

Annular Erythematous Plaques With Central Hypopigmentation on Sun-Exposed Skin

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A 67-year-old White woman presented to our dermatology clinic with pruritic annular erythematous plaques with central hypopigmentation on the forearms, dorsal aspect of the hands, neck, and fingers of 3 to 4 months' duration. The patient rated the severity of pruritus an 8 on a 10-point scale. A review of symptoms was positive for fatigue, joint pain, and headache. The patient had a history of type 2 diabetes mellitus, osteoarthritis, thyroid disease, and stage 3 renal failure. A punch biopsy from the left forearm was performed.

WHAT'S YOUR DIAGNOSIS?

- a. annular elastolytic granuloma
- b. annular lichen planus
- c. erythema annulare centrifugum
- d. necrobiosis lipoidica
- e. tinea corporis

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The authors report no conflict of interest.

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THE DIAGNOSIS:

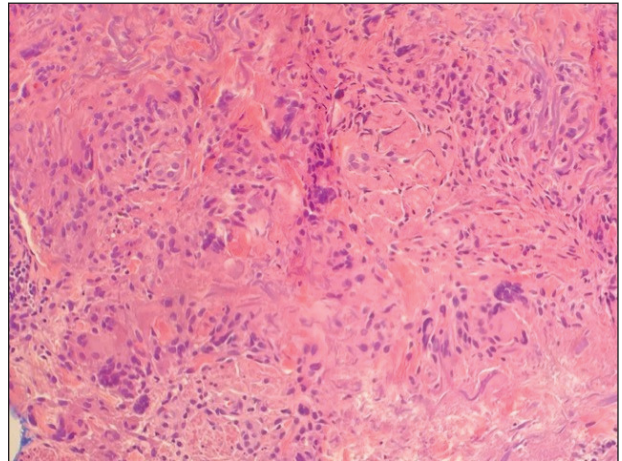
Annular Elastolytic Granuloma

A biopsy showed a markedly elastotic dermis consisting of a palisading granulomatous inflammatory infiltrate and numerous multinucleated histiocytes (Figure). These histopathologic findings along with the clinical presentation confirmed a diagnosis of annular elastolytic granuloma (AEG). Treatment consisting of 3 months of oral minocycline, 2 months of oral doxycycline, and clobetasol ointment all failed. At that point, oral hydroxychloroquine was recommended. Our patient was lost to follow-up by dermatology, then subsequently was placed on hydroxychloroquine by rheumatology to treat both the osteoarthritis and AEG. A follow-up appointment with dermatology was planned for 3 months to monitor hydroxychloroquine treatment and monitor treatment progress; however, she did not follow-up or seek further treatment.

Annular elastolytic granuloma clinically is similar to granuloma annulare (GA), with both presenting as annular plaques surrounded by an elevated border.¹ Although AEG clinically is distinct with hypopigmented atrophied plaque centers,² a biopsy is required to confirm the lack of elastic tissue in zones of atrophy and the presence of multinucleated histiocytes.^{1,3} Lesions most commonly are seen clinically on sun-exposed areas in middle-aged White women; however, they rarely have been seen on frequently covered skin.⁴ Our case illustrates the striking photodistribution of AEG, especially on the posterior neck area. The clinical diagnoses of AEG, annular elastolytic giant cell granuloma, and GA in sun-exposed areas are synonymous and can be used interchangeably.^{5,6}

Pathologies considered in the diagnosis of AEG include but are not limited to tinea corporis, annular lichen planus, erythema annulare centrifugum, and necrobiosis lipidica. Scaling typically is absent in AEG, while tinea corporis presents with hyphae within the stratum corneum of the plaques.⁷ Papules along the periphery of annular lesions are more typical of annular lichen planus than AEG, and they tend to have a more purple hue.⁸ Erythema annulare centrifugum has annular erythematous plaques similar to those found in AEG but differs with scaling on the inner margins of these plaques. Histopathology presenting with a lymphocytic infiltrate surrounding vasculature and no indication of elastolytic degradation would further indicate a diagnosis of erythema annulare centrifugum.⁹ Histopathology showing necrobiosis, lipid depositions, and vascular wall thickenings is indicative of necrobiosis lipidica.¹⁰

Similar to GA,¹¹ the cause of AEG is idiopathic.² Annular elastolytic granuloma and GA differ in the fact that elastin degradation is characteristic of AEG compared to collagen degradation in GA. It is suspected



Histopathology revealed a palisading granulomatous inflammatory infiltrate and numerous multinucleated histiocytes (H&E, original magnification $\times 100$).

that elastin degradation in AEG patients is caused by an immune response triggering phagocytosis of elastin by multinucleated histiocytes.² Actinic damage also is considered a possible cause of elastin fiber degradation in AEG.¹² Granuloma annulare can be ruled out and the diagnosis of AEG confirmed with the absence of elastin fibers and mucin on pathology.¹³

