

Single Case

A Case of Annular Elastolytic Giant Cell Granuloma Associated with Syphilis

Xiaoping Liu^a Wei Zhang^b Yeqiang Liu^c Mingyuan Xu^c

^aDepartment of Mycology, Shanghai Skin Disease Hospital, Shanghai, China;

^bDepartment of Cosmetology, Shanghai Skin Disease Hospital, Shanghai, China;

^cDepartment of Dermatopathology, Shanghai Skin Disease Hospital, Shanghai, China

Keywords

Annular elastolytic giant cell granuloma · Syphilis · Elastophagocytosis

Abstract

Annular elastolytic giant cell granuloma (AEGCG) is a rare granulomatous skin disease characterized by annular patches with slightly raised borders, hypopigmented and/or atrophic centers found mainly on sun-exposed skin. Histologically, it is characterized by phagocytosis of elastic fibers by multinucleated giant cells. The pathogenesis of the disease is unclear. We report a case of 55-year-old man with AEGCG in association with syphilis, whose condition improved when hydroxychloroquine sulfate and topical tacrolimus were administered over a 2-month period.

© 2018 The Author(s)
Published by S. Karger AG, Basel

Introduction

Annular elastolytic giant cell granuloma (AEGCG) is a rare dermatosis characterized clinically by annular plaques with raised erythematous borders and histologically by granulomatous inflammation accompanied by loss of elastic tissue and elastophagocytosis. AEGCG has been reported in association with several skin disorders. We report a case of AEGCG accompanied by syphilis.

Case Report

A 55-year-old man presented with mild itchy erythematous papules on the forehead that had gradually increased in number and size for 1 year. The red papules first appeared on the forehead with 2–3 mm in size and then spread to the scalp. The diameter of the largest plaque enlarged to 6 cm in half a year and was well circumscribed with a slightly elevated border and central atrophy. He reported that he did not receive any topical or oral medications. He had a history of drug abuse and received 6 months' mandatory rehab. He was referred for a serologic reactive rapid plasma reagin test, which was positive at a titer of 1:128 (normal value: <1:2). HIV antibody test results were negative. Syphilis was diagnosed and treated with intramuscular penicillin G benzathine after skin biopsy. The patient received a weekly dose of penicillin G benzathine for 3 weeks. After 3 weeks of treatment, the titer dropped to 1:32. Dermatological examination revealed multiple erythematous plaques with raised borders and atrophic centers spread over the forehead and scalp (Fig. 1a). There were no other skin or mucosal lesions. A biopsy was performed with a clinical suspicion of discoid lupus erythematosus. Histopathological examination revealed dermal infiltrate with histiocytes and multinucleated giant cells (Fig. 2a); PAS staining revealed no fungal organisms (Fig. 2b). Van Gieson special staining revealed loss of elastic fibers in the center of the lesion (Fig. 2c). Based on these findings, a diagnosis of AEGCG was made and the patient was treated with oral administration of hydroxychloroquine sulfate 200 mg once daily and topical application of tacrolimus. Within 2 months, the erythema ceased and the elevated lesions flattened (Fig. 1b, c).

Discussion

AEGCG is an entity that was proposed originally by Hanke [1] and is characterized by elastolysis, elastophagocytosis, and an infiltrate of multinucleated giant cells. The pathogenesis of AEGCG remains unclear. It has been suggested that solar radiation, heat, or other unknown factors transform the antigenicity of the elastic fibers and induce the cellular immunological reaction [2]. This theory is supported by immunohistochemical findings that the inflammatory infiltrates are T lymphocytes and predominantly of CD 4+ cells [3]. The main clinical differential diagnosis includes actinic granuloma, granuloma annulare, atypical facial necrobiosis lipoidica, and granuloma multiforme. Histopathology is the key factor in differential diagnosis.

AEGCG follows a chronic course; however, there have been reports of spontaneous remission [4]. Therapy options include topical or intralesional glucocorticoids, clofazimine, cyclosporine, topical calcineurin inhibitors, hydroxychloroquine, methotrexate, psoralen plus ultraviolet A therapy, and narrowband ultraviolet B therapy [5]. In our case, the patient was treated with hydroxychloroquine sulfate at a daily dose of 200 mg and topical tacrolimus. The patient showed significant improvement after a treatment period of 2 months.

AEGCG has been associated with diabetes mellitus, systemic sarcoidosis, cutaneous amyloidosis, molluscum contagiosum, and cutaneous T-cell lymphoma [3]. In our case, the patient had associated syphilis. It is possible that this association was a coincidence. Syphilis is a sexually transmitted infection transmitted by *Treponema pallidum* subspecies *pallidum*, and several dermatological disorders are reported that are accompanied by syphilis [6, 7]. Chung et al. [8] found that *T. pallidum* can stimulate host human fibroblasts to increase the synthesis of matrix metalloproteinase-1 (MMP-1), which may act as a virulence factor of the organism. The pathogenic mechanism of *T. pallidum* was similar to ultraviolet irradiation in AEGCG, which

induces the formation of reactive oxygen species in the skin. Human dermal fibroblasts exposed to reactive oxygen species show increased expression of mRNA for MMP-1 and MMP-2, which have the ability to degrade collagen and elastic fibers [9]. In addition, elastophagocytosis as the characteristic feature of AEGCG has been associated with numerous infections including leprosy, bacterial folliculitis, and granulomatous syphilis [10]. It may be postulated that syphilis infection may be involved in the pathogenesis of AEGCG. Therefore, the role of syphilis infection in AEGCG should be clarified in the future.

