

Recurrent Arciform Plaque on the Face

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An otherwise healthy 31-year-old woman presented with a gradual growth of a semiannular, arciform, mildly pruritic plaque around the mouth of 10 years' duration that recurred biannually, persisted for a few months, and spontaneously remitted without residual scarring. She denied joint pain, muscle aches, sores in the mouth, personal or family history of autoimmune diseases, or other remarkable review of systems. Physical examination revealed a well-defined, edematous, smooth, arciform plaque on the face with no mucous membrane involvement. Laboratory evaluation, including complete blood cell count, comprehensive metabolic panel, and antinuclear antibody titer, was unremarkable. A punch biopsy was obtained.

WHAT'S YOUR DIAGNOSIS?

- erythema annulare centrifugum
- Jessner lymphocytic infiltration of the skin
- localized granuloma annulare
- lupus erythematosus tumidus
- reticular erythematous mucinosis

PLEASE TURN TO **PAGE E14** FOR THE DIAGNOSIS

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The authors report no conflict of interest.

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THE DIAGNOSIS: Lupus Erythematosus Tumidus

Histopathologic evaluation of a punch biopsy revealed focal dermal mucin deposition and CD123⁺ discrete clusters of plasmacytoid dendritic cells without interface changes (Figure 1), favoring a diagnosis of lupus erythematosus tumidus (LET) in our patient. There was no clinical improvement in symptoms when she previously was treated with topical antifungals or class III corticosteroid creams. Tacrolimus ointment 0.1% twice daily for 1 month did not result in substantial improvement in the appearance of the plaque, and it spontaneously resolved after 2 to 3 months. She declined treatment with hydroxychloroquine.

Lupus erythematosus tumidus is an uncommon subtype of chronic cutaneous lupus erythematosus with no distinct etiology. It is clinically characterized by edematous, urticarial, single or multiple plaques with a smooth surface affecting sun-exposed areas that can last for months to years.¹ In contrast to other variations of chronic cutaneous lupus such as discoid lupus erythematosus, LET lesions lack surface papulosquamous features such as scaling, atrophy, and follicular plugging.¹⁻⁴ Based solely on histologic findings, LET may be indistinguishable from reticular erythematous mucinosis and Jessner lymphocytic infiltration of the skin (JLIS) due to a similar lack of epidermal involvement and presence of a perivascular lymphocytic infiltrate (Figure 2).

The average age at disease onset is 36 years, nearly the same as that described in discoid lupus erythematosus.⁴ Lupus erythematosus tumidus has a favorable prognosis and commonly presents without other autoimmune signs, serologic abnormalities, or gender preference, with concomitant systemic lupus erythematosus sometimes reported.⁵

The absence of clinical and histological epidermal involvement are the most important clues to aid in the diagnosis. It has been postulated that JLIS could be an early cutaneous manifestation of LET.⁶ The differential diagnosis also may include erythema annulare centrifugum, granuloma annulare, and urticarial vasculitis. Lesions typically respond well to photoprotection, topical corticosteroids, and/or antimalarials. The addition of tacrolimus ointment 0.1% may result in complete regression without recurrence.⁷

Erythema annulare centrifugum is a reactive erythema that classically begins as a pink papule that gradually enlarges to form an annular erythematous plaque with a fine trailing scale that may recur.⁸ The histopathology of erythema annulare centrifugum shares features seen in LET, making the diagnosis difficult; however, secondary

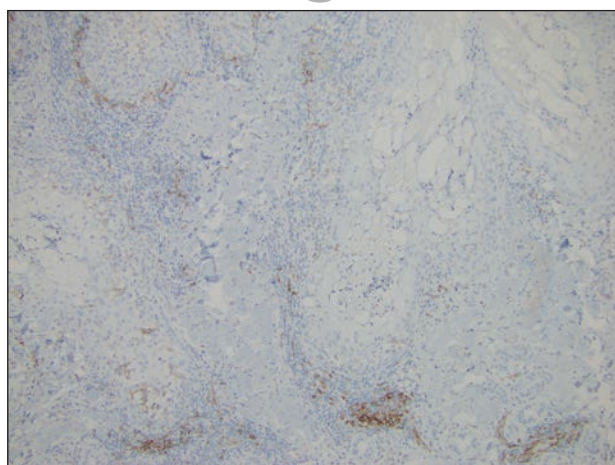


FIGURE 1. Colloidal iron staining demonstrated focal dermal mucin deposition (original magnification $\times 100$).