Although there is no established first-line treatment of AEG, successful treatment has been achieved with antimalarial drugs paired with topical steroids.¹⁴ Treatment recommendations for AEG include minocycline, chloroquine, hydroxychloroquine, tranilast, and oral retinoids, as well as oral and topical steroids. In clinical cases where AEG occurs in the setting of a chronic disease such as diabetes mellitus, vascular occlusion, arthritis, or hypertension, treatment of underlying disease has been shown to resolve AEG symptoms.¹⁴

Although light therapy is not common for AEG, UV light radiation has demonstrated success in treating AEG.^{15,16} One study showed complete clearance of granulomatous papules after narrowband UVB treatment.¹⁵ Another study showed that 2 patients treated with psoralen plus UVA therapy reached complete clearance of AEG lasting at least 3 months after treatment.¹⁶

REFERENCES

- Lai JH, Murray SJ, Walsh NM. Evolution of granuloma annulare to mid-dermal elastolysis: report of a case and review of the literature. *J Cutan Pathol*. 2014;41:462-468. doi:10.1111/cup.12292
- Klemke CD, Siebold D, Dippel E, et al. Generalised annular elastolytic giant cell granuloma. *Dermatology*. 2003;207:420-422. doi:10.1159/000074132

3. Limas C. The spectrum of primary cutaneous elastolytic granulomas and their distinction from granuloma annulare: a clinicopathological analysis. *Histopathology*. 2004;44:277-282. doi:10.1111/j.0309-0167.2004.01755.x
4. Revenga F, Rovira I, Pimentel J, et al. Annular elastolytic giant cell granuloma—actinic granuloma? *Clin Exp Dermatol*. 1996;21:51-53.
5. Hawryluk EB, Izikson L, English JC 3rd. Non-infectious granulomatous diseases of the skin and their associated systemic diseases: an evidence-based update to important clinical questions. *Am J Clin Dermatol*. 2010;11:171-181. doi:10.2165/11530080-000000000-00000
6. Berliner JG, Haemel A, LeBoit PE, et al. The sarcoid variant of annular elastolytic granuloma. *J Cutan Pathol*. 2013;40:918-920. doi:10.1111/cup.12237
7. Pflederer RT, Ahmed S, Tonkovic-Capin V, et al. Annular polycyclic plaques on the chest and upper back [published online April 24, 2018]. *JAAD Case Rep*. 2018;4:405-407. doi:10.1016/j.jdc.2017.07.022
8. Traves KP, Savage K, Studdiford JS. Annular lesions: diagnosis and treatment. *Am Fam Physician*. 2018;98:283-291.
9. Weyers W, Diaz-Cascajo C, Weyers I. Erythema annulare centrifugum: results of a clinicopathologic study of 73 patients. *Am J Dermatopathol*. 2003;25:451-462. doi:10.1097/00000372-200312000-00001
10. Dowling GB, Jones EW. Atypical (annular) necrobiosis lipidica of the face and scalp. a report of the clinical and histological features of 7 cases. *Dermatologica*. 1967;135:11-26. doi:10.1159/000254156
11. Piette EW, Rosenbach M. Granuloma annulare: pathogenesis, disease associations and triggers, and therapeutic options. *J Am Acad Dermatol*. 2016;75:467-479. doi:10.1016/j.jaad.2015.03.055
12. O'Brien JP, Regan W. Actinically degenerate elastic tissue is the likely antigenic basis of actinic granuloma of the skin and of temporal arteritis [published correction appears in *J Am Acad Dermatol*. 2000; 42(1 pt 1):148]. *J Am Acad Dermatol*. 1999;40(2 pt 1):214-222. doi:10.1016/s0190-9622(99)70191-x
13. Rencic A, Nousari CH. Other rheumatologic diseases. In: Bologna JL, Jorizzo JL, Rapini RP, et al, eds. *Dermatology*. 2nd ed. Elsevier Limited; 2008:600-601.
14. Burlando M, Herzum A, Cozzani E, et al. Can methotrexate be a successful treatment for unresponsive generalized annular elastolytic giant cell granuloma? case report and review of the literature. *Dermatol Ther*. 2021;34:E14705. doi:10.1111/dth.14705
15. Takata T, Ikeda M, Kodama H, et al. Regression of papular elastolytic giant cell granuloma using narrow-band UVB irradiation. *Dermatology*. 2006;212:77-79. doi:10.1159/000089028
16. Pérez-Pérez L, García-Gavín J, Allegue F, et al. Successful treatment of generalized elastolytic giant cell granuloma with psoralen-ultraviolet A. *Photodermatol Photoimmunol Photomed*. 2012;28:264-266. doi:10.1111/j.1600-0781.2012.00680.x