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Case Report

Fracture Reveals a Case of Spiny Keratoderma!

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ABSTRACT

Spiny keratoderma is a rare and likely underreported condition characterised by asymptomatic multiple pinpoint, keratotic projections over palms and soles. We report a case of a 50-year-old female with hereditary spiny keratoderma without any associated disease. Interestingly, the patient consulted us after a fracture of her right middle finger splinted, due to which she was unable to use her hand and wash it as usual and the consequent absence of exfoliation caused by hand usage and washing made the spines prominent. The characteristic clinical presentation with histopathologic and dermoscopic findings helped us to diagnose this case. The differential diagnosis of palmoplantar hyperkeratosis can be extensive. Therefore, it is important to distinguish spiny keratoderma from other palmoplantar hyperkeratosis.

Keywords: Spiny keratoderma, Musicbox spine, Histopathology

INTRODUCTION

Brown reported the first case of spiny keratoderma in 1971 and termed it as 'punctate keratoderma'.[1] It is characterised by pinpoint keratotic spicules over palms and soles and may be hereditary or acquired.

CASE REPORT

A 50-year-old female presented to the dermatology outpatient department with a 20-year history of asymptomatic spiny lesions on both palms, gradually increasing in size and number. Her sister and father had similar lesions. She had a fracture of her right middle finger 1 month back, for which a splint was applied. As she was unable to wash her hands frequently, she noticed an exacerbation of lesions. Cutaneous examination revealed multiple, discrete, firmly adherent and hyperkeratotic papules of 1–2 mm on both palms [Figure 1]. There were no other cutaneous, mucosal, hair or nail abnormalities. The differential diagnoses of spiny keratoderma, multiple filiform verrucae and arsenic keratosis were considered. Complete hemogram, lipid profile, chest X-ray and abdominal ultrasound were normal. Dermoscopy showed brownish, translucent and ovoid projections with normal skin markings. Histopathology showed compact orthokeratotic stratum corneum and a sharply defined column of parakeratotic cells with underlying hypogranulosis in the epidermis [Figure 2]. A diagnosis of hereditary spiny keratoderma was made and she was treated with 5-flurouracil (5%) cream which led to slight improvement.

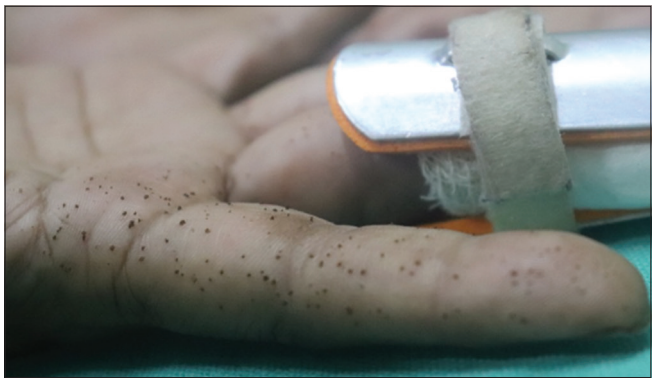

Figure 1: Spiny papules over the palms.

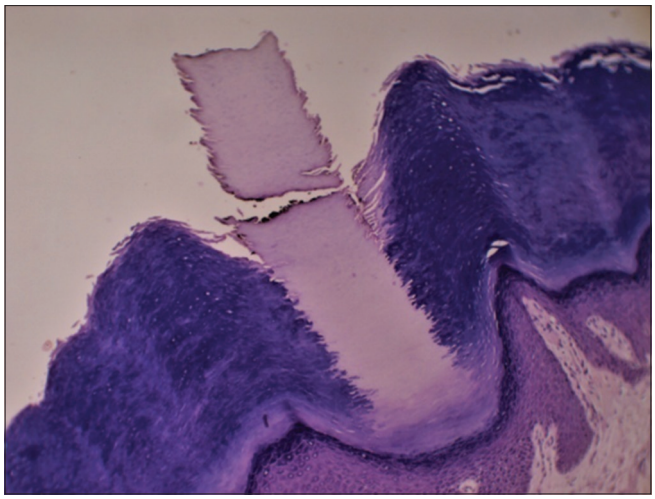

Figure 2: Compact orthokeratotic stratum corneum, sharply defined parakeratotic columns (×400).

DISCUSSION

Spiny keratoderma (porokeratosis punctata palmaris et plantaris, filiform hyperkeratosis and music spine box keratoderma) is a rare entity with 84 cases reported worldwide till now[2] since its initial description by Brown[1] in 1971 as punctate keratoderma. The term 'spiny keratoderma' was coined by Osman *et al*. in 1992.[3]

Several theories have been proposed for the aetiology of spiny keratoderma. The presence of Anti-hair cortex Cytokeratin/ K40 antibody (AE13) (a monoclonal antibody specific to hair, usually seen in normal hair cortex) in the keratotic spicules suggested ectopic hair formation on palms and soles, as the aetiology behind spiny keratoderma.[4] A proposed mechanism is repeated trauma, as cases have been documented in manual labourers. However, this is contradictory to our case, where lesions reappeared due to the absence of friction. We propose that the exfoliation due to the use of hands and hand washing was instrumental in keeping the lesions in check, leading to the prominence of lesions after the patient started wearing a splint and avoiding water. There may also be over- expression of keratins 6 and 16 causing epidermal hyperproliferation.[5] The lesions may precede the malignancy for many years and can persist post-treatment. Some experts suggest that spiny keratoderma might be a paraneoplastic phenomenon, though this association is not firmly established.

The differential diagnoses for spiny keratoderma are – multiple filiform verrucae, punctate porokeratosis and arsenic keratosis. Arsenic exposure can occur occupationally or through contaminated drinking water and may lead to spiny keratosis of the palms and soles. Arsenic is associated with an increased risk of squamous cell carcinoma of the skin and other malignant conditions such as lung cancer and bladder cancer. These conditions can be excluded based on the characteristic histopathology seen in spiny keratoderma.

The condition is either hereditary or acquired. The hereditary type manifests between the ages of 12 and 50 years and has an autosomal dominant mode of inheritance. It is usually not linked to any malignancy or systemic disease. The acquired form is usually seen after the age of 50 years and is associated with malignancies and various systemic diseases, including various carcinomas, leukaemia and melanoma.[6] Other associated systemic diseases are Type IV hyperlipoproteinemia, chronic renal failure, pulmonary tuberculosis, myelofibrosis, Darier's disease and adult polycystic kidney disease with liver cysts.[6]

Various topical treatments have been used with varying degrees of success and recurrence rate, including emollients along with urea cream, 12% salicylic acid, 12% ammonium lactate and 5% 5-fluorouracil. Other treatment modalities include paring and dermabrasion. Management of spiny keratoderma is challenging, and complete resolution is rare. The characteristic clinical, histopathologic and dermoscopic findings helped us to diagnose this case.

CONCLUSION

Spiny keratoderma is a rare condition, making it crucial to differentiate it from other types of palmoplantar hyperkeratosis, which can have extensive range of differential diagnoses.

Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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