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A man with diffuse hyperkeratotic papules and plaques

Harleen Arora

Henry Ford Health System, harora2@hfhs.org

Danielle Yeager

Henry Ford Health System, dyeager2@hfhs.org

Alison Tisack-Boucher

Henry Ford Health System, atisack1@hfhs.org

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Harleen Arora MD¹, Danielle Yeager MD¹, Allison Tisack-Boucher MD¹

¹ Department of Dermatology
Henry Ford Health System, Detroit, Michigan

History

- A 47-year-old African American man with no relevant past medical history presented to Dermatology with a dermatitis that was diagnosed at an outside clinic as hypertrophic lichen planus (LP).
- This dermatitis began 2-3 years prior with a sudden onset on the arms and knees and has since spread diffusely, including oral involvement.
- Denied any history of hepatitis C, syphilis, or HIV. No family history of similar condition

Examination

- Many hyperkeratotic scaly papules coalescing into plaques with underlying violaceous macules and patches on the arms, legs, buttocks, and hips
- Many papules and plaques in a linear distribution, suggesting koebnerization.
- Mouth with hyperkeratotic papules along the oral commissures, cobblestoning of the palate, and reticulated white streaks consistent with Wickham's striae on the upper and lower mucosal lip and the left buccal mucosa.

Differential Diagnosis

- Hypertrophic lichen planus (HLP)
- Keratosis lichenoides chronica (KLC)
- Diffuse verrucae

Laboratory

- CBC and CMP normal
- Hepatitis C, syphilis, and HIV negative

Histopathology

- Verrucous epidermal hyperplasia with overlying parakeratosis and occasional neutrophils in the stratum corneum.
- Lichenoid infiltrate of lymphocytes at dermal epidermal junction.
- Necrotic keratinocytes and melanin incontinence
- Staining for HPV RNA in-situ hybridization (ISH) 16/18, low risk cocktail, and high risk cocktail were all negative.

Clinical Photos



Figure 1

Hyperkeratotic scaly papules coalescing into plaques on a violaceous background on the arms, elbows, and abdomen



Figure 2

Linear distribution of hyperkeratotic papules indicative of koebnerization

Diagnosis

- Verruca was ruled out with negative HPV staining.
- KLC and HLP both are clinically similar and present with hyperkeratosis and lichenoid infiltrate with interface change histologically.
- KLC was favored due to chronicity of individual lesions and due to the confluent parakeratosis and corneal neutrophils seen on pathology which are uncharacteristic of HLP.

Course and Therapy

- At time of presentation, patient was on a regimen of acitretin 50 mg daily, betamethasone dipropionate ointment to the body, and tacrolimus 0.1% ointment to the face without significant improvement.
- Patient was continued on topical steroids and acitretin, and was also started on CellCept 500mg BID.

Discussion

- HLP and KLC can present very similarly with erythematous to violaceous hyperkeratotic papules and plaques. These are usually present on the lower extremities but may spread more diffusely, including trunk and face.
- **Keratosis Lichenoides Chronica (KLC)**
 - Rarer condition with a chronic course
 - May present at any age and has a slight male predominance
 - Etiology unknown; given its similarity to other keratotic skin diseases and its rarity, it has not been well-studied
 - Lesions can be pruritic and are typically in a reticular or linear pattern.
 - Oral and nail involvement in 20-30%
 - KLC also may present with facial eruption similar to seborrheic dermatitis or rosacea.
 - Difficult to treat
 - Oral retinoids and phototherapy have had some efficacy
 - Systemic corticosteroids, immunosuppressants, antimalarial medications, and antibiotics have been used with limited efficacy.
- **Hypertrophic Lichen Planus (HLP)**
 - Hyperkeratotic, pruritic papules and plaques
 - Average course of 6 years duration
 - Koebnerization leads to linear formations of plaques
 - May include oral involvement (Wickham striae) or nail changes
 - LP involves activation of T lymphocytes at the dermal-epidermal junction with apoptosis of basal keratinocytes.
 - Due to longstanding inflammation, hypertrophic LP may develop malignant transformation to squamous cell carcinomas or keratoacanthomas.
 - Treatment
 - Topical or intralesional corticosteroids, with or without cryotherapy
 - Acitretin and mycophenolate mofetil (CellCept) have shown efficacy in resistant hypertrophic LP.
 - Other treatment options for cutaneous LP include methotrexate, metronidazole, cyclosporine, dapsone, azathioprine, and phototherapy.

References

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