

UC Davis

Dermatology Online Journal

Title

Spiny keratoderma

Permalink

<https://escholarship.org/uc/item/5kn3k24j>

Journal

Dermatology Online Journal, 19(12)

Authors

Nagler, Arielle
Boyd, Kevin P
Patel, Rishi R
et al.

Publication Date

2013

DOI

10.5070/D31912020706

Copyright Information

Copyright 2013 by the author(s). This work is made available under the terms of a Creative Commons Attribution-NonCommercial-NoDerivatives License, available at <https://creativecommons.org/licenses/by-nc-nd/4.0/>

Peer reviewed

Case Presentation

Arielle Nagler, MD, Kevin P. Boyd, MD, Rishi R. Patel MD, and Hyun-soo Lee, MD

Dermatology Online Journal 19 (12): 2

New York University School of Medicine

Abstract

We present a case of a 48-year-old man with an approximately 30-year history of spiny projections on the palms, which were histopathologically consistent with spiny keratoderma. Spiny keratoderma is a rare entity of unknown etiology that has been described with both hereditary and acquired variants. The hereditary form, which is most likely the diagnosis in our patient, manifests at a younger age and is benign. The acquired variant, which presents in older adults, has been associated with a variety of systemic diseases and malignant conditions. In patients suspected of having acquired spiny keratoderma, an evaluation for malignant conditions may be warranted.



Case synopsis

A 48-year-old man with a past medical history of hypertension presented to the dermatology clinic at Bellevue Hospital Center for evaluation of small spines on his palms. At the time of presentation, the patient recounted that he had first noticed the projections on his palms around the time that he was enrolled in high school. He states that they are asymptomatic and he has not noticed any change in the number of lesions since they first developed. He grew up on the Lower East Side of Manhattan, which makes exposure to arsenic unlikely. He did, however, spend the summers in the southern part of the United States where he would drink well water. He currently has a desk job. He was unsure if other family members had similar lesions on their palms.

At the time of evaluation the patient was also complaining of recurrent draining jaw cysts. These cysts developed after jaw trauma four years prior. At that time, open reduction and fixation of the mandible was performed. No other family members had similar jaw cysts.

Physical Examination: On the palmar aspects of the hands, including both the palms and fingers, there were many brown, keratotic projections that were approximately 1-mm in size. The dorsa of the hands were unaffected and there were no hair or nail changes. A fibrotic sinus tract was present on his right jaw without drainage, erythema, warmth, or fluctuance. His dentition was good. There were no oral lesions.

Histopathology: A punch biopsy specimen was obtained from his left palm. There are tall, narrow columns of parakeratosis with preservation of the underlying granular layer that protrudes above the surface of the adjacent cornified layer. At the base, there is slight invagination of the epidermal surface with a thinner granular layer.

Diagnosis: Spiny keratoderma

Discussion: Spiny keratoderma is a rare disorder of multiple, firmly-attached, keratotic projections on the palms and soles. These spines resemble those of a music box [1]. Typically, the lesions are asymptomatic, but some patients report slight pain with pressure or mild pruritus. It was first described in 1971 by Brown as punctate keratoderma [2]. Confusingly, this entity has been referred to by a variety of terms, which include punctate porokeratotic keratoderma, palmoplantar filiform hyperkeratosis, and minute digitate hyperkeratosis [3]. Spiny keratoderma is now the favored term because it more accurately describes the clinicopathological findings.

Histopathologically, the spiny projections represent columns of parakeratosis that arise from the inter-adnexal epidermis. The underlying epidermis tends to have an attenuated granular layer but no dyskeratosis or vacuolar changes.

The etiology of spiny keratoderma is unknown. Both hereditary and acquired cases have been described. One review of the literature found 28 cases of spiny keratoderma reported. Of those, approximately 19% were thought to be hereditary [4]. The hereditary form, which is an autosomal dominant trait, typically appears between the ages of 12 and 50 [5]. Hereditary cases are considered benign and isolated. No malignant conditions or other systemic diseases have been associated with these hereditary cases [6].

Acquired spiny keratoderma, however, has been associated with both systemic disease and malignant conditions. This variant typically presents after the age of 50 [5]. Spiny keratoderma has been found in association with rectal [7], bronchial [8], renal [9], and breast carcinomas [10], as well as with leukemia [11], squamous-cell carcinoma of the skin [12], and melanoma [13]. Many of the cases of spiny keratoderma preceded the diagnosis of malignant conditions; however, some cases were coincidental [4]. Although there appears to be a clear association with malignant conditions, only one case of spiny keratoderma has been reported to improve after the successful treatment of the cancer [12]. Many authors favor a paraneoplastic phenomenon but whether the occurrence of spiny keratoderma and a malignant condition is coincidental or truly paraneoplastic remains unclear. No malignant transformation of the spiny lesions themselves has ever been reported [3].

Cases of spiny keratoderma also have been associated with systemic diseases. In one review, 34% of cases of spiny keratoderma were associated with other systemic diseases, which included Darier's disease [14], type IV hyperlipoproteinemia [15], chronic renal failure [16], and pulmonary tuberculosis (TB) [17]. It is unclear whether some of these cases are coincidental. However, in one case the patient developed TB and spiny keratoderma simultaneously and the spiny keratoderma improved with treatment of the TB [17].

