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“I FORGOT TO SHAVE MY HANDS”: A CASE OF SPINY KERATODERMA

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Abstract

A 57 year old Caucasian man presented with multiple asymptomatic spiny papules on the palms and soles that he had been shaving off with a razor for many years. He was otherwise healthy with no personal or family history of skin disease or malignancy. A diagnosis of spiny keratoderma of the palms and soles or “music box spine dermatosis” was made. The clinical, histologic, and electron microscopic features of spiny keratoderma are distinct. This entity previously had multiple classifications and it is important to distinguish it from other keratodermas as some keratodermas can be linked to cutaneous and internal malignancies and conditions: poly cystic kidney disease, liver cysts, Darier’s disease, and hyperlipoproteinemia among others. Spiny keratodermas can have systemic associations and do not resolve spontaneously. Treatment is generally ineffective.

Introduction

Individuals with spiny keratoderma, (“music box spine” keratoderma) have keratotic papules limited to the palms and soles. Spiny keratoderma has multiple classifications and encompasses several entities. Associations with systemic disease or malignancy occur in some acquired cases. We present the case of a man with acquired spiny keratoderma who had no known associated disease.

Case report

A healthy 57 year-old Caucasian man presented to his dermatologist for regular follow-up. As he shook his hand, the dermatologist noted a spiny texture to the patient’s palms. Upon questioning, the patient responded matter-of-factly: “I forgot to shave my hands today.”

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Additional history revealed the man had noted the gradual appearance of hard, asymptomatic lesions on both palms for several years and had self-treated these by shaving. The patient had no known exposure to arsenic or well water and did not have this condition in his first four decades of life. A review of systems was unremarkable and regular visits with his primary care physician for age-appropriate screenings had not revealed any findings concerning for systemic disease. Family history was negative for keratoderma, ichthyosis or other dermatologic diseases.

Skin examination revealed multiple 1–2 mm spiny papules and hyperkeratotic plugs symmetrically involving the palms and soles as well as the lateral surfaces of digits. These lesions resembled the spines of a music box (Figure 1a). The patient had not been aware of the lesions on his soles.

Punch biopsy of one of the palmar papules revealed large columns of keratin arising from the epidermis in an area with focally decreased granular layer and parakeratosis (Figure 1b).

Discussion

In 1971, Brown¹ reported the first case of a keratosis marked by numerous tiny spicules on the palmoplantar surface. These spines have been likened to the spines of an old-fashioned music box and correspond to columns of keratotic material over a hypogranular epidermis.

There are multiple classifications of “music box spine” keratotic papules limited to the palms and soles, including filiform hyperkeratosis,² minute digitate hyperkeratosis,³ punctate keratoderma,¹ punctate porokeratotic keratoderma,⁴ porokeratosis punctata palmaris et plantaris⁵ and spiny keratoderma.⁶ The term spiny keratoderma of the palms and soles^{4,6–10} is now favored because it emphasizes the distribution and morphology of the spiny projections and because the clinical, histologic and electron microscopic features¹¹ of this entity are distinct from porokeratosis.

Zarour and colleagues classified various filiform hyperkeratoses according to sites of involvement and histopathology¹². McGovern and Gentry proposed a similar scheme that replaces “porokeratotic” with “parakeratotic” and replaces “filiform hyperkeratosis” with “spiny keratoderma.”⁷ (Table 1) These alterations in terminology place the diversely labeled cases of music box spine lesions into a more accurate classification scheme based on the morphology, distribution, and histopathology of lesions...

The differential diagnosis of palmoplantar hyperkeratosis can be extensive. Since the filiform appearance is typical, clinical inspection can often distinguish spiny keratoderma from entities such as verrucae, nevoid basal cell carcinoma lesions, and pitted keratolysis. In addition, patients with multisystem disorders that display palmoplantar keratoses, such as Cowden’s disease, will have other mucocutaneous and systemic findings to support these diagnoses. The main distinction, therefore, is between spiny keratoderma and other punctate keratodermas (Table 2). In unclear cases, histopathology can aid in making a diagnosis.

Associations with internal malignancy or systemic disease have been reported in some acquired cases necessitating a good medical history and regular screenings. Reported malignancies include bronchial carcinoma,¹³ renal cancer,¹⁰ rectal carcinoma,¹⁴ breast cancer,⁹ and malignant melanoma.¹⁵ The spiny keratoderma of the palms and soles often begins years before the cancer and persists after its removal. Malignant transformation of spiny keratoderma lesions themselves has not been reported to date. We cannot rule out that our patient’s spiny keratoderma is not an early manifestation of an as yet undetected internal malignancy.

