# Idiopathic Spiny Keratoderma: A Report of Two Cases and Literature Review

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## Abstract

*Spiny keratoderma is a rare and likely underreported condition that presents with punctate hyperkeratotic growths localized to the palms and soles. We present two cases of clinically diagnosed spiny keratoderma. Although the lesions were asymptomatic, patients are at risk of an underlying internal malignancy with this condition, so diagnosis is crucial. Neither men were seeking treatment for the lesions when they were discovered, suggesting that this condition may be much more common than reported. Patients with histories of manual labor, increased UV exposure, and non-melanoma skin cancer (NMSC) may also be at higher risk for developing spiny keratoderma.[1] The epidemiology, histopathologic features, differential diagnosis, and current treatments for spiny keratoderma are reviewed.*

## Introduction

Spiny keratoderma is a rare palmoplantar keratoderma that presents with keratotic, pinpoint papules on the palms and soles. There are both hereditary and acquired forms. When found, a thorough history and physical examination are warranted as there are case reports of spiny keratoderma being associated with underlying internal disease and malignancy of the kidney, colon, breast, lung, and skin.[2] Acquired spiny keratoderma usually manifests after 50 years of age and may be associated with manual labor.[1,3] We present two cases in older men with spiny keratoderma of one to 20 years' duration, and with no underlying malignancy or systemic disease to date.

## Case Report

**Case 1**

An 84-year-old male presented for a full-body skin examination. Upon shaking hands with the patient, we noted diffuse, 2 mm to 3 mm spiny papules on both palms (Figures 1-3) without involvement of the soles. The patient stated he slowly developed these lesions in his 60s, and the lesions are and have always been asymptomatic. His past medical history was negative for any internal malignancies, and he was followed regularly with a family practitioner. He was also current with age-appropriate screenings and examinations. His social history was significant for a long career performing outdoor manual labor while working for a phone company. He had no known direct arsenic exposure or prior radiation treatment. Previous dermatologic history included three basal cell carcinomas in his 70s and 80s that were successfully treated with surgical excision. To treat the spiny projections, he had attempted to "sand" them for a period with some success, but they would always return, and eventually he lost the enthusiasm to do so. He also used trials of salicylic acid and urea, which helped to soften the spines but never provided complete resolution. Although he was embarrassed for many years about his condition, it now no longer bothered him.

**Case 2**

A 67-year-old Caucasian male presented with a one-year history of insidiously growing, pinpoint hyperkeratotic papules projecting from his palms bilaterally (Figures 4-5). He presented to the clinic for skin examination at six-month follow-up for removal of cutaneous squamous cell carcinomas. Upon shaking his hand, the spiny projections were noted. He stated they were present during the last surgery but were less noticeable and not concerning to him at the time. His past medical history included surgical removal of squamous cell carcinomas from his right temple and left forearm. He had been a gun and weapons enthusiast for his entire life, spending significant time using his hands to maintain and fire his weapons and many hours outside without sun protection. The patient was referred back to his primary care physician for internal evaluation. After colonoscopy, chest X-ray and blood work, no internal derangements were noted.

## Discussion

Brown reported the first case of spiny keratoderma in 1971 when he described punctate keratotic projections on the palms of a 20-year-old male.[3] Spiny keratoderma presents with numerous, flesh-colored, well-marginated keratotic papules on

**Table 1. Treatment options for spiny keratoderma[6,8-14]**

| Treatment | Course | Results | Follow-up |
|---|---|---|---|
| Oral acitretin | 10 mg start dose; gradually increased to 30 mg for 8 weeks | Improvement over 4 weeks | At 18 months, still clear |
| Topical tazarotene gel | 0.1% applied once daily for 1 week | Brisk irritant dermatitis with residual improvement of lesions | Not reported |
| Topical 5-FU cream | 5.0% applied twice daily for 2 weeks (with occlusion for resistant lesions) | Decrease in size and number of lesions | Recurrence within a few weeks of discontinuation |
| Topical tacalcitol ointment | 0.002% applied once daily | Dramatic improvement over 3 months | Not reported |
| Topical ammonium lactate lotion | 5% twice a day | Complete resolution in 2 out of 5 patients | Recurrence within a few weeks of discontinuation |
| Salicylic acid in petrolatum and curettage | 40% applied at night, followed by curettage in the morning | Improvement of lesions (thinner and less painful) | Not reported |
| Salicylic acid gel | 6% applied under occlusion at night | Resolution after four days | Recurrence with treatment cessation |

