# A rare case of systemic amyloidosis presenting as palmar purpuric eczematous eruption

Dear Editor,

A 55-year-old man presented to the dermatology outpatient department with minimally itchy, red-coloured, bumpy lesions on his palms for 8 months. Examination revealed two morphologies. There were yellow to skin-coloured shiny translucent to waxy papules on the thenar eminence, palmar aspect of the distal and proximal interphalangeal and metacarpophalangeal joints. A few were ill-defined, purpuric, minimally scaly plaques on creases and thenar eminence of the right palm [Figures 1a and 1b]. He was previously diagnosed with palmar eczema and treated with topical steroids but showed only minimal improvement. An unusual morphology led us to examine the oral mucosa, which showed macroglossia with prominent tooth indentations, waxy infiltrated papules on the dorsum of the tongue, and multiple areas of purpura [Figure 1c]. There were multiple non tender boggy swellings on the occipital area of the scalp [Figure 1d]. The patient denied any weight loss, paraesthesia, light-headedness, syncope, hoarseness of voice or bony pains. The prominent macroglossia led us to consider primary systemic amyloidosis. We biopsied both tongue and palmar lesions, which showed amorphous eosinophilic deposits in the lamina propria, papillary and perivascular locations [Figure 2a]. Congo red stain showed khaki red to brown material on light microscopy, which demonstrated apple-green birefringence under polarised microscopy. Immunohistochemistry showed significant positivity positive for serum amyloid associated proteins [Figures 2b and 2c]. Serum for immunoglobulins IgM and IgG were elevated (1.2 and 17.9 g/dL respectively). Serum electrophoresis showed elevated free κ and λ chains (28.9g/L and 2164 g/L, respectively) with a reversal of the κ to λ ratio. Skeletal survey, M band electrophoresis, peripheral smear, serum calcium, and urine electrophoresis were normal. Bone marrow aspiration showed 17% plasma cells with λ

