Music box spine keratoderma without any systemic manifestation

Sir,

The term spiny keratoderma is used for a dermatosis, presenting with multiple keratotic protrusions that resemble music box spines, and located on palms and soles.[1,2] There are many synonyms, including punctate keratoderma, punctate porokeratotic keratoderma, and porokeratosis punctata palmaris et plantaris.[1] Both familial and acquired cases were reported.

Systemic diseases or malignancies may be associated with acquired cases.

We report a woman with acquired spiny keratoderma without any associated disease. A 28-year-old woman visited the dermatology outpatient department with the complaints of thickening and roughness of hands and feet due to keratotic papules starting on palmar aspect of fingers since the age of 12 years. She had no other significant medical problem. She had no history of environmental arsenic exposure and family history was not contributory.

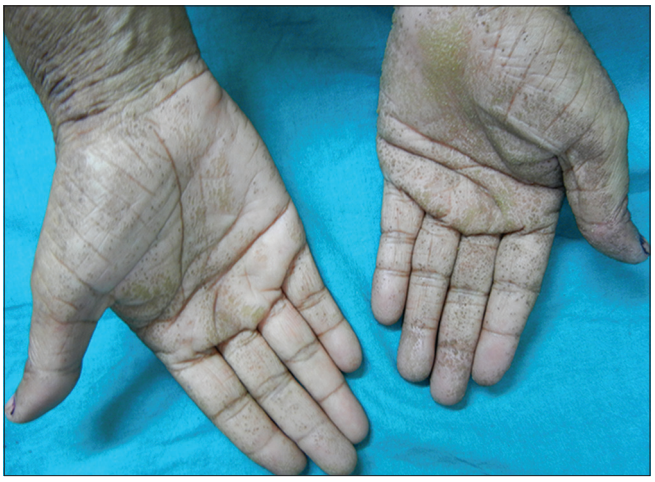

Figure 1: Numerous minute keratotic papules over palms

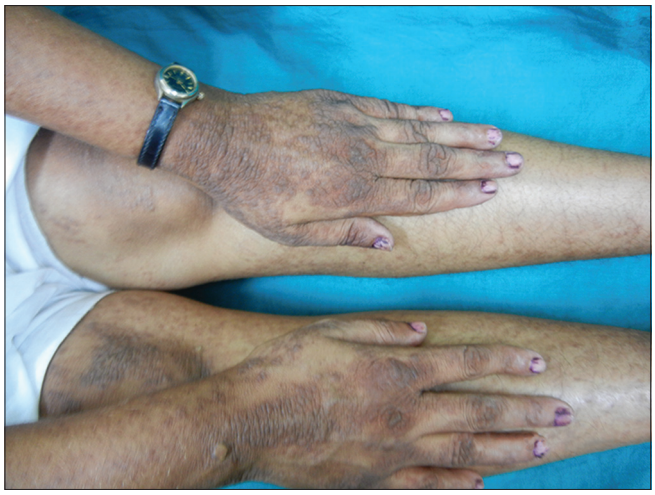

Figure 2: Multiple papules over dorsum of hands and knee

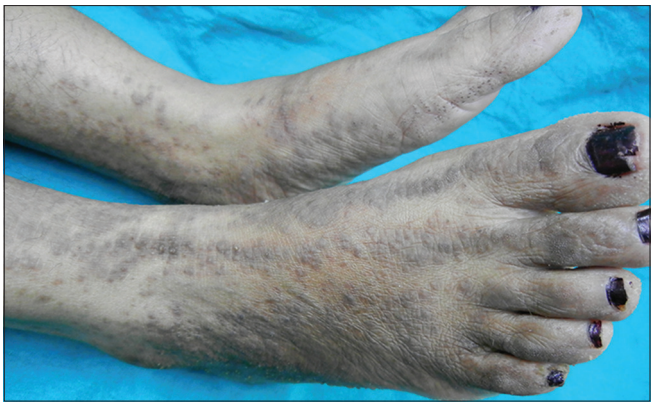

Figure 3: Multiple papules over dorsum of foot

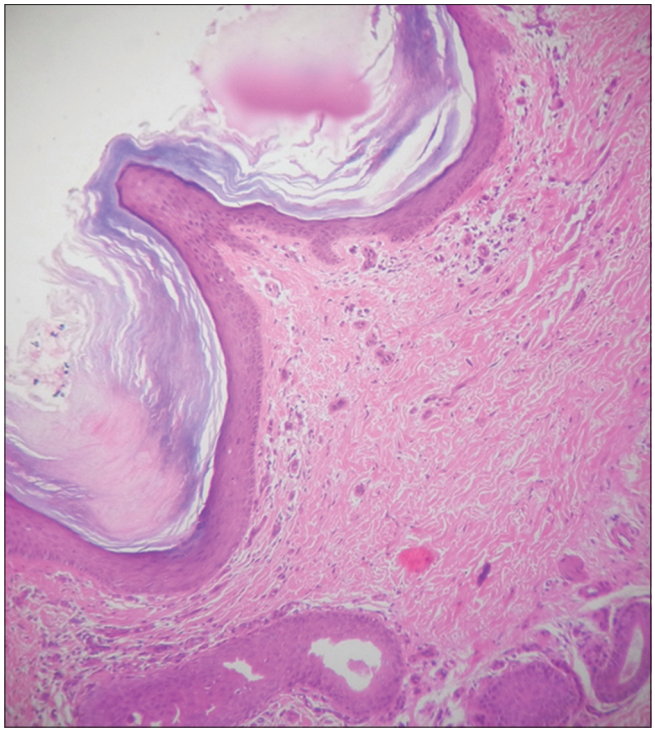

Figure 4: Broad parakeratotic column with thick underlying granular cell layer. At the crest of epidermal cell proliferation, the granular cell layer is reduced gradually (H and E, ×100)

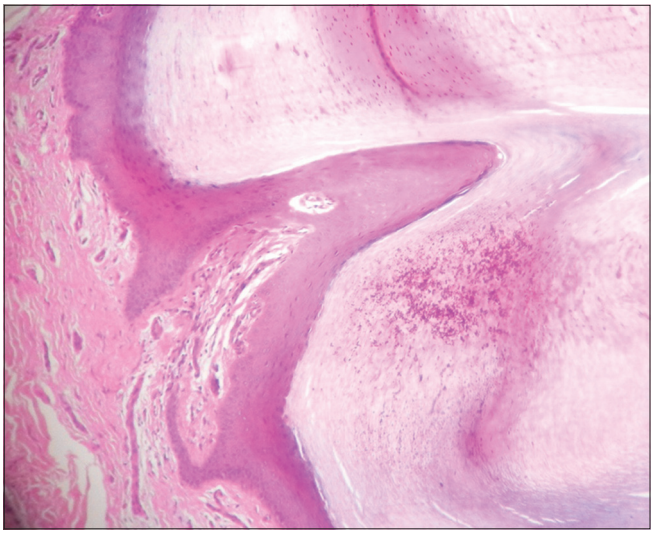

Figure 5: Thick cup-like hyperkeratotic column invaginating at acanthotic epidermis and with a thin underlying granular cell layer (H and E, ×100)

On examination, multiple keratotic papules over palms [Figure 1] and soles were noted. Multiple papules were also observed over dorsum of hands [Figure 2] and feet [Figure 3]. Complete hemogram, lipid profile, and chest radiography revealed no marked abnormalities.

Skin biopsy from a keratotic papule over the palm revealed a broad parakeratotic column with thick underlying granular layer. At the crest of epidermal proliferation, the granular layer is reduced gradually [Figure 4]. Histopathology of a papular lesion of dorsum of hand showed thick cup-like hyperkeratotic column invaginating at acanthotic epidermis with a thin underlying granular layer [Figure 5]. Based on the clinical manifestations and histopathologic findings, wemade a diagnosis of music box spine keratoderma.