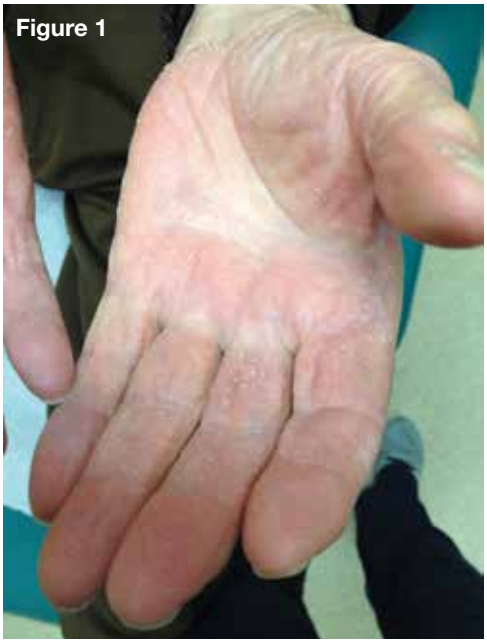
Figure 1


Figure 2

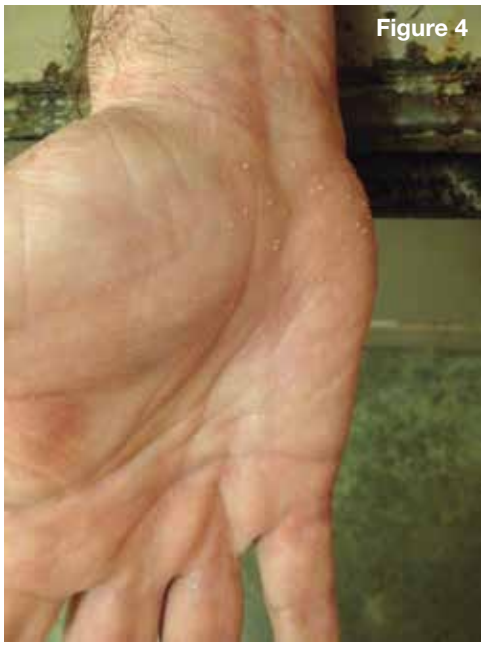
Figure 4


Figure 3

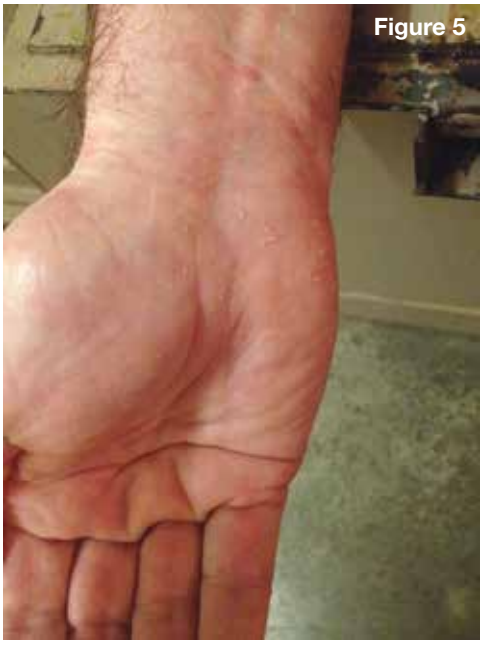
Figure 5

the palms, fingers, and soles. Spiny keratoderma has recently been classified as one of the digitate keratoses. It has been alternatively referred to as punctate porokeratotic keratoderma, music box spine keratosis, multiple minute palmar-plantar digitate hyperkeratosis, and filiform hyperkeratosis, but spiny keratoderma is now preferred.[4]

Spiny keratoderma consists of both inherited and acquired forms, with the acquired form more common in males over 50 and possibly associated with internal malignancy.[5] Risk factors for the acquired variant, as seen in both of our patients, include a history of manual labor.[1] Others include immunosuppression and underlying malignancy of the kidney, colon, breast, lung, and skin.[5] Our patients both had a history of significant UV exposure, which could be another risk factor for spiny keratoderma. However, UV exposure may be a confounding variable in patients with histories of manual labor, too, as our patients invariably performed their years of manual labor under UV exposure.

