Annular elastolytic giant cell granuloma in association with Hashimoto's thyroiditis

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ABSTRACT

Annular elastolytic giant cell granuloma (AEGCG) is a rare granulomatous skin disease characterized clinically by annular plaques with elevated borders and atrophic centers found mainly on sun-exposed skin and histologically 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. We report a case of AEGCG in a 50-year-old woman and is highlighted for the classical clinical and histological findings of the disease and its rare co-existence with Hashimoto's thyroiditis.

Key words: Annular elastolytic giant cell granuloma, Hashimoto's thyroiditis, mid-dermal elastolysis

INTRODUCTION

Annular elastolytic giant cell granuloma (AEGCG), is a rare granulomatous disorder characterized by annular plaques with elevated borders and central atrophy with a tendency to occur over sun exposed areas, rarely involving covered areas.[1] AEGCG falls under noninfectious granulomatous diseases of the skin, in which the granulomatous reactions represent immune reactions to an inciting antigen and may be associated with systemic disease.[2] A case of AEGCG associated with Hashimoto's thyroiditis (HT), in a 50-year-old female patient is reported for its rare co-occurrence.

CASE REPORT

A 50-year-old, nondiabetic, nonhypertensive female patient presented with multiple asymptomatic raised skin lesions involving back, upper limbs, and legs for two years. She also had diffuse alopecia, dryness of skin, and thyroid swelling of four years duration. There was no history of photosensitivity, oral ulcers, arthralgia, or drug intake. Personal history and family history were not contributory.

Dermatological examination revealed multiple, skin-colored and erythematous, annular, arciform, and polycyclic plaques of varying sizes involving the back, extensor aspects of forearms, dorsa of both hands, and anterolateral aspects of both legs [Figures 1a and 2]. The goiter was diffuse, firm in consistency, with a bosselated surface and was not fixed to underlying structures [Figure 3]. There was no mucous membrane or nail involvement. Systemic examination did not reveal any abnormality.

Routine hematological and biochemical investigations, including blood sugar levels, were within normal limits. However, she had raised TSH levels (8 μ/L), with normal T3 (8 pmol/L) and T4 (2.5 pmol/L) levels. Thyroid peroxidase antibody was positive. Chest radiograph and abdominal ultrasonography did not reveal any abnormality. Antinuclear antibody test, Mantoux test, VDRL, and KOH examination for fungus were negative.

Fine-needle aspiration cytology (FNAC) of the thyroid swelling showed follicular epithelial cells in sheets and clusters, with Hurthle

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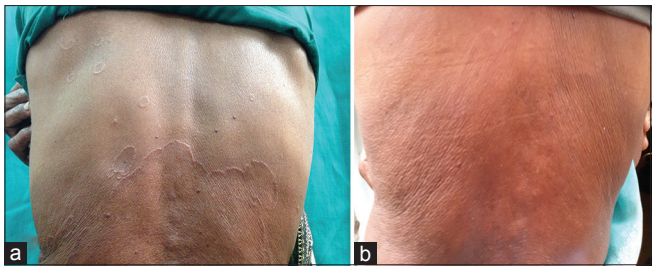

Figure 1: (a) Annular elastolytic giant cell granuloma (AEGCG) involving back, and (b) regressed AEGCG lesions in the back

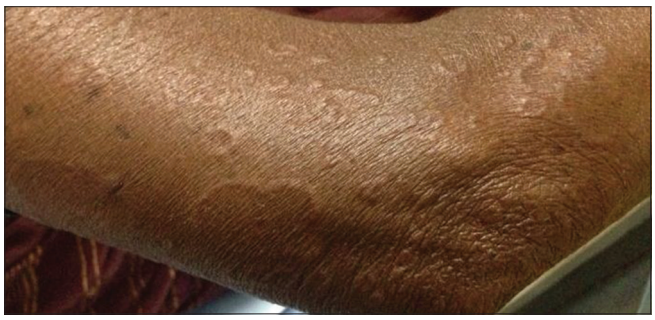

Figure 2: Annular elastolytic giant cell granuloma involving extensor aspect of elbow

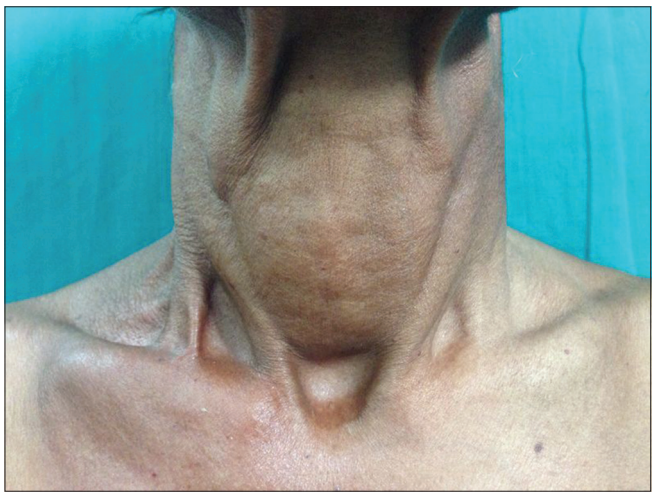

Figure 3: Diffuse thyroid swelling with bosselated surface

cells, lymphocytes, and fibrosis against a hemorrhagic background admixed with colloid, which was suggestive of HT.