Besides neoplasia, associations with Darier's Disease,¹² chronic renal failure,¹⁶ myelofibrosis,² type IV hyperlipoproteinemia,⁸ and adult polycystic kidney disease with liver cysts¹⁷ have been reported. The connection between spiny keratoderma and these systemic diseases remains unclear, but since this dermatosis is likely underdiagnosed, more reported cases will enable further understanding of this relationship.

Treatment of spiny keratoderma is difficult and unsatisfying. The spiny lesions are bothersome primarily for cosmetic reasons. Management involves mechanical debridement, such as paring and dermabrasion, and topical treatments, such as urea, salicylic acid and vitamin A or retinoids. Maintenance therapy is required to prevent relapse with all forms of treatment. Beyond addressing the aesthetic aspect of spiny keratoderma, the awareness and management of possible underlying malignancy and systemic conditions in the acquired form are important. Complete personal and family history, regular medical examinations, and age-appropriate cancer screenings are essential in these patients for early detection of any associated internal disease.

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References

1. Brown F. Punctate keratoderma. *Arch Dermatol* 1971;104:682–683. [PubMed: 5131713]
2. Mehta RK, Mallet RB, Green C, Rytine E. Palmar filiform hyperkeratosis (FH) associated with underlying pathology? *Clin Exp Dermatol* 2002;27:216–219. [PubMed: 12072012]
3. Guarneri C, Guarneri F, Vaccaro M, Borgia F, Cannavo SP. Multiple minute digitate hyperkeratosis. *International Journal of Dermatology* 2005;44:664–667. [PubMed: 16101869]
4. Friedman SJ, Herman PS, Pittelkow MR, Su WPD. Punctate Porokeratotic Keratoderma. *Arch Dermatol* 1988;124:1678–1682. [PubMed: 2460029]
5. Saka EL, Gentry RH. Porokeratosis punctata palmaris et plantaris: A new entity? *J Am Acad Dermatol* 1985;13:908–912. [PubMed: 2933431]
6. Osman Y, Daly TJ, Don PC. Spiny keratoderma of the palms and soles. *Acad Dermatol* 1992;26:879–881.
7. McGovern TW, Gentry RH. Spiny Keratoderma: case report, classification, and treatment of music box spine dermatoses. *Cutis* 1994;54:389–394. [PubMed: 7867380]
8. Urbani CE, Moneghini L. Palmar spiny keratoderma associated with type IV hyperlipoproteinemia. *J Eur Acad Dermatol Venereol* 1998;10:262–266. [PubMed: 9643333]
9. Hillion B, Le Bozec P, Moulouget-Michau I, Blanchet-Bardon C, Petit A, Stefan J, Civatte J. Hyperkeratose palmo-plantaire filiforme et cancer du sein. *Ann Dermatol Venereol* 1990;834–836. [PubMed: 2151590]
10. Beylot C, Taieb A, Bioulac P, Doutre MS, Houdee G. Hyperkeratose palmoplantaire filiforme et neoplasie viscerale. *Ann Dermatol Venereol* 1982;109:747–748.
11. Hashimoto K, Toi Y, Horton S, Sun TT. Spiny keratoderma—a demonstration of hair keratin and hair type keratinization. *J Cutan Pathol* 1999;26(1):25–30. [PubMed: 10189241]
12. Zarour H, Grob J, Andrac L, Bonerandi J. Palmoplantar orthokeratotic filiform hyperkeratosis in a patient with associated Darier disease. *Dermatology* 1992;185:205–209. [PubMed: 1446087]
13. Bianchi L, Orlandi A, Iraci S, Spagnoli LG, Nini G. Punctate porokeratotic keratoderma : its occurrence with internal neoplasia. *Clin Exp Dermatol* 1994;19:139–141. [PubMed: 8050143]
14. Fegueur S, Bilet S, Crickx B, Perron J, Grossin M, Belaich S. Hyperkeratose palmoplantaire filiforme et cancer recto-sigmoidien. *Ann Dermatol Venereol* 1988;115:1145–1146. [PubMed: 2977074]
15. Kaddu S, Soyer P, Kerl H. Palmar filiform hyperkeratosis: a new paraneoplastic syndrome? *J Am Acad Dermatol* 1995;33:337–340. [PubMed: 7615881]

16. Feldman R, harms M. Multiple filiforme hyperkeratosis. *Hautarzt* 1993;44:658–661. [PubMed: 8225976]
17. Anderson D, Cohen DE, Lee HS, Thellman C. Spiny keratoderma in association with autosomal dominant polycystic kidney disease with liver cysts. *J Am Acad Dermatol* 1996;34:935–936. [PubMed: 8621833]
18. Kong MS, Harford R, O'Neill JT. Keratosis Punctata Palmoplantaris controlled with topical retinoids: A case report and review of the literature. *Cutis* 2004;74:173–179. [PubMed: 15499759]
19. Rustad OJ, Vance JC. Punctate keratoses of the palms and soles and keratotic pits of the palmar creases. *J Am Acad Dermatol* 1990;22:468–476. [PubMed: 2138179]
20. Al Mutairi N, Joshi A, Nour-Eldin O. Punctate palmoplantar keratoderma (Buschke-Fischer-Brauer disease) with psoriasis: a rare association showing excellent response to acitretin. *J Drugs Dermatol* 2005;4(5):627–634. [PubMed: 16167422]

Figure 1.

