

Familial spiny keratoderma without systemic disease in a woman

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DESCRIPTION

Spiny keratoderma is a dermatosis presenting with of multiple projections located on the palms and soles, with a distinct histology characteristic of a parakeratotic column above a hypogranular epidermis.¹

A woman in her 60s presented with gradually increasing, spiny projections on both palms and soles for 3 years. On cutaneous examination, multiple, discrete, firmly adherent, keratotic papules of the size 1–2 mm were noted over both the palms (figure 1A,B) and soles (figure 2A,B). There were no other cutaneous findings and systemic examination was within normal limits. There was no history of any manual labour being done by her except minor household works. There was no history of any treatment with drugs like statins. She was not associated with any comorbid conditions like diabetes, hypertension or hyperlipidaemia. Patient's son around 30 years has noticed similar lesions over both palms and soles since 3 years. Complete hemogram, lipid profile, chest X-ray and abdominal ultrasound were normal. Histopathology of the skin biopsy showed a sharply defined column of parakeratotic cells with underlying hypogranulosis suggestive of spiny keratoderma. A diagnosis of familial spiny keratoderma or 'music box spine keratoderma' was made based on clinical and histological findings. She was treated with topical 20% urea and oral retinoid.

Three types of palmoplantar localised digital keratoses have been classified: spiny keratoderma, arsenic keratosis and multiple filiform verrucae. The index patient did not have history of exposure to arsenic as well as well water. There were no histological features of arsenic keratosis or multiple filiform verrucae visualised in the index case. This condition can be familial, typically presenting in adolescence or adulthood. The list of

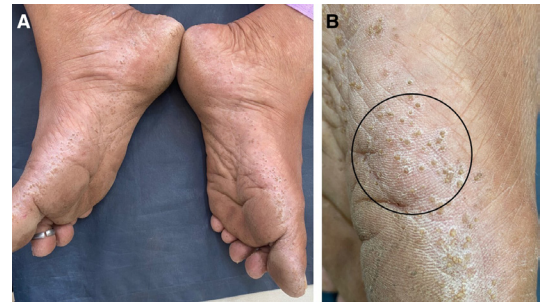


Figure 2 Multiple, small spine like symmetrically distributed lesions over soles (A) with close up view (B) of firmly adherent, keratotic papules (shown by circle mark).

clinical differentials to be considered are—punctate porokeratosis, Darier's disease, epidermodysplasia verruciformis, arsenic keratosis, multiple filiform verrucae, Buschke–Fisher–Brauer disease, acrokeratoelastoidosis lichenoides and nevoid basal cell carcinoma²

There are reports of its association with internal malignancies when the condition presents in later part of adulthood.³ Solid tumours such as squamous cell carcinoma, oesophageal carcinoma, malignant melanoma, colonic cancer and multiple myeloma are the reported malignancies associated with spiny keratoderma of later age onset.⁴ Non-neoplastic conditions include asthma, Darier's disease, tuberculosis and renal abnormalities.⁵ A thorough workup is a must to identify any potential underlying cancer in every case of spiny keratoderma. Spontaneous late adulthood onset of spiny keratoderma without any underlying systemic disease is rare. The index case is one such presentation without any underlying systemic disease and hence the reporting for its rarity.

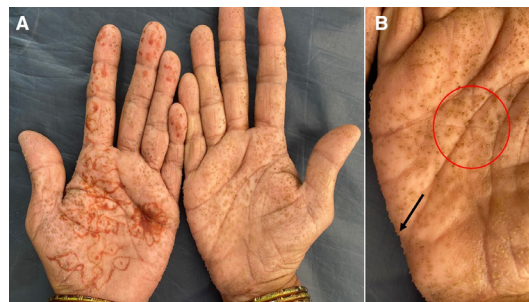


Figure 1 Multiple, small spine like symmetrically distributed lesions over palms (A) with close up view (B) of firmly adherent, keratotic papules (shown by arrow and circle mark).

Learning points

- ▶ Systemic workup is warranted once you diagnose late adulthood spiny keratoderma and further monitor for systemic internal malignancies.
- ▶ Proper history of well water consumption and exposure to arsenic needs to be elicited.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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