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Spiny keratoderma

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Abstract

Spiny keratoderma is a rare entity characterized by filiform keratotic lesions on palms and soles. Although there are some inherited cases the majority are acquired. This last variant can be idiopathic or associated with neoplasms and chronic systemic diseases. We report a new case of spiny keratoderma associated with endometrial carcinoma.

Keywords: spiny keratoderma, parakeratotic spinulose keratoderma, neoplasm

Introduction

Spiny keratoderma has been described in association with chronic systemic diseases and neoplasms. We wish to report a new case of spiny keratoderma associated with endometrial carcinoma.

Case Synopsis

An 83-year-old woman presented with a history of growths on both palms for approximately 5 years. She denied suffering from itching or pain. She had no relevant medical history. Physical examination showed millimetric keratotic papules of both palms (**Figures 1, 2**) that were consistent with parakeratotic spiny keratoderma histologically (**Figure 3**). There was no family history of spiny keratoderma. She was sent to her medical doctor to rule out internal malignancy and the diagnosis of an endometrial carcinoma was made after a computed

tomography scan of the chest, abdomen, and pelvis. It was treated with radical surgery without adjuvant treatment because of the age of the patient. There was no improvement of the cutaneous lesions after the antineoplastic surgery and topical treatment with salicylic acid.

Case Discussion

Spiny keratoderma, also known as palmoplantar filiform hyperkeratosis or music box spine dermatosis [1] is an entity with unknown etiology with less than a hundred cases described in the literature. There are some genetically transmitted cases with autosomal dominant inheritance with common onset during childhood. The majority of the cases (80%) are acquired with onset in the second half of life and can be associated with chronic diseases such as myelofibrosis [2], Darier disease,



Figure 1. *Millimetric keratotic papules of both palms.*

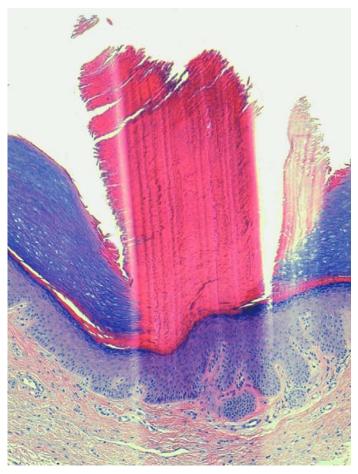


Figure 3. Column of parakeratotic hyperkeratosis projected on the surface of the skin. H&E, $10\times$.

polycystic kidney disease, type IV hyperlipoproteinemia, and neoplastic diseases such as melanoma, leukemia, and lung, breast, and renal carcinomas [3]. This correlation is still unclear and could be coincidental because of the higher incidence of malignancy in patients over 50 years old [4].

There are several hypotheses that try to explain the pathophysiology of this entity: 1) an ectopic hair formation based on a hair-specific antibody (AE13) in the lesions has been detected; 2) overexpression of keratins 6 and 16 (hyperproliferative cells markers) in the keratotic lesion [5] attributable to repeated trauma; 3) overexpression of p63, a transcription factor that regulates the proliferation and differentiation of the keratinocytes [6]; and 4) a disruption of cholesterol synthesis with an induction

of epidermal hyperplasia caused by the coenzyme A reductase inhibitors [1]. The possible association of parakeratotic spiny keratoderma with malignancy would justify the workup for an underlying neoplasm in older age groups [4].

The clinical differential diagnosis includes punctate keratoderma, verrucae, pitted keratolysis, and palmoplantar porokeratosis. It is important to rule out arsenic exposure, also related with keratosis on the palms and soles [7]. However, the history of exposure and cutaneous hyperpigmentation would be helpful in the differential diagnosis. Histologically, it is easier to differentiate these entities from spiny keratoderma, which is characterized by focal filiform columns of orthokeratotic or parakeratotic hyperkeratosis that are projected on the surface of the skin.

Treatment should be offered if the patient complains of functional or cosmetic discomfort. Some therapeutic options include topical creams (urea emollients, 6-10% salicylic acid, 0.003% tacalcitol, 5-fluorouracil or 0.05% tretinoin), oral retinoids, and mechanical debridement with varying results [8]. The lesions usually recur after treatment cessation. Resolution of the keratotic lesions have been described after successful antineoplastic treatment in cases relating to cancers [3].

Conclusion

Spiny keratoderma is a rare condition that can cause functional and cosmetic discomfort. Its diagnosis is important owing to the possible association with an underlying neoplastic disease.

Potential conflicts of interest

The authors declare no conflicts of interests

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