Net Case

Effective treatment with hydroxychloroquine in a case of annular elastolytic giant cell granuloma

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ABSTRACT

Annular elastolytic giant cell granuloma (AEGCG) is a rare granulomatous and elastolytic skin disease of unknown pathogenesis. Therapy for AEGCG is controversial. The data about the effectiveness of chloroquine in the treatment of AEGCG are also variable. Here, we report a case of AEGCG with significant improvement after a total treatment period of 22 weeks with hydroxychloroquine. Although a possibility of spontaneous remission cannot be ruled out, we think that chloroquine can be considered as an effective treatment of this chronic disorder.

Key words: Annular elastolytic giant cell granuloma, treatment, hydroxychloroquine

INTRODUCTION

Annular elastolytic giant cell granuloma (AEGCG) is a rare granulomatous and elastolytic skin disease of unknown pathogenesis and variable clinical picture.[1-5] The differential diagnosis consists of a large spectrum of skin diseases from histopathological and clinical point of view.[1,2,5] Therapeutic results are usually unsatisfactory with currently utilized treatment modalities. The data about the effectiveness of chloroquine in the treatment of AEGCG are also controversial.[1,2,5]

Here, we report a patient of AEGCG with generalized lesions which significantly improved with hydroxychloroquine treatment.

CASE REPORT

A 55-year-old fair-skinned Turkish woman who wore traditional dress covering her whole body except the face and hands presented with sudden-onset generalized erythematous eruption developing 8 months ago. The lesions were asymptomatic, but progressive in size and number. She was using atorvastatin for hyperlipidemia and esomeprazole for gastric ulcer over the last year and denied any exposure to sunlight.

On dermatological examination, generalized, symmetric, erythematous papular lesions forming sharply demarcated annular plaques on the neck, upper chest, right foot and extensor surfaces of the hands and forearms were detected [Figures 1a-d].

Histopathology of a punch biopsy which was taken from the papular lesions on the left wrist revealed non-palisading granuloma on the upper and mid reticular dermis with elastolysis [Figures 2a-d].

A diagnosis of AEGCG was made both with clinical and histopathological findings. Complete blood count, erythrocyte sedimentation rate, and biochemistry were normal. Ocular examination revealed no abnormality.

Hydroxychloroquine 200 mg/day bid was started in the treatment. The patient was also advised to avoid sun exposure and to use sunscreen cream on the face. The

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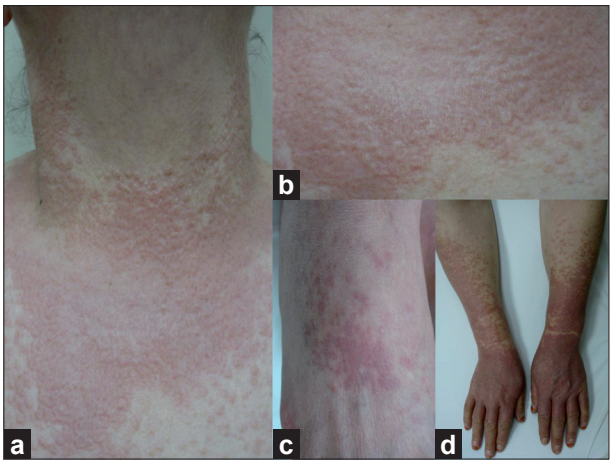

Figure 1: Erythematous papular lesions forming sharply demarcated annular plaques both on the sun-exposed and covered areas: (a) neck, (b) upper chest, (c) right foot, (d) extensor surfaces of the hands and forearms (before treatment)

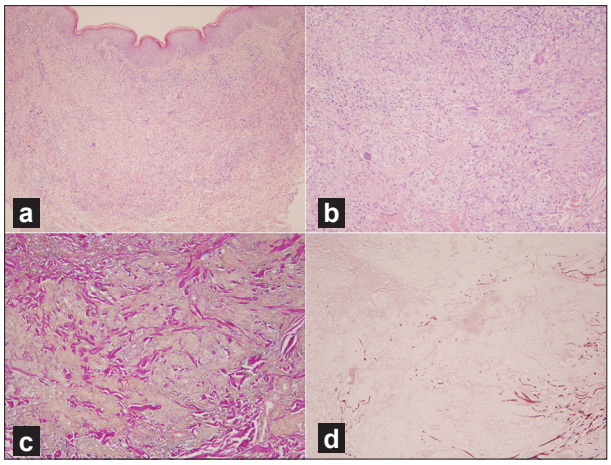

Figure 2: (a) Under low power, there is a heavy cellular infiltrate which contains many giant cells on the upper and mid reticular dermis (H and E, ×100); (b) In the central area of the lesion, a cellular infiltrate composed of many giant cells, histiocytes, and lymphocytes around degenerated collagen fibers (H and E, ×200); (c) Hale's colloidal iron stain fails to show obvious mucin deposition on this area (colloidal iron, ×200); (d) Orcein stain disclose a striking elastolysis on the lesional area, compared to the periphery (Orcein, ×100)