Histopathologial examination (HPE) of a lesion from the back revealed diffuse granulomatous infiltrate composed of multinucleated giant cells, histiocytes, and lymphocytes in the dermis. No features of active vasculitis, necrobiosis, or mucin deposition were noted [Figures 4 and 5]. Special staining with Verhoeff–van Gieson stain showed loss of elastic fibers in upper and mid-dermis and fragmentation of elastic fibers with occasional elastophagocytosis, all features suggestive of AEGCG [Figures 6 and 7].

Based on HPE findings and FNAC study, a clinical diagnosis of AEGCG in association with HT was entertained. She was prescribed topical steroids and emollients, but there was no significant improvement after two weeks of treatment. She was referred to an endocrinologist for the management of HT.

Two months later, she presented with much regressed skin lesions [Figure 1b] and gave a history of having undergone thyroid surgery at a nearby tertiary care hospital. Details of the surgical procedure are unavailable.

DISCUSSION

Hanke *et al*. in the year 1979 described a rare granulomatous cutaneous disorder, AEGCG, clinically characterized by solitary or grouped papules forming annular plaques with elevated borders and central atrophy. The lesions tend to occur mainly in photoexposed areas and on covered areas rarely,[1] similar to the clinical picture in the present patient, who had asymptomatic plaques in non–photo-distributed locations also.

The diagnosis of AEGCG is mainly based on distinct histopathological findings, which includes granulomatous infiltrate with lymphocytes, histiocytes, and multinucleated giant cells in the upper and mid-dermis. Characteristic and unique features of AEGCG include fragmentation of elastic fibers, presence of scanty elastic fibers in the areas of the granulomatous infiltrate, and elastophagocytosis by giant cells,[3] all of which were noticed in HPE in the present case. The importance of dermatopathology cannot be overemphasized in this case.

The principal differential diagnosis for AEGCG is granuloma annulare (GA), which is considered as an artificial distinction by some authors.[4] Although some elastolysis has also been described in GA, the complete loss of elastic tissue in the central zone is used as the basis for the separation of these two conditions. The presence of larger and more numerous giant cells is also a feature in favor of AEGCG, as was observed in the present patient.[4] In view of the many giant cells and elastolysis, authors too wish to hold the view that AEGCG and GA are two separate entities. Also, absence of mucin and collagen necrobiosis helps to establish the distinct identities of the two conditions.

Regarding the genesis of AEGCG lesions, speculations point to ultraviolet radiation, heat, or other unknown factors that might change the antigenicity of elastic fibers, triggering a cellular immune reaction directed to them.[1] AEGCG has been associated with acute myelogenous leukemia, CD4 T-cell lymphoma, adult T-cell leukemia, and prostate

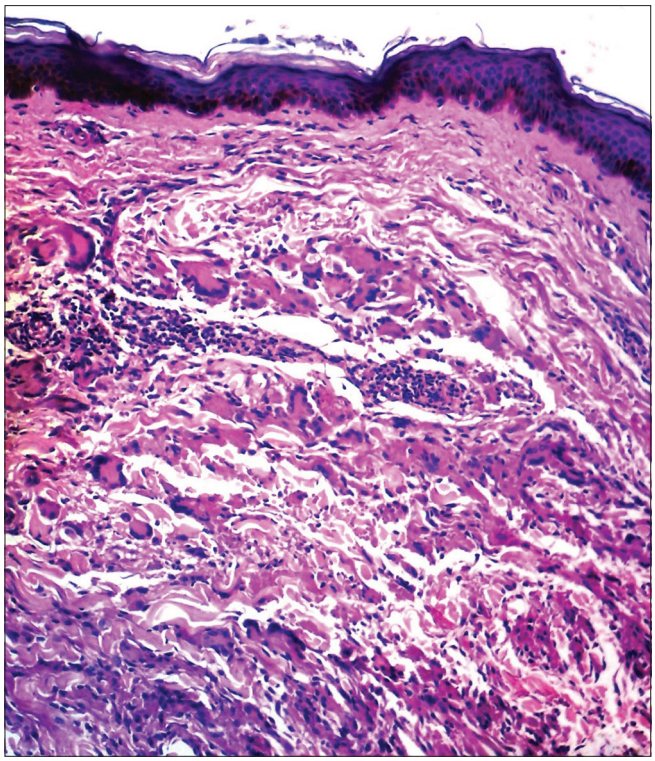

Figure 4: Diffuse granulomatous infiltration composed of multinucleated giant cells, histiocytes, and lymphocytes in the dermis (H and E, ×200)

