 12 and 50 years. It is not associated with any malignancy or systemic disease. It shows male predominance and no racial predilection. The acquired form usually appears after the age of 50 years and shows malignant and systemic disease associations. The malignancies that are associated include various carcinomas, leukemia, and melanoma. The lesion often begins years before malignancy and can persist after treatment. There has been only one case of reversal of the disease after treatment of the malignancy. Many authors suggest that spiny keratoderma can be a paraneoplastic phenomenon, but whether this association holds true ground or not has not been established. More so, there are no reports of any malignant changes in the spiny spicules itself. The other systemic diseases associated are Darier’s disease, Type IV hyperlipoproteinemia, chronic renal failure, pulmonary tuberculosis, myelofibrosis, and adult polycystic kidney disease with liver cysts. The other risk factors associated with the disease are a history of manual labor which is postulated to cause hyperproliferation and parakeratosis and significant ultraviolet exposure.

Figure 1: Spiny papules over the palms

Figure 2: Spiny papules over the soles

Figure 3: Remarkable narrow column of parakeratosis (×10)

Figure 4: Column of parakeratosis with orthokeratotic stratum corneum elsewhere (×40)
The pathophysiology is yet not established. It has been associated with overexpression of keratins 6 and 16 which are responsible for epidermal hyperproliferation which manifest as the keratotic spicules. A study done by Hashimoto et al. suggested the presence of AE 13, a monoclonal hair-specific antibody expressed in normal hair cortex in the keratotic spicules, suggesting spiny keratoderma to be ectopic hair formation of the palms and soles.

The clinical differential diagnosis of palmoplantar hyperkeratoses is varied and includes punctate porokeratosis, arsenic keratosis, multiple filiform verrucae, Buschke–Fisher–Brauer disease, acrokeratoelastoidosis lichenoides, and nevoid basal cell carcinoma. All the above differentials can be ruled out based on typical histopathology demonstrated by spiny keratoderma. The closest histopathological differential is palmar porokeratosis. Histopathologically, it can be differentiated from porokeratosis by the absence of dyskeratotic cells in epidermis below the cornoid lamella and basal cell vacuolization. The following table helps in distinguishing spiny keratoderma from the above differentials [Table 1].

Management involves mechanical debridement such as paring and dermabrasion. Topical treatments such as emollients along with 40% salicylic acid and urea cream, 0.1% tazarotene gel, 5% 5-fluorouracil, 12% ammonium lactate, and 0.002% tacalcitol cream have been used with variable success and recurrences. Oral acitretin 10 mg increasing to 30 mg given for 8 weeks has been shown to give excellent result with at least 18-month clearance. Our patient denied any treatment and was lost to follow-up.

We report this case due to rarity of this condition and to reinforce the subtle histopathological differences between punctate porokeratosis and spiny keratoderma.

Table 1: Histopathological differentials

| Layers of skin | Spiny keratoderma | Porokeratosis | Arsenic keratosis | Verruca |
|----------------|-------------------|---------------|-------------------|--------|
| Stratum corneum | Narrow column of parakeratosis | Coronoid lamella | Hyperkeratosis | Hyperkeratosis with parakeratosis |
| Stratum granulosum | Hypo to agranulosum | Absent | Acanthosis with dysplastic changes | Hypergranulosis |
| Stratum spinosum | Unremarkable | Dyskerotic | Acanthosis with koilocytes | Unremarkable |
| Stratum basale | Unremarkable | Vacuolization | Increased pigmentation | Lichenoid lymphocytic infiltrate with dilated capillaries |
| Dermis | Unremarkable | Lymphocytic infiltrate | Unremarkable | |

Figure 5: No basal cell layer vacuolization (×100)

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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