# Scratch amyloidosis over nose: A rare site of cutaneous amyloidosis

Dear Editor,

Primary localised cutaneous amyloidosis is a rare group of skin disorders characterised by the deposition of extracellular homogenous hyaline material (amyloid) in the dermis without systemic involvement.[1] The different subtypes of primary localised, cutaneous amyloidosis include lichen amyloidosis, macular amyloidosis and nodular amyloidosis. A joint manifestation of lichen and macular amyloidosis is called biphasic amyloidosis.[2,3] The disease is more common in females with macular amyloidosis being the commoner variant. The disease manifests as hyperpigmented macules and patches in a rippled pattern on the extensor aspect of forearms and upper back.

Frictional melanosis is a close differential diagnosis with a clinical and histopathological overlap. Demonstration of amyloid deposits is diagnostic of cutaneous amyloidosis.[3] Here, we discuss an uncommon presentation of cutaneous amyloidosis on the nose.

A 52-year-old woman, with no known comorbidities, presented with a dark discolouration of the nose with itching of 9 months duration. The patient gave a history of frequent scratching of the nose with no oozing or pain. Dermatological examination revealed a well-defined hyperpigmented plaque over the nose [Figure 1].

Dermoscopy showed a hyperpigmented reticular pigment network over a dark background [Figure 2]. Histopathological examination showed pigment incontinence, and the papillary dermis had nodular acellular deposits that were pale eosinophilic in character [Figure 3a]. High power highlighted the focal basal cell vacuolar degeneration with melanophages in the upper dermis [Figure 3b], while the Congo red stain showed birefringence of nodular material in the papillary dermis [Figure 3c]. Immunohistochemistry (IHC) (400x magnification) with serum-associated amyloid (SAA) stained the deposits in the papillary dermis [Figure 4]. All routine investigations to rule out systemic causes of amyloidosis were normal. A final diagnosis of cutaneous amyloidosis was made, and the patient was advised not to scratch the nose and was treated with mid-potent topical corticosteroid cream (0.1% mometasone). After 2 weeks, her

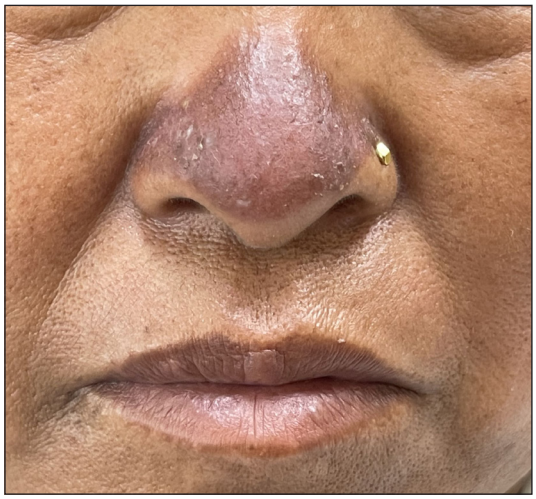

**Figure 1:** Well-defined hyperpigmented plaque over the nose.

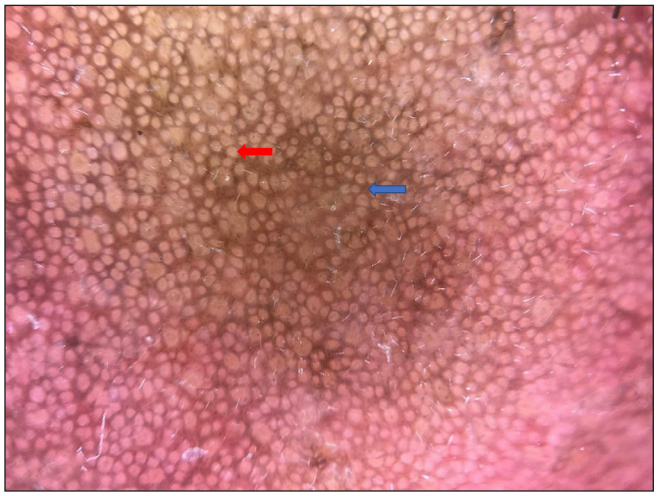

**Figure 2:** Dermoscopy revealed a reticular pigmented network over a brown background. Red arrow indicates accentuation of reticular pigment network and blue arrow indicates sparing of hair follicles. (Dermlite dl4, polarised mode with 10x magnification)