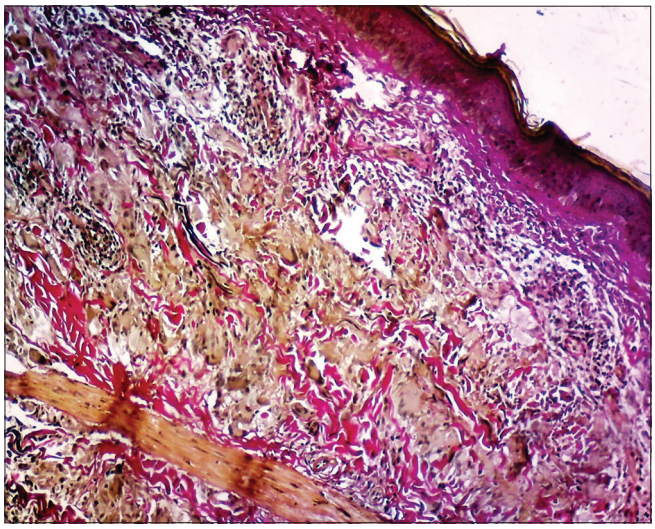

Figure 6: Verhoeff–van Gieson stain showing loss of elastic fibers in upper and mid-dermis (Verhoeff–van Geison stain, ×100)

carcinoma,[5] and this association with malignancy has been interpreted as systemic immunologic host defence against the tumor antigen.[6] In the present patient there was no clinical or laboratory evidence for any underlying malignancy.

HT is an autoimmune disease, known to occur usually in elderly females. Based on the typical clinical features, positive antithyroid peroxidase antibody, and classical FNAC

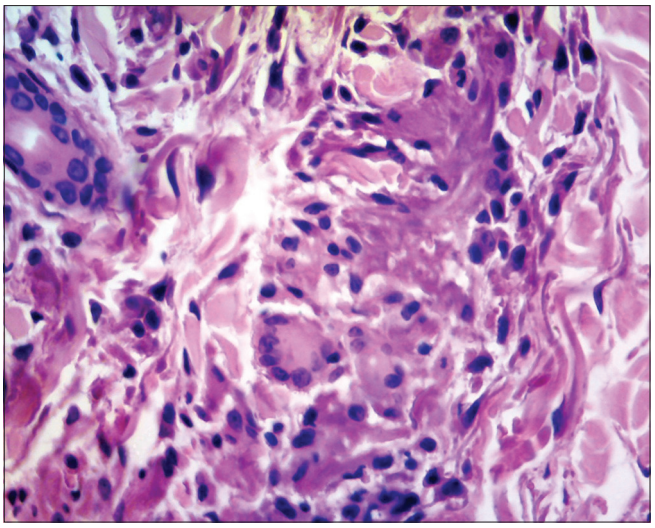

Figure 5: Granulomatous dermal infiltrate made up of histiocytes and multinucleated giant cells containing fragments of elastic fibers (H and E, ×400)

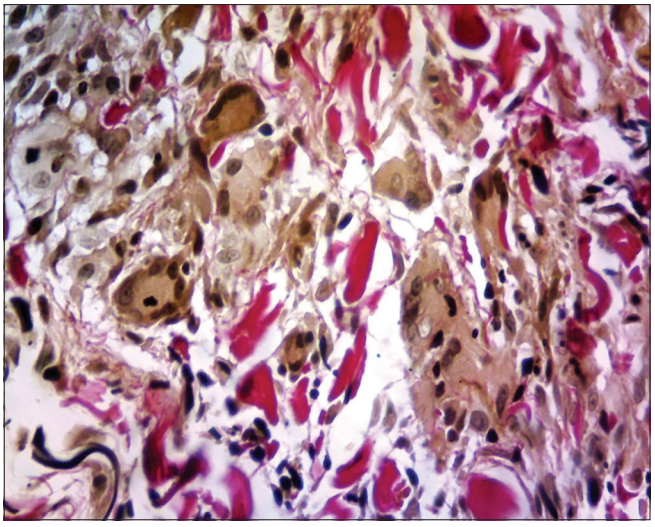

Figure 7: Fragmentation of elastic fibers with elastophagocytosis by multinucleated giant cells (Verhoeff–van Geison stain, ×400)

appearances, the diagnosis of HT was established in this case.

Muller and his coworker considered AEGCG as a prodromal stage of mid-dermal elastolysis (MDE) and also suggested that AEGCG and MDE might represent different stages in the clinical spectrum of dermal elastolysis.[7] Also Gambichler *et al*. has reported a case of MDE associated with HT, suggesting an autoimmune mechanism.[8] In the present case the association of AEGCG with HT, which is often of an autoimmune etiology, could be attributed to a similar mechanism.

AEGCG tends to run a chronic course; however, cases of spontaneous remission have been recorded.[9] Treatment

options include topical and intralesional steroids, topical tacrolimus and pimecrolimus, hydroxychloroquine, isotretinoin, dapsone, cyclosporine, and PUVA therapy with variable response.[10] Whether the near complete resolution of the AEGCG following thyroidectomy is due to the elimination of underlying contributory immune mechanism or a spontaneous phenomenon, cannot be ascertained conclusively in the present patient.

Alhough there are previous reports of AEGCG occurring in isolation or in association with systemic conditions, especially malignancies, the present case is being reported for the rare co-existence of AEGCG with HT.

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Conflicts of interest

There are no conflicts of interest.

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