

Not Lichen This Rash

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Introduction:

- ❖ Skin rashes are amongst the most common conditions seen by primary care physicians (PCPs).
- ❖ It can be a formidable challenge to arrive at an accurate diagnosis because of the large number of dermatologic and systemic conditions that can manifest as a rash.
- ❖ The case below highlights the importance of PCPs in identifying red flag features of chronic rashes and making timely referral to Dermatology in order to ultimately diagnose cutaneous amyloidosis.

Case Presentation:

- ❖ A 73-year-old Hispanic man presented to his PCP office with 15-year history of recurrent episodes of intensely pruritic, hyperpigmented pustular rash on the anterior aspect of his legs.
- ❖ He had completed multiple courses of topical steroids of varying potency for undifferentiated dermatitis with only mild relief.
- ❖ On subsequent visits, the patient reported worsening pruritis affecting his sleep and noted progression of his rash to his back and arms.



Image 1 & 2: Lichenified, hyperkeratotic pigmented papules with excoriation

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DIAGNOSIS :  
A. SKIN, UPPER BACK, PUNCH:  
1. Changes consistent with lichen amyloidosis, see microscopic  
description.  
2. Small superficial dermal angioma.  
(Multiple additional levels obtained)  
B. SKIN, RIGHT ARM, PUNCH:  
Changes consistent with lichen amyloidosis, see microscopic  
description.
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Image 3: Skin biopsy results.

- ❖ He was referred to Dermatology and at that office visit he was noted to have lichenified, hyperkeratotic pigmented papules involving his shins, back, and sacral cleft with active excoriation
- ❖ Skin biopsy revealed patchy globular aggregates within the papillary dermis, superficial dermal perivascular chronic inflammation, hyperkeratosis and papillomatous epidermal acanthosis.
- ❖ Globular aggregates stained positive for Congo red, Crystal Violet and CK5/6 consistent with amyloid protein of epidermal origin. He was hence diagnosed with lichen amyloidosis and started on augmented betamethasone with significant improvement. Subsequently, he was maintained on oral bethamethasone three times weekly to prevent relapse. Fortunately, further clinical and laboratory evaluation showed no evidence of systemic amyloidosis.

Discussion:

- ❖ Lichen amyloidosis is the most common form of Primary Limited Cutaneous Amyloidosis (PLCA) which presents as multiple, intensely pruritic, discrete hyperpigmented papules that may coalesce into plaques.
- ❖ It is most common in males from South Asia, China, and South America in the 5th and 6th decade of life.
- ❖ It is characterized by deposition of amyloid derived from the degeneration of keratin filaments in the papillary dermis without any visceral involvement.
- ❖ It can be associated with MEN 2A syndrome or other autoimmune skin and systemic conditions. Fortunately, it does not progress to systemic amyloidosis.
- ❖ Treatment is centered on symptomatic and cosmetic relief. Therapeutic options include topical or intralesional steroids, antihistamines, UV-therapy, laser therapy, dermabrasion, scalpel scraping, retinoid agents and cyclophosphamide.

Conclusion:

- ❖ This case aptly demonstrates the importance of training PCPs to recognize red flag features while treating skin conditions such as chronic and progressive skin disease, no response or minimal response to standard treatment, uncertain diagnosis or systemic features.
- ❖ An expedited referral to Dermatology in such cases can improve quality of patient care by prompt diagnosis and initiation of appropriate treatment.

References:

- 1) <https://emedicine.medscape.com/article/1102672-overview>.
- 2) <https://medlineplus.gov/genetics/condition/primary-localized-cutaneous-amyloidosis/>
- 3) Ladizinski B, Lee KC. Lichen amyloidosis. *CMAJ*. 2014;186(7):532. doi:10.1503/cmaj.130698