1a. Music box spines present on palms.

1b. Routine histology of biopsy of one of the palmar keratoses shows large columns of keratin arising from the epidermis in an area with focally decreased granular layer and parakeratosis (Hematoxylin and eosin 10X).

Table 1**Classification of Filiform Hyperkeratosis**

Types of filiform hyperkeratosis <i>Spiny keratoderma</i>	Topography	Histology	Other nomenclature
Type Ia: Palmoplantar porokeratotic FH <i>Palmoplantar parakeratotic SK</i>	Palms and soles	Columns of parakeratosis	Punctate keratoderma Punctate porokeratotic keratoderma, Porokeratosis punctata palmaris et plantaris neoplasie viscerale, Hyperkeratose filiforme et cancer du sein
Type Ib: Disseminated porokeratotic FH <i>Disseminated parakeratotic SK</i>	Shoulder, chest, arms Widespread with palmoplantar sparing	Column of parakeratosis	Porokeratotic minute digitate hyperkeratosis
Type IIa: Palmoplantar orthokeratotic FH <i>Palmoplantar orthokeratotic SK</i>	Palms and soles	Orthokeratosis	Multiple minute palmar-plantar digitate hyperkeratosis Hyperkeratose palmoplantaire filiforme et cancer recto-sigmoïdien
Type IIb: Disseminated orthokeratotic FH <i>Disseminated orthokeratotic SK</i>	Widespread with palmoplantar sparing	Orthokeratosis	Hyperkeratose disseminee familiale Multiple minute digitate hyperkeratosis Minute aggregate keratosis Hiperkeratosis multiple minuta y digitata Minute aggregate keratosis Hyperkeratose filiforme diffuse
Type III: FH in eccrine hamartoma <i>SK in eccrine hamartoma</i>	Linear, involving the palms, soles, limbs, neck and axillae	Column of parakeratosis associated with the sweat duct structures or skin appendages	Porokeratotic eccrine ostial and dermal duct nevus

Classification of filiform hyperkeratosis according to Zarour et al¹² with modifications proposed by McGovern and Gentry⁷ FH filiform hyperkeratosis, SK spiny keratoderma

Table 2Clinical Variants of Punctate Palmoplantar Keratoderma Adapted from Kong¹⁸, Rustad¹⁹, Al-Mutairi²⁰

Acquired keratoses	Classic clinical description	Associations
Arsenical keratoses	Round, verrucous, or acuminate keratotic; palmoplantar surface	Angiosarcoma of the liver Nonmelanoma skin cancer Bronchial adenocarcinoma
Idiopathic punctuate PPK	Multiple, small, hyperkeratotic papules, spicules, or nodules on the palms and soles	Internal malignant conditions, seen with increasing age
Spiny keratoderma, acquired form	Multiple asymptomatic, thin, spiny keratotic projections with music box appearance; palmoplantar surface	Breast, renal, colorectal, and lung carcinoma Melanoma Type IV hyperlipoproteinemia Darier's disease Chronic renal failure Myelofibrosis Polycystic kidney disease with liver cysts
Punctate PPK of the palmar creases	Discrete, sharply margined, hyperkeratotic, conical, 1–5 mm depressions confined to flexural creases; may be painful, most common in Afro-Caribbean descent	Dupuytren's contracture Striate keratoderma Knuckle pads
Hereditary keratoses	Classic clinical description	Associations
Buschke-Fisher-Brauer disease Keratosis punctata Keratosis papulosa	Multiple 1–2 mm punctuate keratoses of the palmoplantar surface; AD; develops between ages 12–30	Longitudinal nail dystrophy Lichen nitidus Ichthyosis Atopy Recalcitrant warts Increased risk of transformation to malignancy
Spiny keratoderma, hereditary form	Small keratotic spines over entire palmoplantar surface, resembles the spines of music box; AD; develops between ages 12–50	No known increased risk of malignancy
Acrokeratoelastoidosis lichenoides	2–4 mm round to oval papules on the borders of hands, feet and wrists, may be umbilicated and become confluent; AD; late onset	Darier's disease Cowden's disease