Statement of Ethics

Patient consent for publication was obtained.

Disclosure Statement

The authors have no conflicts of interest to disclose.

References

- 1 Hanke CW, Bailin PL, Roenigk HH Jr. Annular elastolytic giant cell granuloma. A clinicopathologic study of five cases and a review of similar entities. *J Am Acad Dermatol*. 1979 Nov;1(5):413–21.
- 2 Klemke CD, Siebold D, Dippel E, Hildenbrand R, Bleyl U, Goerdts S. Generalised annular elastolytic giant cell granuloma. *Dermatology*. 2003;207(4):420–2.
- 3 Doulaveri G, Tsagrioni E, Giannadaki M, Boseberg E, Limas C, Potouridou I et al. Annular elastolytic giant cell granuloma in a 70-year-old woman. *Int J Dermatol*. 2003 Apr;42(4):290–1.
- 4 Hermes B, Haas N, Czarnetzki BM. [Annular elastolytic giant cell granuloma with a spontaneous healing tendency]. *Hautarzt*. 1995 Jul;46(7):490–3.
- 5 Nanbu A, Sugiura K, Kono M, Muro Y, Akiyama M. Annular Elastolytic Giant Cell Granuloma Successfully Treated with Minocycline Hydrochloride. *Acta Derm Venereol*. 2015 Jul;95(6):756–7.
- 6 Pinto-Almeida T, Rosmaninho A, Sanches M, Alves R, Selores M. Secondary syphilis on a psoriatic patient under cyclosporine: a challenging case. *Int J Dermatol*. 2014 Jan;53(1):e35–8.
- 7 Di Biagio A, Taramasso L, Gustinetti G, Riccardi N, Viscoli C. Onychomadesis in a male patient with secondary syphilis. *Int J STD AIDS*. 2016 Jul;27(8):704–5.
- 8 Chung KY, Kim KS, Lee MG, Chang NS, Lee JB. *Treponema pallidum* induces up-regulation of interstitial collagenase in human dermal fibroblasts. *Acta Derm Venereol*. 2002;82(3):174–8.
- 9 Watabe D, Akasaka T. Annular elastolytic giant cell granuloma developing on lesions of vitiligo. *Int J Dermatol*. 2013 Nov;52(11):1458–60.
- 10 El-Khoury J, Kurban M, Abbas O. Elastophagocytosis: underlying mechanisms and associated cutaneous entities. *J Am Acad Dermatol*. 2014 May;70(5):934–44.

Xiaoping Liu and Wei Zhang contributed equally to this work.

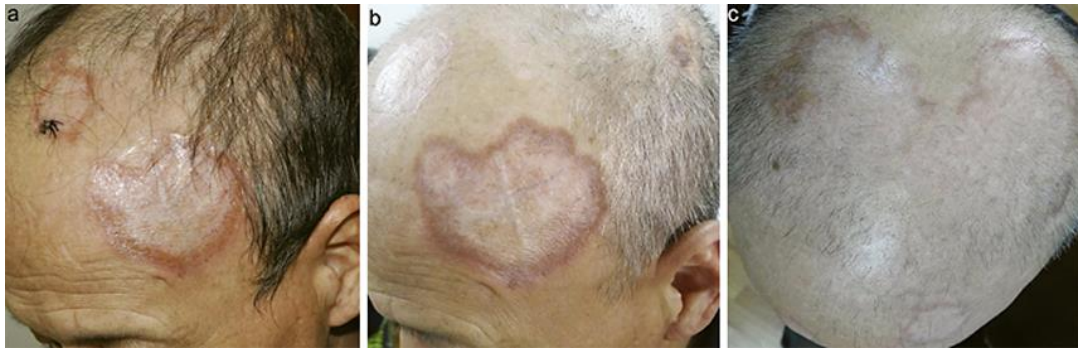


Fig. 1. **a** Multiple erythematous plaques with raised borders and atrophic centers distributed over the forehead and scalp. **b, c** Clinical presentation after a period of 2 months.

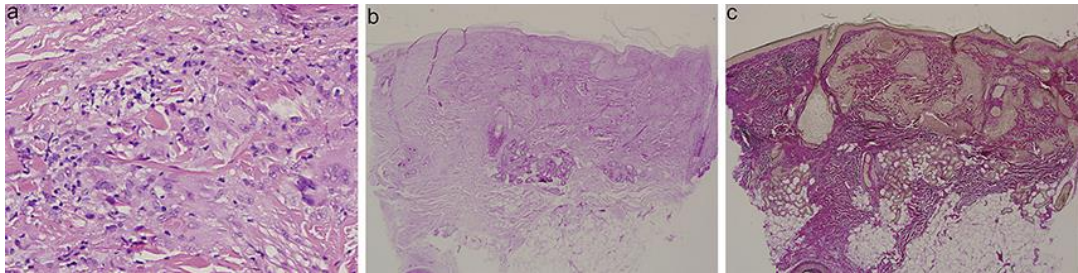


Fig. 2. **a** Dermal infiltrate made up of histiocytes and multinucleated giant cells. **b** PAS staining revealed no fungal organisms. **c** Verhoeff staining showing the absence of elastic fibers in the region of the granuloma.