The pathophysiology of spiny keratoderma has yet to be elucidated. Several studies have attempted to investigate the mechanism. One study found that keratins 6 and 16, which are markers of hyperproliferating cells, were over expressed in the keratotic projections, which explained the parakeratosis observed in these lesions [18]. In another case series, the authors analyzed six patients with spiny keratoderma using monoclonal hair specific anti-keratin antibodies. AE13, which is an antibody that is specific to a keratin expressed in normal hair cortex, was positive in the columnar lesions. Using electron microscopic examination, the authors also found keratinization without production of keratohyaline granules, which is another feature of the normal hair cortex. Taken together, the authors concluded that spiny keratoderma represents ectopic hair, albeit aberrant, formation on the palms and soles.[19]

The differential diagnosis for a patient with spiny keratoderma includes porokeratosis, arsenical keratoses, and multiple filiform verrucae [20]. The diagnosis of porokeratosis can be excluded on the basis of the histopathologic features. In porokeratosis, in addition to the column of parakeratosis, there is underlying dyskeratosis and vacuolated keratinocytes in the epidermis, which are not observed in spiny keratoderma. Porokeratosis and spiny keratoderma also are clinically distinct; porokeratosis often presents with plaques or annular lesions with a raised ridge at the periphery. The distinction between spiny keratoderma and porokeratosis is important because porokeratosis can be associated with an increased risk of squamous-cell carcinoma at the site. Arsenic exposure may present with spiny keratoses of the palms and/or soles. Arsenic exposure may occur through its presence in drinking water or as an occupational hazard. Arsenic is associated with an increased risk for squamous-cell carcinoma of the skin and other internal malignant conditions. Multiple filiform verrucae, although typically on the face, may also be localized to the palms and soles. Histopathological features include columns of parakeratosis, but unlike spiny keratoderma, these projections are overlying papillomatosis with areas of coarse, thick granular layer and koilocytes.

Once the diagnosis of spiny keratoderma has been made, its treatment has not been well established. Positive results have been obtained with a variety of treatments, which include 5-fluorouracil [21], topical tacalcitol 0.002% ointment (an active form of vitamin D) [22], and acitretin [23]. It also is important to obtain a detailed clinical history from the patient because although spiny keratoderma can be benign and familial, sporadic cases may be a marker for a predisposition to developing malignant conditions.

References

1. McGovern T, Gentry R. Spiny keratoderma: case report, classification, and treatment of music box spine dermatoses. *Cutis* 1994;54:389
2. Brown F. Punctate keratoderma. *Arch Dermatol* 1971;104:682
3. Torres G, *et al.* "I forgot to shave my hands": a case of spiny keratoderma. *J Am Acad Dermatol* 2008;58:344
4. Mevorah B, *et al.* Unilateral spiny hyperkeratosis: case report and review of the literature. *Dermatology* 2008;217:181
5. Anderson D, *et al.* Spiny keratoderma in association with autosomal dominant polycystic kidney disease with liver cysts. *J Am Acad Dermatol* 1996;34:935
6. Walsh S, Bell R. Spiny projections on the palms and soles: spiny keratoderma of the palms and soles. *Am J Dermatopathol* 2010;32:281
7. Fegueux S, *et al.* Hyperkeratose palmo-plantaire filiforme et cancer rectosigmoidien. *Ann Dermatol Venereol* 1988;115:1145
8. Bianchi L, *et al.* Punctate porokeratotic keratoderma--its occurrence with internal neoplasia. *Clin Exp Dermatol* 1994;19:139
9. C CB, *et al.* Hyperkeratose palmo-plataire filiforme et neoplasie viscerale. *Ann Dermatol Venereol* 1982;109:747
10. Hillion B, *et al.* Hyperkeratose palo-plantaire filiforme et cancer du sein. *Ann Dermatol Venereol* 1990;117:478
11. Bernal A, *et al.* A patient with spiny keratoderma of the palms and a lymphoproliferative syndrome: an unrelated paraneoplastic condition? *Dermatology* 2000;201:379
12. Horton S, *et al.* Spiny keratoderma: a common under-reported dermatosis. *J Dermatol* 1998;25:353
13. Kaddu S, *et al.* Palmar filiform hyperkeratosis: a new paraneoplastic syndrome? *J Am Acad Dermatol* 1995;33:337
14. Zarour H, *et al.* Palmoplantar orthokeratotic filiform hyperkeratosis in a patient with associated Darier's disease. Classification of filiform hyperkeratosis. *Dermatology* 1992;185:205
15. Urbani C, Moneghini L. Palmar spiny keratoderma associated with type IV hyperlipoproteinemia. *J Eur Acad Dermatol Venereol* 1998;10:262
16. Feldmann R, Harms M. Multiple filiform hyperkeratoses. *Hautarzt* 1993;44:658
17. Gimenez-Arnau A, Camarasa J. Palmar filiform or spiny hyperkeratosis associated with pulmonary tuberculosis. *J Eur Acad Dermatol* 1994;3:400
18. Kondo S, *et al.* Punctate porokeratotic keratoderma: some pathogenetic analyses of hyperproliferation and parakeratosis. *Acta Derm Venereol* 1990;70:478
19. Hashimoto K, *et al.* Spiny keratoderma--a demonstration of hair keratin and hair type keratinization. *J Cutan Pathol* 1999;26:25
20. Caccetta T, *et al.* Multiple minute digitate hyperkeratosis: a proposed algorithm for the digitate keratoses. *J Am Acad Dermatol* 2012;67:e49
21. Osman Y, *et al.* Spiny keratoderma of the palms and soles. *J Am Acad Dermatol* 1992;26:879
22. Yukawa M, *et al.* Spiny keratoderma of the palms successfully treated with topical tacalcitol. *Acta Derm Venereol* 2007;87:172
23. Scott-Lang V, McKay D. Spiny keratoderma successfully treated with acitretin. *Clin Exp Dermatol* 2013;38:91