Violaceous Papules on the Plantar Feet: An Unusual Presentation of a Nodular Amyloidosis

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Primary cutaneous amyloidosis refers to the deposition of amyloid within normal skin without evidence of internal organ involvement. Typically, it is characterized by hyperpigmented, asymptomatic, or pruritic patches (macular variant) or firm, skin-colored, or hyperpigmented papules and plaques (lichenoid variant). A third variant, nodular amyloidosis, is rare.

A 51-year-old man presented for evaluation of a 7-year history of an asymptomatic, nonprogressive violaceous papules and nodules involving the plantar feet (Figure 1A and 1B).

A skin biopsy of a nodule revealed a mild superficial dermal perivascular and

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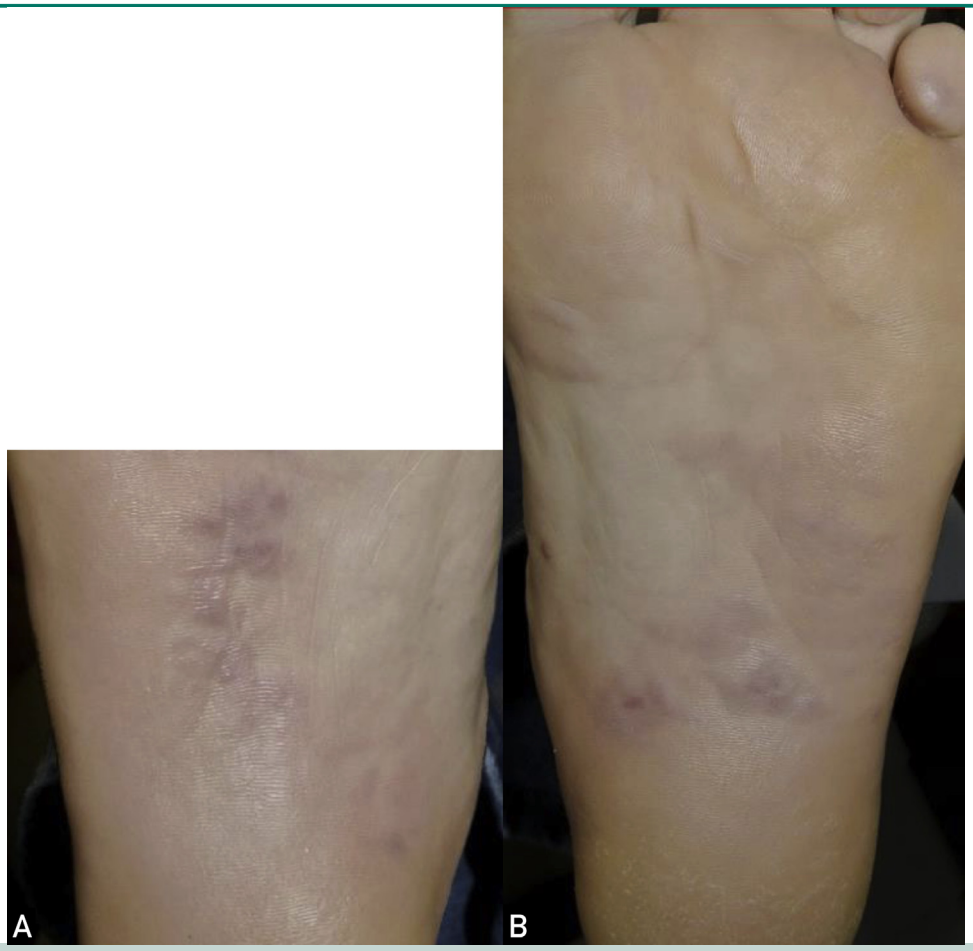

FIGURE 1. A and B, Multiple bilateral violaceous papules and nodules on the plantar feet.

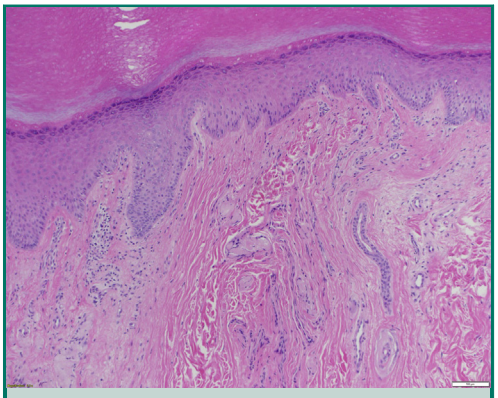

FIGURE 2. Mild superficial dermal perivascular and periadnexal lymphoplasmacytic inflammation with amorphous eosinophilic deposits in the dermis (hematoxylin and eosin, original magnification ×10).

periadnexal lymphoplasmacytic inflammation with Congo red–positive material involving the papillary dermis, periadnexal structures, and vessel walls (Figure 2; Supplemental Figure A and B, available online at http://www.mayoclinicproceedings.org).

Liquid chromatography tandem mass spectrometry revealed a peptide profile consistent with AL (lambda)–type amyloid deposition.

Further evaluation found no systemic involvement with amyloid. A bone marrow biopsy revealed a normocellular bone marrow without morphologic or immunophenotypic features of a plasma cell proliferative disorder. Subcutaneous fat aspirate from the abdomen was negative for amyloid. Serum and urine protein electrophoresis found no monoclonal protein.

Based on the clinical presentation, skin biopsy findings, and extensive evaluation revealing lack of systemic involvement, a final diagnosis of nodular localized primary cutaneous AL-lambda amyloidosis (NLPCA) was rendered.

Although lichenoid and macular (keratin-derived) amyloidosis are relatively common, NLPCA is the rarest form of primary cutaneous amyloidosis.

By definition, primary cutaneous amyloidosis is limited to the skin without internal organ involvement. However, the largest British series on NLPCA (15 patients) with the longest clinical follow-up period reported a 7% risk of progression to systemic amyloidosis.[1] The Mayo Clinic experience (16 patients) reported no evidence of progression in 14 patients over a 10-year follow-up period.[2]

In keeping with previous reports,[2] our patient presented with acral involvement. Nonetheless, the sole of the feet is a rare location for this condition. Because the histologic changes can be subtle, a high index of suspicion of this entity is needed to ensure an accurate diagnosis.

SUPPLEMENTAL ONLINE MATERIAL

Supplemental material can be found online at: http://www.mayoclinicproceedings.org. Supplemental material attached to journal articles has not been edited, and the authors take responsibility for the accuracy of all data.

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