A Ringed Enigma: A Case of Annular Elastolytic Giant Cell Granuloma Successfully Treated with Hydroxychloroquine

Dear Editor,

Annular Elastolytic Giant Cell Granuloma (AEGCG) was first reported by O'Brien in 1975. O'Brien used "actinic" in the name because he believed that its etiology was linked to ultraviolet and infrared radiation. Also known as actinic granuloma, O'Brien granuloma and Miescher's granuloma, this entity is a non-infectious granulomatous and elastolytic skin disorder of unknown pathogenesis, characterized by annular plaques with elevated borders and central atrophy with a tendency to occur over sun-exposed areas predominantly in middle-aged women.[1]

A 35-year-old female, laborer by occupation, presented with multiple asymptomatic reddish to skin-colored raised lesions which were gradually progressive in size and number for the past two years. She first noticed a few tiny asymptomatic reddish lesions over bilateral forearms which gradually progressed to the present size and number. Although there was a history of aggravation of lesions during summers, the lesions persisted even during winter. She did not suffer from any comorbidities. Personal and family history was unremarkable. General and systemic examination was within the normal limits. Cutaneous examination revealed multiple well-defined, erythematous, annular and serpiginous plaques measuring approximately two–ten cm in size, with indurated erythematous and elevated borders and central atrophic hypopigmented areas present over the bilateral forearms, thighs, and flank region [Figure 1]. Mucosae and nails were uninvolved. Sensations over the plaques were intact, and there was no evident nerve thickening. Routine hematological and biochemical investigations, including blood sugar levels, thyroid function test, and serum anti-nuclear antibody titer were within normal limits. Histopathology showed normal epidermis with non-palisading granulomatous infiltrate of histiocytes, lymphocytes, and Langhans type multinucleated (more than ten nuclei) giant cells with mild elastophagocytosis and elastotic degeneration present outside the granuloma in the upper dermis [Figure 2]. Necrobiosis was absent and dermal mucin was not seen on alcian blue staining. Other special stains like periodic acid-Schiff stain and Fite-Faraco stains were negative. Based on clinical and histopathological findings, a diagnosis of AEGCG was made. She was advised strict photoprotection with broad spectrum sunscreen. She was prescribed hydroxychloroquine PO 200 mg once daily after ophthalmological screening, and white petroleum jelly for topical application. On subsequent monthly follow-ups, there was significant flattening of the plaques and reduction of erythema at six months [Figure 3]. The patient did not report any significant side effects with the usage of hydroxychloroquine.

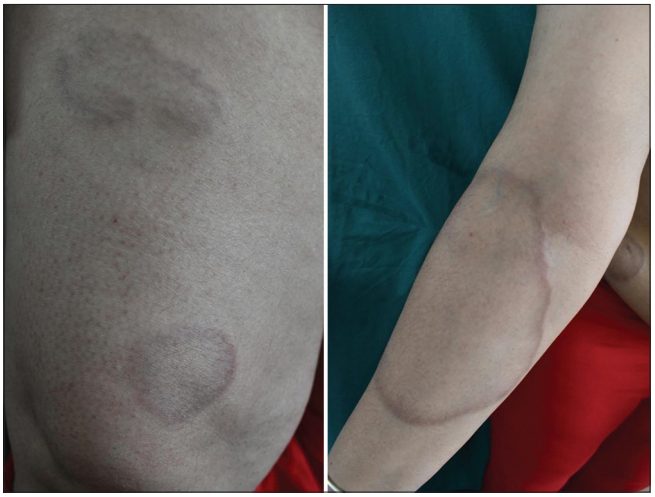

Figure 1: Annular plaques with erythematous raised border and atrophic center over thigh and forearm

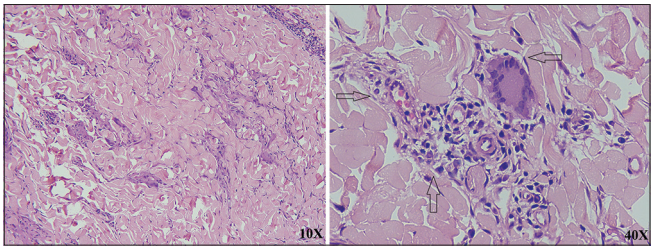

Figure 2: Non-palisading granulomatous infiltrate of histiocytes, foreign body type giant cells, with the arrow showing elastophagocytosis (H & E 40x)

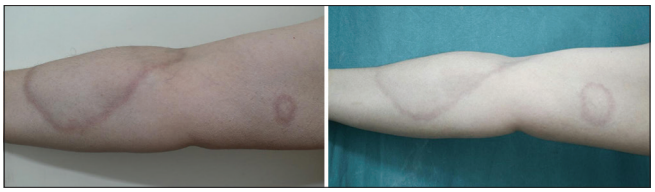

Figure 3: Significant flattening of lesions and reduction in erythema

AEGCG is an uncommon, idiopathic inflammatory skin disease of middle-aged women. Its pathogenesis is not well understood, but may be related to inflammation precipitated by actinic damage. Specifically, O'Brien postulated that "a state of auto-aggression develops in relation to the damaged elastic fibers," in an attempt to repair or remodel damaged skin. The most frequently associated risk factor is intense sun exposure or/tanning beds. Recently, many atypical variants of this condition, including papular, reticular, generalized forms, concomitant conditions,[2] and lesions involving

sun-protected areas had been reported. The clinical differential diagnosis includes the annular variants of the diseases like tinea corporis, granuloma annulare (GA), granuloma multiforme, necrobiosis lipoidica, sarcoidosis, and erythema annulare centrifugum. Larger lesions with central atrophy and hypopigmentation distributed on the trunk and extremities in AEGCG, contrast from the smaller rings of GA seen on the distal aspects of the limbs, particularly, hands and feet. In GA, histology shows foci of necrobiosis surrounded by granulomatous zone, giant cells with few nuclei (two-three), elastotic material, and mucin deposition within the granulomas. Granuloma multiforme is very similar in clinical presentation but it can be differentiated on the basis of histology. Although, a similar zonal pattern is seen with a granulomatous response unlike AEGCG, necrobiosis is seen along with mucin deposition in the dermis. Various treatment modalities have been employed like intralesional/systemic corticosteroids, topical calcineurin inhibitors, hydroxychloroquine, psoralen plus ultraviolet-A radiation therapy, clofazimine, methotrexate, and azathioprine. There are case reports of improvement with dapsone, systemic retinoids, and chloroquine therapy.[3] The mechanism of action of antimalarials is incompletely understood and is under active scrutiny. They act on multiple cellular targets, including ion channels, nucleic acids, and cellular organelles. Recent research has implicated their role in modifying innate immunity pathways including toll-like receptors, which ordinarily induce Interferon type 1 production.[4] Hydroxychloroquine in AEGCG has been studied in multiple reports, but the response has been variable.[5] We prescribed hydroxychloroquine for a period of six months and the patient was followed up for a period of one year with no recurrence of lesions.

Here, we report a rather unusual presentation of AEGCG over photo exposed as well as protected areas. Hence, histopathology played a critical role in clinching the diagnosis. Hydroxychloroquine given over six months emerged as a safe and effective treatment modality for this condition.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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