Annular Plaques Overlying Hyperpigmented Telangiectatic Patches on the Neck

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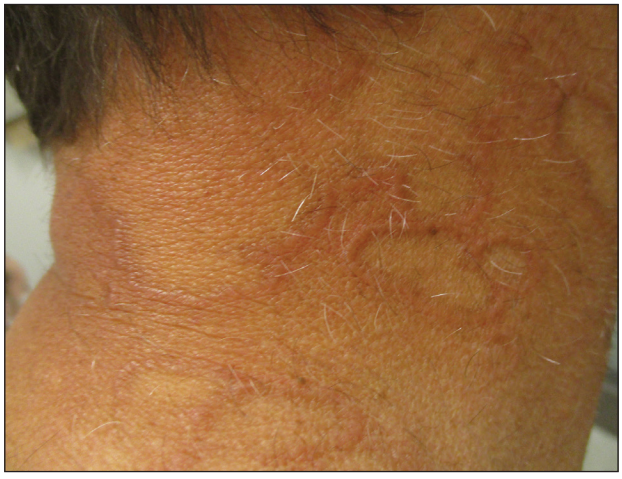

A 58-year-old man with a history of type 2 diabetes mellitus, nephrolithiasis, hypovitaminosis D, and hypercholesterolemia presented to our dermatology clinic for a follow-up total-body skin examination after a prior diagnosis of basal cell carcinoma on the vertex of the scalp. Physical examination revealed extensive photodamage and annular plaques overlying hyperpigmented telangiectatic patches on the dorsal portion of the neck. The eruption persisted for 1 year and failed to improve with clotrimazole cream. His medications included simvastatin, metformin, chlorthalidone, vitamin D, and tamsulosin. Two shave biopsies from the posterior neck were performed.

WHAT'S YOUR **DIAGNOSIS?**

a. annular elastolytic giant cell granuloma
b. granuloma annulare
c. necrobiosis lipoidica
d. secondary syphilis
e. tinea corporis

PLEASE TURN TO PAGE 78 FOR THE DIAGNOSIS

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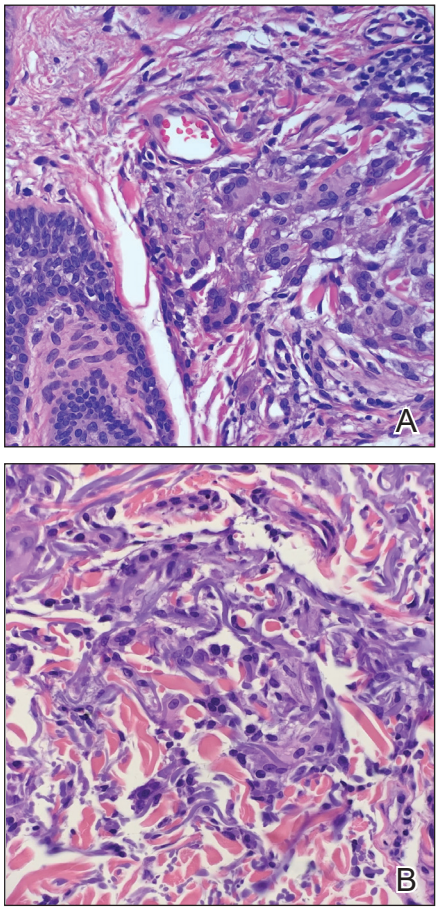
doi:10.12788/cutis.0707

THE **DIAGNOSIS:** Annular Elastolytic Giant Cell Granuloma

Histologic examination of the shave biopsies showed a granulomatous infiltrate of small lymphocytes, histiocytes, and multinucleated giant cells. The giant cells have abundant eosinophilic cytoplasm, with several also containing fragments of basophilic elastic fibers (elastophagocytosis)(Figure). Additionally, the granulomas revealed no signs of necrosis, making an infectious source unlikely, and examination under polarized light was negative for foreign material. These clinical and histologic findings were diagnostic for annular elastolytic giant cell granuloma (AEGCG).

Annular elastolytic giant cell granuloma is a rare chronic inflammatory disorder that classically presents on sun-exposed skin as annular plaques with elevated borders and atrophic centers.[1-4] Histologically, AEGCG is characterized by diffuse granulomatous infiltrates composed of multinucleated giant cells, histiocytes, and lymphocytes in the dermis, along with phagocytosis of elastic fibers by multinucleated giant cells.[5] The underlying etiology and pathogenesis of AEGCG remains unknown.[6]



A and B, A biopsy showed basophilic elastic fibers in the cytoplasm of the multinucleated giant cells in the dermis (H&E, original magnifications ×400). Images courtesy of Anne Bussian, MD (Birmingham, Alabama).

Annular elastolytic giant cell granuloma commonly affects females aged 35 to 75 years; however, cases have been reported in the male and pediatric patient populations.[1,2] Documented cases are known to last from 1 month to 10 years.[7,8] Although the mechanisms underlying the development of AEGCG remain to be elucidated, studies have determined that the skin disorder is associated with sarcoidosis, molluscum contagiosum, amyloidosis, diabetes mellitus, and cutaneous T-cell lymphoma.[9] Diabetes mellitus is the most common comorbidity associated with AEGCG, and it is theorized that diabetes contributes to the increased incidence of AEGCG in this population by inducing damage to elastic fibers in the skin.[10] One study that examined 50 cases of AEGCG found that 38 patients had serum glucose levels evaluated, with 8 cases being subsequently diagnosed with diabetes mellitus and 6 cases with apparent impaired glucose tolerance, indicating that 37% of the sample population with AEGCG who were evaluated for metabolic disease were found to have definitive or latent type 2 diabetes mellitus.[11] Although AEGCG is a rare disorder, a substantial number of patients diagnosed with AEGCG also have diabetes mellitus, making it important to consider screening all patients with AEGCG for diabetes given the ease and widely available resources to check glucose levels.

Actinic granuloma, granuloma annulare, atypical facial necrobiosis lipoidica, granuloma multiforme, secondary syphilis, tinea corporis, and erythema annulare centrifugum most commonly are included in the differential diagnosis with AEGCG; histopathology is the key determinant in discerning between these conditions.[12] Our patient presented with typical annular plaques overlying hyperpigmented telangiectatic patches. With known type 2 diabetes mellitus and the clinical findings, granuloma annulare, erythema annulare centrifugum, and AEGCG remained high on the differential.

No standard of care exists for AEGCG due to its rare nature and tendency to spontaneously resolve. The most common first-line treatment includes topical and intralesional steroids, topical pimecrolimus, and the use of sunscreen and other sun-protective measures. UV radiation, specifically UVA, has been determined to be a causal factor for AEGCG by changing the antigenicity of elastic fibers and producing an immune response in individuals with fair skin.[13] Further, resistant cases of AEGCG successfully have been treated with cyclosporine, systemic steroids, antimalarials, dapsone, and oral retinoids.[14,15]

Some studies reported partial regression or full resolution with topical tretinoin; adalimumab; clobetasol ointment; or a combination of corticosteroids, antihistamines, and hydroxychloroquine.[2] Only 1 case series using sulfasalazine reported worsening symptoms after treatment initiation.[16]

Our patient deferred systemic medications and was treated with 4 weeks of topical triamcinolone followed by 4 weeks of topical tacrolimus with minimal improvement. At the time of diagnosis, our patient also was encouraged to use sun-protective measures. At 6-month follow-up, the lesions remained stable, and the decision was made to continue with photoprotection.

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