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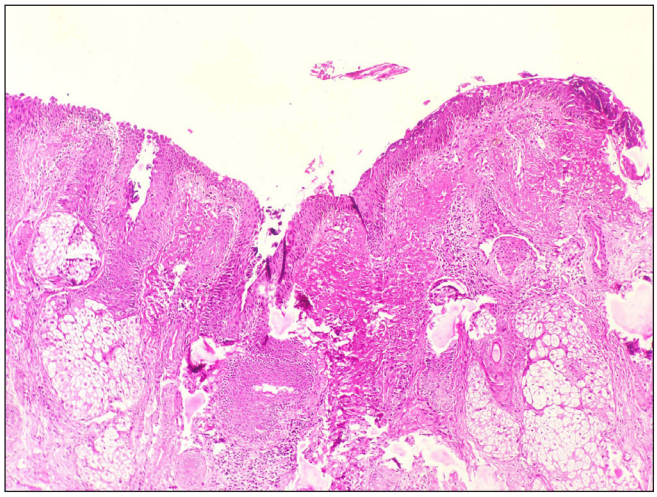

**Figure 3a:** Pale eosinophilic nodular acellular deposits in the papillary dermis (Haematoxylin & eosin, 100x).

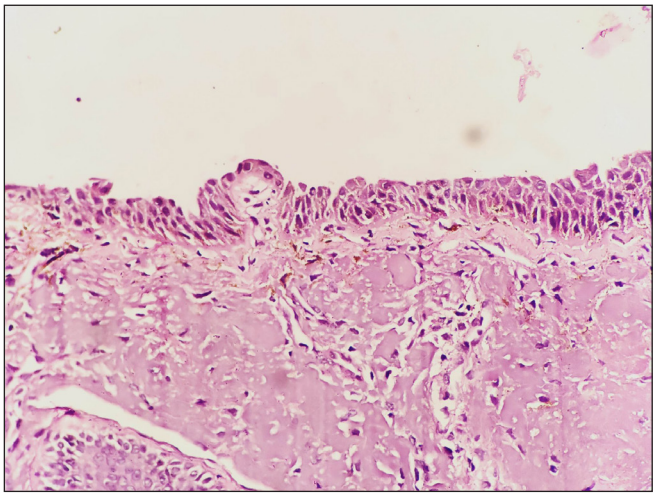

**Figure 3b:** Focal basal cell vacuolar degeneration with melanophages in the upper dermis (Haematoxylin & eosin, 400x).

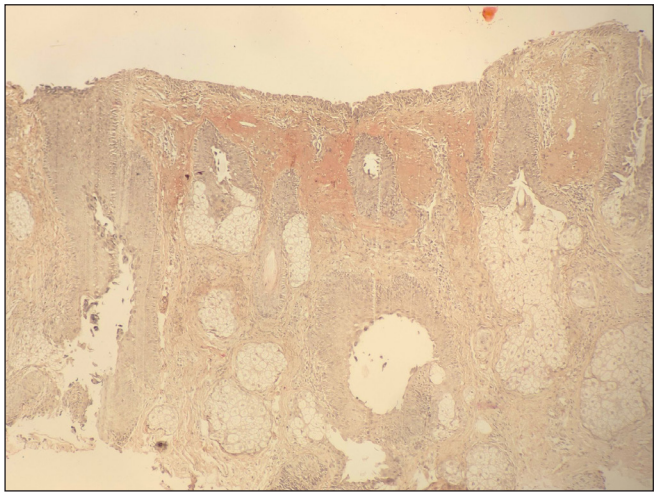

**Figure 3c:** Polarised microscopy showed congophilic nodular acellular material in the papillary dermis (Congo red stain, 100x).