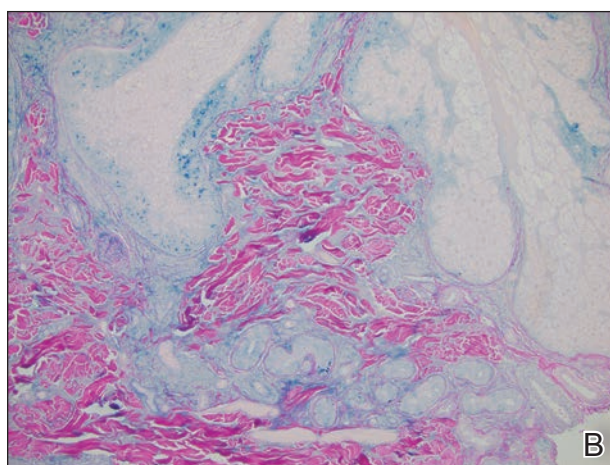
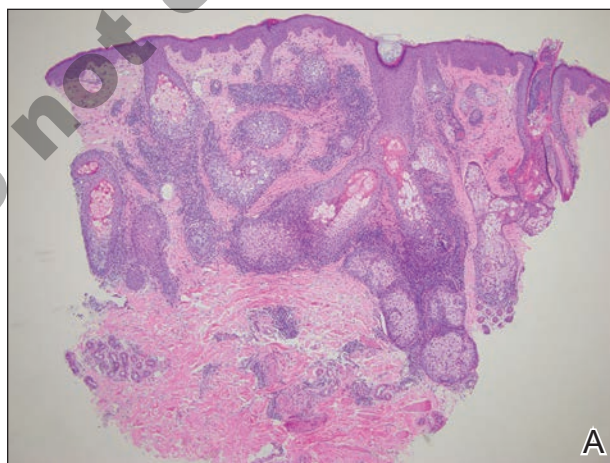


FIGURE 2. A and B, Punch biopsy specimen demonstrated a superficial and deep perivascular and periadnexal lymphocytic infiltrate (H&E, original magnifications $\times 40$ and $\times 100$).

changes to the epidermis (eg, spongiosis, hyperkeratosis) may be seen. This condition has been associated with lymphoproliferative malignancies.⁸

Reticular erythematous mucinosis is clinically distinguished from LET, as it presents as reticular, rather than arciform, erythematous macules, papules, or plaques that may be asymptomatic or pruritic.⁹ Histopathology typically shows more superficial mucin deposition than in LET as well as superficial to mid-dermal perivascular and periadnexal lymphocytic infiltrates. Reticular erythematous mucinosis more frequently is reported in women in their 30s and 40s and has been associated with UV exposure and hormonal triggers, such as oral contraceptive medications and pregnancy.⁹

Granuloma annulare typically presents as asymptomatic, erythematous, annular plaques or papules in young women.¹⁰ There are several histologic subtypes that show focal collagen degeneration, inflammation with palisaded or interstitial histiocytes, and mucin deposition, regardless of clinical presentation. Granuloma annulare has been associated with systemic diseases including type 2 diabetes mellitus and thyroid disease. Localized granuloma annulare most commonly presents on the dorsal aspects of the hands or feet.¹⁰

We present a case of LET on the face. Although histologically similar to other dermatoses, LET often lacks epidermal involvement and presents on sun-exposed areas of the body. Jessner lymphocytic infiltration of the skin also should be considered in the differential, as there is an overlap of clinical and histopathological features; JLIS lacks mucin deposits.⁶ This case reinforces the importance of correlating clinical with histopathologic findings. Our patient was treated with tacrolimus ointment 0.1%, and the plaque eventually resolved in 2 to 3 months without

recurrence. This condition should be included in the differential diagnosis of recurring annular plaques on sun-exposed areas, particularly in middle-aged adults, even in the absence of systemic involvement.

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