Sir,

A 90-year-old male was referred from ICU in view of increased roughness of the palmar aspects of both the hands for the past 6 years associated with the appearance of small asymptomatic spicule-like lesions [Figure 1]. The patient was hepatitis C virus (HCV) positive, with Chronic Obstructive Pulmonary Disease with Cor pulmonale with Congestive Heart Failure with right lower lobe pneumonia. There was no history of previous treatment with Ayurvedic or homeopathic medicines. No other significant history was present. There were no similar complaints in the family members.

On examination, multiple, discrete, firmly adherent, hyperpigmented hyperkeratotic, pinpoint, and shiny papules of approximately 1–2 mm were present symmetrically over bilateral palms [Figure 2a and b] with sparing of dorsal surfaces. Similar lesions were present over bilateral soles but they were very sparse [Figure 3a and b]. There were no other cutaneous, mucosal, hair, and nail abnormalities.

Investigations revealed normal blood profile with deranged Renal Function Test (increased blood urea and creatinine), deranged Liver Function Test (raised SGOT/SGPT/ALP/gamma GT, low total protein, and albumin), raised CKMB (Creatine Kinase-MB isoform), and 2D-ECHO abnormality (presence of Atrial Premature Complexes, Dilated Cardiomyopathy, dilated LA/LV, severe global hypokinesia of LV, severe LV systolic dysfunction+ {LVEF 25%}, moderate MR, sclerosed AV, trace AR, mild TR, mild PAH {RVSP 42 mmHg}). Viral markers revealed HCV reactivity. But there was no evidence of malignancy.

However, the patient went against medical advice and refused further investigations.

A differential diagnosis of spiny keratoderma, punctuate keratoderma, and arsenical keratosis was considered. A final diagnosis of music box spine keratoderma was made after clinical consideration and reviewing the literature. The patient denied any further management and hence was lost to follow-up.

Spiny keratoderma is a rare keratoderma with less than 40 cases reported worldwide. It was first described by Brown in 1971 as punctate keratoderma (punctuate keratotic projections) in a case of a keratosis characterized by multiple tiny spicules on the palmo-plantar surfaces. The spines resemble the spine of an old-fashioned music box corresponding to columns of keratotic materials over a hypogranular epidermis.[1] Clinically, it is seen as fine 1–2 mm papules projecting from the palmoplantar surface as spiked, filiform, prickly, minute digitate, or music-box spines.[2] However, the term “spiny keratoderma” was first coined by Osman et al. in 1992.[3] However, it has been a highly controversial entity. It has also been described as “porokeratosis palmaris et plantaris” or “punctate porokeratotic keratoderma” despite histopathological and electron microscopic variation. However, unlike porokeratosis, this entity does not have increased malignant potential.[4] It may also resemble arsenical keratoses. The other names used to describe spiny keratoderma include PPKP2, multiple minute palmoplantar digitate hyperkeratosis, and filiform hyperkeratosis.

It can either present as hereditary (autosomal dominant inheritance) or acquired condition. The hereditary form usually appears between the age of 12 and 50 years with male predominance. The acquired form usually appears after the age of 50 years and has known malignant and
systemic disease associations. The associated malignancies include various carcinomas like those of the lung, bronchus, renal, colon or esophagus, chronic lymphatic leukemia, and melanoma. Various associated systemic diseases include Darier’s disease, type IV hyperlipoproteinemia, chronic renal failure, pulmonary tuberculosis, myelofibrosis, and adult polycystic kidney disease with liver cysts. A history of manual labor causing hyperproliferation and parakeratosis, and significant ultraviolet exposure are believed to be the risk factors.

A study done by Hashimoto et al. suggested the presence of a monoclonal hair-specific antibody, AE 13 which is expressed in normal hair cortex in the keratotic spicules, suggesting spiny keratoderma to be ectopic hair formation of the palms and soles.

Although the diagnosis is clinical, the histopathology shows dense columns of well-defined parakeratotic cells with underlying hypogranular epidermis.

The clinical differentials to be considered include punctate porokeratosis, Darier’s disease, epidermolyssplasia verruciformis, arsenic keratosis, multiple filiform verrucae, Buschke–Fisher–Brauer disease, acrokeratoelastoidosis lichenoides, and nevoid basal cell carcinoma.

Spiny keratoderma is difficult to treat, with an unsatisfactory prognosis. Management involves mechanical debridement such as paring and dermabrasion. Topical treatments such as emollients along with 40% salicylic acid and urea cream, 5% 5-fluorouracil cream, 12% ammonium lactate, 0.1% tazarotene gel, and 0.002% tacalcitol cream and retinoids have been used with variable success and recurrences. Oral acitretin 10 mg increasing to 30 mg given for 8 weeks has been shown to give excellent result with at least 18-month clearance.

In our case, the patient went against medical advice and denied histopathological examination and treatment. From the review of literature, we also concluded that in a case of localized palmoplantar keratoderma with classical music box spine appearance, a clinical diagnosis is possible without the need for histopathological examination since the histopathological findings are nonspecific. A detailed workup is important to rule out any underlying systemic abnormalities. Even though the only hepatic manifestation that has been described in association with spiny keratoderma is liver cysts, from our case, it is possible that hepatitis may be an unrecognized or underreported association since only a handful of cases have been reported and the ones with associated systemic involvement are even fewer.
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Conflicts of interest
There are no conflicts of interest.

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