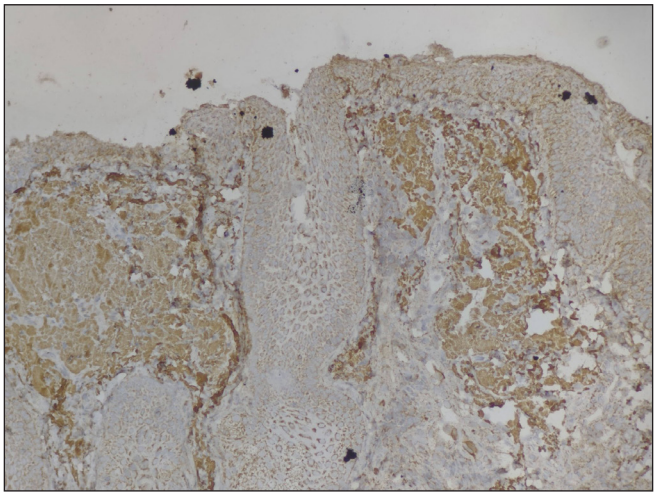

**Figure 4:** Serum-associated amyloid (SAA) highlights the acellular deposits in the papillary dermis (100x).

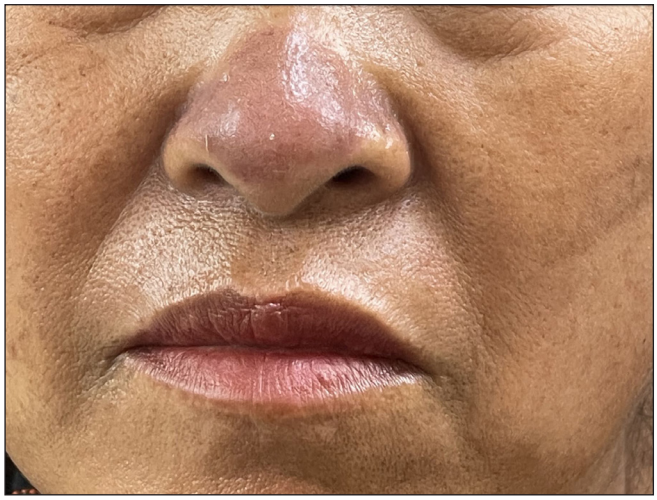

**Figure 5:** Regression of the hyperpigmentation with reduced pruritus after 2 weeks.

lesions showed improvement with remarkable regression of the hyperpigmentation and reduced pruritus [Figure 5].

Primary localised cutaneous amyloidosis is a disorder where amyloid deposits are seen in previously normal skin, with no evidence of deposits in internal organs. Macular amyloidosis, a common variant, has a female preponderance with age of onset ranging between 21 and 50 years. Clinically, macular amyloidosis presents as poorly delineated hyperpigmented lesions of greyish-brown macules with a rippled pattern, associated with deposition of amyloid material in the papillary dermis on histopathology.[4] The sites most commonly involved are the interscapular area and extremities (shins and forearms), while clavicles, breast, face, neck and axillae are rarely involved.

Friction or often repeated trauma by towels, nylon scrubbers and clothes has been implicated in the causation of macular amyloidosis, and is described as friction amyloidosis or nylon friction dermatitis.[5,6] Though the rippled and reticulate

pattern is the most common presentation, many unusual forms, like poikilodermatous, diffuse, bullous, nevoid, linear, amyloidosis cutis dyschromica and incontinentia pigmenti-like have been reported.[6]

We report this case for its unusual appearance as well as rare site of presentation.

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## References

1. Mehrotra K, Dewan R, Kumar JV, Dewan A. Primary cutaneous amyloidosis: A clinical, histopathological and immunofluorescence study. J Clin Diagn Res 2017;11:WC01–5.
2. Guillet C, Steinmann S, Maul JT, Kolm I. Primary localized cutaneous amyloidosis: A retrospective study of an uncommon skin disease in the largest tertiary care center in Switzerland. Dermatol 2022;238: 579–86.
3. Konmettu AP, Gurumurthy C, Rangappa V, Shastry V. Frictional melanosis and macular amyloidosis-exploring the link. Pigment Int 2022;9:166–75.
4. Bandhlish A, Aggarwal A, Koranne R. A clinico-epidemiological study of macular amyloidosis from North India. Indian J Dermatol 2012;57:269.
5. Venkataram M, Shashikant MBA. Frictional melanosis and its clinical and histopathological features. Iran J Dermatol 2018;21:124–7.
6. Somani VK, Somani A, Sarkar R. Primary localized cutaneous amyloidosis − A review. Pigment Int 2023;10:4–13.

# Ectodermal dysplasia-skin fragility syndrome – identification of a novel plakophilin1 (*PKP1*) gene variant through whole exome sequencing

Dear Editor,

A 16-year-old boy, presented with generalised skin erosions, dryness and cracking of lips, thickened skin on the palms and soles, abnormal nails, and woolly hair. The symptoms started at the age of 3 months with gradual progression. He was the first child of a second-degree consanguineous marriage, with his younger sister and both parents being asymptomatic. On physical examination, his height (147 cm) and weight (33 kg) were below the third centile for his age, with a normal gait. Cutaneous examination revealed generalised xerosis with features of skin fragility in the form of numerous superficial skin erosions with crusting on the upper back [Figure 1a], chest, and limbs, with the face being relatively spared. Palmoplantar keratoderma was present with superficial fissures [Figures 1b and 1c]. The scalp showed coarse woolly hair, without alopecia/hypotrichosis, but with remarkably easy pluckability [Figure 1d]. The eyebrows looked thick and dense, with the eyelashes being normal. The lips and perioral region were red, scaly, and fissured [Figure 1e]. Nails were dystrophic with subungual hyperkeratosis and distal curving. A general examination of all other systems, including the cardiovascular system, was normal.

A trichogram of plucked hair showed dystrophic anagen hair. Histopathology revealed widened intercellular spaces with a split in the sub-corneal layer [Figure 2a].

Scanning electron microscopy (SEM) of hair revealed damaged, rough cuticles with paint brush fractures of the cortex and dystrophic anagen roots [Figures 2b and 2c].

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