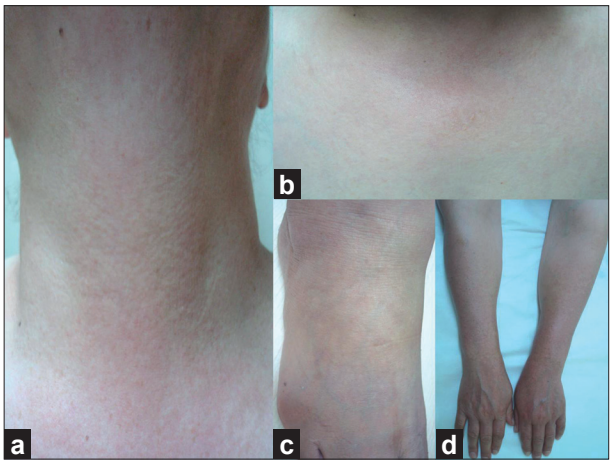

Figure 3: (a-d) Complete resolution of the lesions with only a slight postlesional erythema (after 22 weeks of hydroxychloroquine treatment)

lesions stopped to progress and started to improve. Flattening of the papules and plaques with reduction of the erythema was observed within 8 weeks of treatment. Subsequently, the dose was tapered to 200 mg/day. Hydroxychloroquine was stopped after a total treatment period of 22 weeks hence lesions regressed completely without any side effects [Figures 3a-d]. In the last visit, which was 6 months after the discontinuation of treatment, the patient was still in remission without any relapse.

DISCUSSION

AEGCG has been recognized by Hanke *et al.* in 1979 as a separate disease entity.[1] It has previously been defined as atypical annular necrobiosis lipoidica of the face and scalp, Miescher's granuloma of the face, and actinic granuloma (O'Brien).[2]

The etiopathogenesis of AEGCG is still unclear. Ultraviolet radiation, heat or other unknown factors are suggested to change the antigenicity of elastic fibers leading to the development of a cellular immune reaction.[2]

AEGCG usually affects fair-skinned middle-aged women, similar to our case.

The clinical picture is quite variable.[1-5] Although it mostly presents with solitary or multiple annular or ring-shaped patches with elevated borders, central hypopigmentation and atrophy, generalized papular lesions can also be seen.[2,3] AEGCG is generally considered to be a disease mainly affecting sun-exposed areas, but rarely it can also localize on covered areas.[6,7] Generalized lesions involving both sun-exposed and covered areas with no history of sun exposure was an interesting finding in our case providing further evidence that photosensitivity may not be the only factor in the etiopathogenesis of AEGCG.

The diagnosis of AEGCG is mainly based on clinical and distinct histopathological findings, like the

presence of granulomatous reaction with elastolysis and phagocytosis of elastic fibers by multinucleated giant cells (elastophagocytosis, not specific for AEGCG) with the absence of palisading histiocytes, necrobiosis, lipid, or mucin.[1,2] The main differential diagnosis includes generalized granuloma annulare (GA) and other diseases such as sarcoidosis, annular lichen planus, lichenoid photosensitivity reactions, subacute cutaneous lupus erythematosus, and inflammatory mid dermal elastolysis (type 2).[2,5] Histopathology is the key factor in differential diagnosis.

There is no standard therapy for this chronic disorder which can also show spontaneous remission.[8] Treatments achieving successful results include intralesional or systemic corticosteroids, excision of a solitary lesion, retinoid-PUVA, cyclosporine, topical tacrolimus, tranilast with topical pimecrolimus or topical steroids, dapsone, fumaric acid esters, narrow-band UVB, and quinacrine.[2,3,9-14] In contrast, topical corticosteroids, PUVA, cauterization, cryotherapy, methotrexate, and isotretinoin were found to be unsuccessful.[2,15] In addition to that, clofazimine has been reported as both effective[16] and ineffective[2] in two separate cases.

Hydroxychloroquine is an antimalarial agent, which is also used widely and successfully in the treatment of several connective tissue disorders and granulomatous diseases. The results of chloroquine therapy in AEGCG is quite variable; it has been found to be ineffective,[1] only partially effective[1,5] or solely effective[2,14] in single cases. It was previously found to be effective and safe in a patient with generalized AEGCG, who was unresponsive to clofazamine therapy.[2] Although a possibility of spontaneous remission cannot be ruled out, the disease progression stopped and lesions healed after starting hydroxychloroquine in the presented case. Therefore, we think that hyroxychloroquine seems to be an effective and safe alternative in the treatment of AEGCG.

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