Title
Photodistributed granuloma annulare

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Abstract

Annular elastolytic giant cell granuloma (AEGCG) is a controversial entity that is considered by many to be a variant of granuloma annulare (GA). The majority of cases of AEGCG occur in Caucasian women (3:2) between the ages of 40 and 70, with the distribution of the mostly annular lesions favoring exposed areas of skin and rarely involving covered skin. The most common systemic association has been with diabetes mellitus. We present a 52-year-old woman with an asymptomatic, annular, erythematous, photodistributed eruption of two-years duration. As part of her evaluation, it was detected that she had a hemoglobin A1C of 10.3% and a diagnosis of diabetes mellitus was made. We review the literature on the clinical and histopathologic features of GA and AEGCG and the overlap between these entities.

Case synopsis

History: A 52-year-old woman presented to the Skin and Cancer Unit for evaluation of an asymptomatic eruption of two-years duration. Around the time of experiencing stress, she first developed erythematous papules on her right forearm and the dorsal aspect of the right hand that persisted despite treatment with a topical glucocorticoid. Several months prior to presentation, after another stressful life event, the eruption appeared on her dorsal aspect of the left hand and forearm and later spread to her upper chest. The eruption remained asymptomatic. At the time of presentation, she denied recent sun exposure, although she reported sun exposure as a child. She noted many sunburns and inability to tan. The patient denied fever, chills, nausea, vomiting, and diarrhea. She reported recent weight gain.

Physical examination: On the dorsal aspects of the hands, forearms, and upper chest were annular, erythematous, thin papules and plaques.

Laboratory data: A complete blood count, anti-nuclear antibody, anti-Ro antibody, anti-La antibody, and serum protein electrophoresis were normal or negative. Comprehensive metabolic panel showed glucose of 216 mg/dL and aspartate transaminase and alanine transaminase of at 110 and 108 U/L, respectively. Hemoglobin A1C was at 10.3%.

Histopathology: There is a nodular infiltrate of histiocytes, some of which are multinucleated, that surround foci of degenerated collagen and increased connective tissue mucin, as demonstrated by a colloidal-iron stain.
Discussion

Diagnosis: Photodistributed granuloma annulare

Comment: Granuloma annulare (GA) is a benign, granulomatous dermatosis of unknown etiology, which most commonly has a localized distribution. Cases associated with photodistribution are rare and may overlap with annular elastolytic giant cell granuloma (AEGCG). AEGCG is a controversial entity, which was described as a granulomatous dermatologic condition with clinical similarities to GA [1,2]. It is characterized clinically by papules that evolve into annular and serpiginous plaques with elevated borders and histologically by elastolysis along with elastophagocytosis by multinucleated giant cells. Papular and reticular variants have been described [1]. The majority of cases of AEGCG occur in Caucasian women (3:2) between the ages of 40 and 70, with the distribution of the mostly annular lesions favoring exposed areas of skin and rarely involving covered skin. The most common systemic association has been diabetes mellitus [2].

Specific histopathologic findings of AEGCG include a prominent degree of elastolysis and elastophagocytosis by multinucleated giant cells and the lack of necrobiosis or excessive deposition of mucin [2]. It was originally noted that these histopathologic changes were present in annular lesions in sun-damaged skin, and the term actinic granuloma (AG) was suggested for lesions with
a distribution that correspond to sun-exposed skin and to indicate the pathogenic role of solar radiation [3]. However, investigators challenged the notion of AG as a distinct entity from GA, especially because of case reports of AG-like lesions in sun-protected areas. The term annular elastolytic giant cell granuloma (AEGCG) was initially introduced to specifically describe annular lesions with a major granulomatous elastolytic component, which could occur anywhere in the skin, including actinic sites [4].

In a review of the clinicopathologic data of 20 patients whose biopsies demonstrated the histopathologic features of elastolytic granuloma, as well as previously published cases with identical tissue changes, it was determined that the tissue changes of AG and AEGCG are identical and differ from GA because of predominant elastolysis and elastophagocytosis in the absence of necrobiosis and palisading granuloma [2]. However, elastolysis and elastophagocytosis occur in several diseases, which include generalized GA and cutaneous lymphoma and may be incidental to the activation of histiocytes [5]. Authors have attempted to distinguish AEGCG by the distinct zone of elastophagocytosis that is observed and the differing quantity and distribution of giant cells as compared to GA [6]. Yet, biopsies with mixed histopathologic features of GA and AEGCG have been described and attempts to distinguish these lesions by immunohistological methods have not yielded a consensus as many still consider AG and AEGCG to be variants of GA [7].

Case reports have described successful treatment of AEGCG with topical, intralesional, and systemic glucocorticoids; clofazimine; cryotherapy; dapsone; cyclosporine; methotrexate; narrowband ultraviolet B phototherapy; retinoids; fumaric acid esters; antimalarials; topical calcineurin inhibitors; and minocycline hydrochloride, although spontaneous regression also has been reported [11, 12].

References