The pathophysiology of spiny keratoderma is unknown but may involve either abnormal or ectopic keratinization. One study reported biopsy results with overexpression of keratins 6 and 16.[6] These keratins are responsible for epidermal hyperproliferation, which manifests clinically as keratotic projections.[6] The role of ectopic keratinization on the palms and soles was also suggested in a case series involving six other patients.[7] AE13, a monoclonal hair-specific antibody expressed in the normal hair cortex, was also expressed in the compact columns of keratoderma in these patients.[7] In this particular study, electron microscopy showed features of keratinization of a normal hair cortex, including keratinization but without the production of keratohyalin granules.[7] These findings are similar to that of human hair, which suggests that that spiny keratoderma could be representative of ectopic hair formation on the palms and soles. Furthermore, five out of six patients in this study also worked as manual laborers. It has been postulated that repeated trauma through manual labor may explain the hyperproliferation and parakeratosis seen on microscopy, which would support a theory of manual labor causing hand trauma as a risk factor for this condition.[7,8] Although repeated trauma may be a risk factor, the authors did not postulate why some patients' skin is more susceptible than others.

The differential diagnosis includes arsenical keratosis and multiple filiform verrucae, both of which can present in a similar localized fashion on the palmoplantar surfaces. Patients with Cowden's syndrome can also present with palmoplantar keratosis, and therefore a physical exam should be performed to rule out mucocutaneous abnormalities and other manifestations of this syndrome. Hereditary keratoses, including Buschke-Fisher-Brauer disease, hereditary spiny keratoderma, and acrokeratoelastoidosis lichenoides, should be considered in a younger patient.[9] It should be noted that hereditary spiny

keratoderma usually manifests between the ages of 12 and 50 years; however, age is not always a reliable distinguishing factor between the acquired and hereditary subtypes, as there are reports of acquired spiny keratoderma in patients as young as 35 years old.[2,7]

Although biopsy is not essential to establish a diagnosis in all cases, it will reveal a compact column of hyperparakeratosis originating from the stratum corneum, and a hypogranular epidermis directly beneath it. The column is sharply demarcated from adjacent skin that consists of an orthokeratotic stratum corneum. The pathologic differential includes porokeratosis, as the hyperparakeratosis observed can resemble the cornoid lamella present in porokeratosis. These two entities can be distinguished by the presence of dyskeratosis, vacuolated cells, or inflammatory infiltrate seen in porokeratosis, features that are absent in spiny keratoderma. Distinction between spiny keratoderma and porokeratosis should be made either clinically or histologically, as porokeratosis can evolve into SCC or BCC at the clinical site.

Acquired or idiopathic spiny keratoderma has been associated with an underlying neoplasm in up to 50% of cases.[2] The paraneoplastic phenomena include malignancies of the kidney, rectum/colon, breast, and lung. Squamous cell carcinoma, melanoma and chronic lymphocytic leukemia have also been associated with the acquired form.[6] Despite many associations of spiny keratoderma with these underlying malignancies, there is only one case of clearing of the keratoderma after successful cancer treatment.[6] Acquired spiny keratoderma has also been associated with underlying disease, including autosomal-dominant polycystic kidney disease with liver cysts, chronic renal failure, Darier's disease, type IV hyperlipoproteinemia, and pulmonary tuberculosis.[6,9] As such, a complete physical exam should be performed along with implementation of screening guidelines for colonoscopy and/or mammogram in any patient presenting with spiny keratoderma.

There is reported variability in treatments for this stubborn and persistent condition, outlined in Table 1. Treatments with topical emollients and keratolytics such as salicylic acid and urea cream have resulted in little improvement.[10] However, combination therapy with salicylic acid 40% ointment overnight followed by curettage in the morning has proven more effective.[10] Other options include mechanical debridement with dermabrasion and paring. Recent reports of topical tazarotene or acitretin for four weeks have shown more long-standing success.[10,11] Of note, patients on oral acitretin should be followed with routine blood tests that include lipid panels, especially because spiny keratoderma already has an association with hyperlipidemia. In one patient, 5% 5-FU procured successful results, and topical tacalcitol achieved success in another.[12,13] 5-FU and tacalcitol have shown marked improvement in the spiny projections in treated patients, but recurrences have occurred upon discontinuation.[12,13] For those wishing to be treated, newer medications show some promise in eradicating the lesions; however, treatment must be continued to prevent recurrence.

## Conclusion

Acquired or idiopathic spiny keratoderma is a rare condition that can present exclusively on the palms and fingers, as seen in our patients. Other common presentations involve the soles as well. A thorough intake of family and personal history, appropriate cancer screenings, and regular medical examinations should be performed to rule out underlying disease and malignancy in patients presenting with acquired spiny keratoderma. Furthermore, questioning about risk factors, such as manual labor, UV exposure, and immunosuppression, can help to solidify a diagnosis. Providers must consider the psychological impact and social embarrassment this condition can precipitate and educate patients that, if successful, continued treatment will likely be necessary to prevent recurrence.

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