

# Rippled Macules and Papules on the Legs

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A 34-year-old woman presented to our dermatology clinic with an intensely pruritic rash on the legs of 2 years' duration. The pruritus had waxed and waned in intensity, and the skin lesions were refractory to treatment with low-potency topical steroids. She had no other chronic medical conditions and was not taking any other medications.

## WHAT'S YOUR DIAGNOSIS?

- a. acanthosis nigricans
- b. confluent and reticulated papillomatosis
- c. cutaneous amyloidosis
- d. ichthyosis vulgaris
- e. keratosis pilaris

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The authors report no conflict of interest.

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doi:10.12788/cutis.0545

## THE DIAGNOSIS: Cutaneous Amyloidosis

A punch biopsy confirmed the diagnosis of cutaneous amyloidosis, which is characterized by the deposition of amyloid proteins in the skin without systemic involvement. Subtypes of cutaneous amyloidosis include lichenoid, macular, and nodular amyloidosis. A mixed or biphasic amyloidosis can occur when both lichenoid and macular lesions are present.<sup>1</sup> Lichenoid and macular amyloidosis generally are characterized by moderate to severe pruritus. Lichenoid amyloidosis favors the shins, calves, ankles, and extensor extremities; macular amyloidosis has a predilection for the interscapular area and less frequently the upper arms, chest, and thighs.<sup>2</sup> Atypical variants also have been reported, including amyloidosis cutis dyschromica, poikiloderm-like amyloidosis, and bullous amyloidosis, as well as incontinentia pigmenti–like, linear, and nevoid types.<sup>3</sup> Macular amyloidosis has been reported to occur in association with progressive systemic sclerosis, primary biliary cirrhosis, systemic lupus erythematosus, paronychia, and multiple endocrine neoplasia type 2.<sup>2</sup>

Acanthosis nigricans typically presents on the neck and intertriginous areas as velvety hyperpigmented plaques. Confluent and reticulated papillomatosis also appears as slightly elevated papules; however, it occurs

in the intermammary region in a reticulated pattern. Ichthyosis vulgaris also may occur on the lower extremities but presents with adherent large scales rather than papules. Keratosis pilaris may present on the proximal lower extremities with smaller, folliculocentric, flesh-colored to pink papules.

Treatment of cutaneous amyloidosis has long been challenging for dermatologists. The primary focus should be treatment of any underlying disease that is causing the pruritus and subsequent manipulation of skin lesions. Topical calcipotriol, phototherapy, oral cyclophosphamide, and Nd:YAG laser have demonstrated beneficial outcomes. IL-31 antibodies may be a potential future treatment.<sup>1</sup>

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