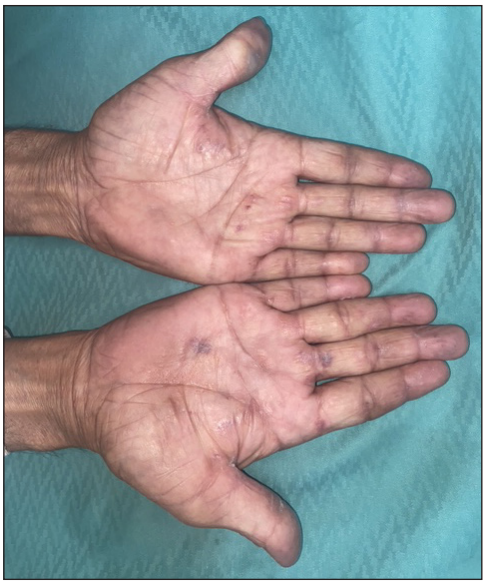

**Figure 1a:** Keratotic plaques and areas of purpura on bilateral palms.

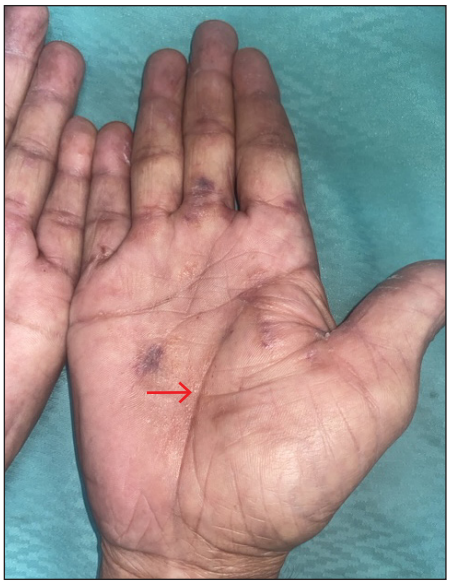

**Figure 1b:** Waxy papules and purpura along the creases of the right palm (indicated by the red arrow).

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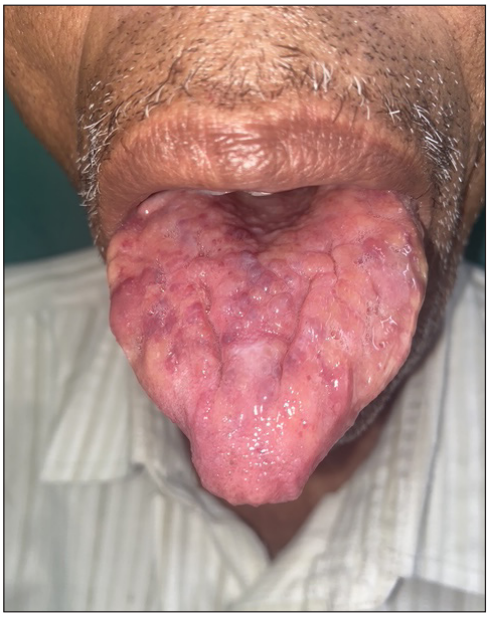