Brown described the first case of a keratosis characterized by multiple tiny spicules on the palmo-planter surface in 1971.[3] The spines are similar to the spine of an old fashioned music box, corresponding to columns of keratotic materials over a hypogranular epidermis.

Clinically, spiny keratoderma appears as spiny protrusions or pits with or without keratin plug.

The etiology of spiny keratoderma is not clear. Familial cases and the association with different malignancies such as lung, bronchial, renal, colon, or esophageal cancers were also reported.[4] Melanoma and chronic lymphatic leukemia, dyslipidemia, renal cysts, and myelofibrosis[5] were also found.

However, no such association with other diseases was found in our patient.

Although the diagnosis is clinical, histopathology shows a column of well-defined parakeratotic cells, with underlying hypogranulosis.[6] The characteristic clinical presentation with histopathologic findings helped us to diagnose this case.

Spiny keratoderma and porokeratosis have similar histologic features. But spiny keratoderma does not show vacuolization and/or dyskeratosis of underlying spinous layer, cornoid lamella, or lymphocytic infiltration of the papillary dermis as seen in porokeratosis. Differentiation between punctate keratoderma and porokeratosis is essential because the latter is associated with basal and squamous cell carcinoma.

Spiny keratoderma is difficult to treat, with an unsatisfactory prognosis. Management involves mechanical debridement, such as paring and dermabrasion, and topical treatments such as 5% 5-fluorouracil cream, urea, salicylic acid, 12% ammonium lactate, and retinoids. The frequency of associated malignant disease is unclear, but malignancy may come up even within 30 years.[4] Besides the cosmetic problem of spiny keratoderma, the awareness and management of possible underlying malignancy and systemic conditions are important. We intend to evaluate and follow up the case in a half-yearly manner for as long as possible in the interest of patient and investigation.

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