**Figure 1c:** Macroglossia with pebbly surface.

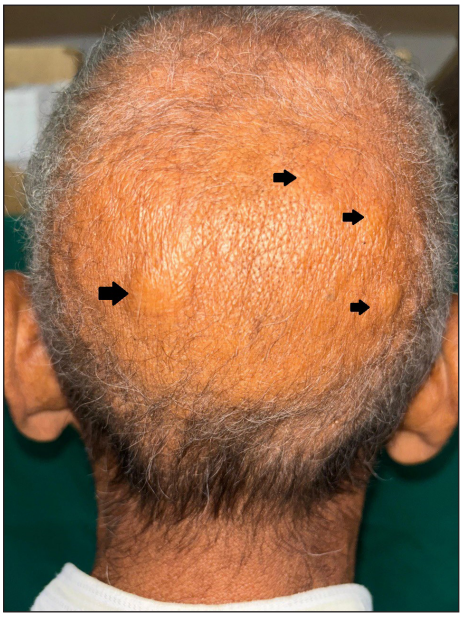

**Figure 1d:** Multiple soft boggy swelling over occiput (black arrows).

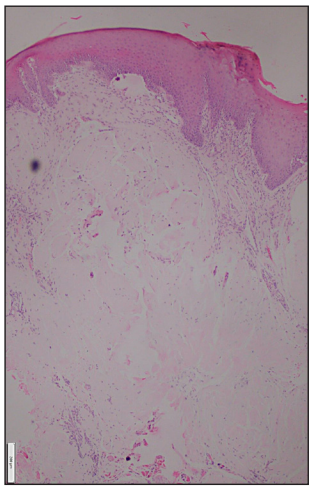

**Figure 2a:** Eosinophilic homogenous deposit in the dermal papilla (Haematoxylin & eosin, 50x).

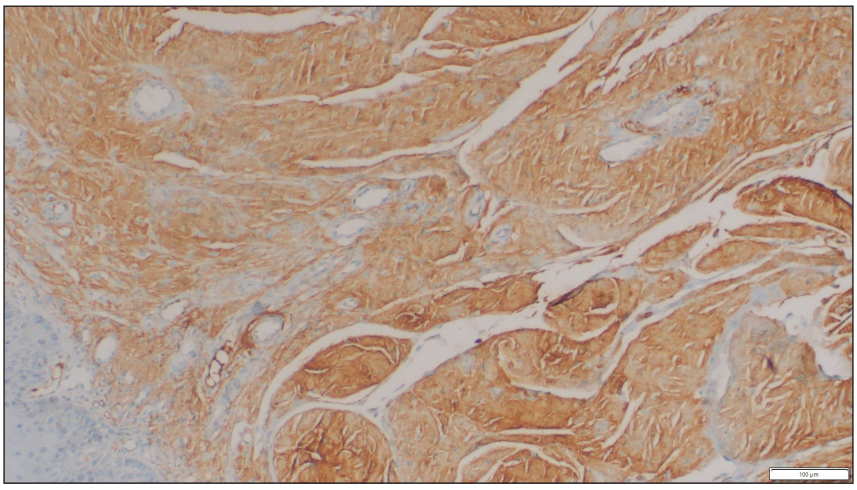

**Figure 2b:** Amyloid associated protein deposits on the dermal region of the tongue (Immunohistochemistry, 50x).

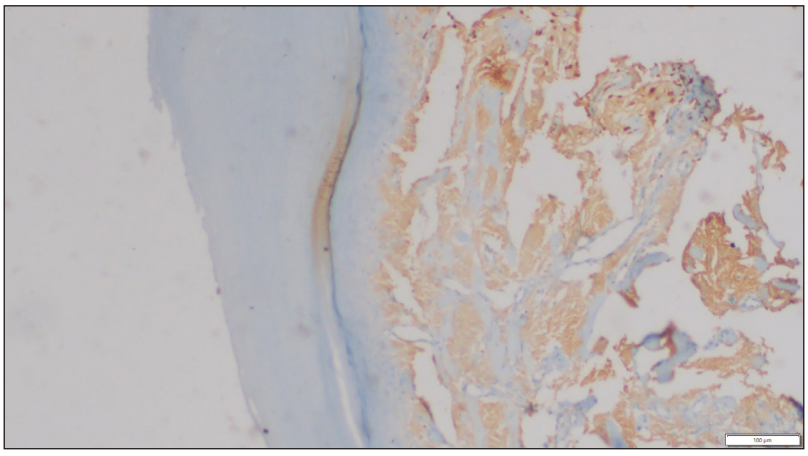

**Figure 2c:** Amyloid-associated protein deposits on the palm (Immunohistochemistry, 50x).

restriction on flow cytometry. The skeletal survey was normal. Primary systemic amyloidosis with multiple myeloma was diagnosed, and the patient was planned for chemotherapy with bortezomib, dexamethasone and lenalidomide.

Primary systemic amyloidosis is a rare disease resulting from skin and organ deposition of monoclonal light chains either due to multiple myeloma or Waldenstrom macroglobulinemia.[1] The complex interaction of these light chains with elastic tissue in the skin leads to dysfunction of the latter, which results in ecchymosis, easy bruising, cutis laxa, scleroderma, purpura, and waxy thickening (as in our case). Another significant pointer to the diagnosis is often macroglossia with scalloped margins over the lateral aspect of the tongue, as in our patient. Other cutaneous presentations of multiple myeloma include follicular hyperkeratotic spicules, diffuse alopecia, cutis verticis gyrata, nail dystrophy, and digital nodular swelling.[2,3] When faced with a case demonstrating purpuric and ecchymotic lesions on the palms, the differential diagnoses include papular purpuric gloves and socks syndrome, palmoplantar erythrodysesthesia secondary to chemotherapy, dermatitis herpetiformis, and trauma.[4,5] There are a few case reports of palmar purpura and nail fold waxy papules as manifestations of amyloidosis.[6,7] However, in these reports, easy bruising and purpura were seen at other sites as well.

We hereby want to highlight the atypical presentation of primary systemic amyloidosis as palmar purpura and encourage an oral mucosal examination, which can often clinch the diagnosis of systemic amyloidosis.

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***Anamika Choudhary[1], Shreya K Gowda[1], Ritu Sharma[1], Sudheer Arava[2], Kanika Sahni[1]***

Departments of [1]Dermatology and Venereology, and [2]Pathology, All India Institute of Medical Sciences, New Delhi, India

**Corresponding author:**
Dr. Kanika Sahni,
Department of Dermatology & Venereology,
All India Institute of Medical Sciences,
Delhi, India.
kanu.sah@gmail.com

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