Annular Elastolytic Giant Cell Granuloma In A Ten Year-Old Child

HN Azakli, Ş Ağirgöl, A Çağlar, B Taş

ABSTRACT

Annular elastolytic giant cell granuloma is a rare granulomatous disease characterized by

annular erythematous plaques with central atrophy, which is generally observed in sun-

exposed areas of the skin. Actinic injury is thought to be involved in pathogenesis, yet its

exact reason has not been elucidated. Elastolysis along with elastophagocytosis constitute its

typical histopathological features. The disease is reported to show a tendency towards being

observed in middle aged women. In this paper, we presented a ten year-old patient diagnosed

with annular elastolytic giant cell granuloma.

Keywords: Annular elastolytic giant cell granuloma granuloma annulare, annular elastolytic

giant cell granuloma, granuloma

¹Bağcilar Education and Research Hospital, Department of Dermatology, Istanbul, ²Milas Government Hospital, Department of Dermatology, Mugla, ³Bağcilar Education and

Research Hospital, Department of Pathology.

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Correspondence: Dr HN Azakli, Bağcilar Eğitim ve Arastirma Hastanesi, Bağcilar/Istanbul,

Turkey. E-mail: cenuraz@hotmail.com

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INTRODUCTION

Annular elastolytic giant cell granuloma (AEGCG) is a rare granulomatous skin disease characterized histopathologically by phagocytosis of elastic fibers by multinucleated giant cells and clinically by annular patches with elevated borders and atrophic centers found mainly on sun-exposed skin. Although its etiology remains unknown, actinic injury is thought to play role as underlying mechanism considering its predominance in favor of sun-exposed areas of the skin. The disease is usually seen in middle aged women. We presented this case report of AEGCG, since it is a rare condition which should be included in the differential diagnosis of confounding diseases such as granuloma annulare, sarcoidosis, and tuberculosis and that it has a distinguishable feature as being a pediatric patient (1,2).

CASE REPORT

A ten year-old boy was admitted to our outpatient clinic with the chief complaint of wounds on his forehead which started about one year ago as raised lesions whose number increased gradually. Past medical or family history revealed no specific features. Physical examination was normal, except that in dermatological examination, frontal region showed several plaques of 0.5-2 cm in diameter, characterized by central atrophy and hypopigmentation with elevated and erythematous borders (Figure 1). No pathological finding was seen either in routine blood tests and urinalysis or in chest X-ray. Histopathological examination of the two punch biopsy specimens obtained separately from elevated margin and central atrophic area showed non-palisading, non-mucinous granuloma formations having peripheral lymphocytic inflammatory infiltration associated with multinuclear giant cell histiocytes in focal interstitial area and between collagen fibers in epidermis and dermis (Figure 2 & 3); and loss of elastic fibers in dermis (Figure 4). The diagnosis of AECGC was established upon these clinical and

histopathological findings. Since the patient was lost to follow-up, treatment and subsequent actions could not be initiated.

DISCUSSION

Although AEGCG was initially thought to be a form of granuloma annulare manifested at sun-damaged areas and hence termed as actinic granuloma (3), later reports of rare cases affecting skin parts not exposed to sun and of those cases with no evidence of actinic injury have made this condition to be considered as a distinct clinical entity (1).

AEGCG is usually seen in 30-50 year-old women. Literature search showed two large case series, first by O'Brien consisting of 19 female patients with an age range of 33-48 years (3) and another case series by Hanke et al. reporting 5 female patients whose ages ranges from 31 to 59 years (1). While majority of remaining case reports points at this pattern of age distribution, presentation of cases of adolescence and childhood is limited (4).

Etiopathogenesis of AEGCG, presenting with a clinical manifestation of dermal plaques characterized by elevated margins and atrophic centers on sun-exposed parts, was suggested to involve the development of elastic fiber injury by humoral immune response activated against elastic fibers which acquire antigenic properties under the influence of UV radiations (8,9). Histopathologically, while erythematous margins of lesions indicate a granulomatous infiltration with phagocytosis of elastic fibers by multinuclear giant cells, central zones of lesions reveal diminishment or complete loss of elastic fibers (8,10,11). Distinctive features such as elastolysis, elastophagocytosis, and non-palisading structure of granuloma along with the absence of mucin accumulation, necrosis, or necrobiosis favors AEGCG regarding the differential diagnosis among granuloma annulare, necrobiosis lipoidica, and sarcoidosis (12).

Having a chronic course with spontaneous healing potential, treatment options of AEGCG include intralesional corticosteroids, or systemic steroids, isotretinoin, acitretin, chloroquine, cyclosporine, and PUVA in case of presence of diffuse lesions (2,13).

In conclusion, AEGCG being already a rare condition that should be considered in differential diagnosis of annular and granulomatous diseases, is seen in childhood even rarer. We presented a case of pediatric patient showing typical clinical and histopathological characteristics of AEGCG, with the aim of making modest contribution to the literature.

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Fig 1: Atrophic plaques surrounded with erythema on the forehead

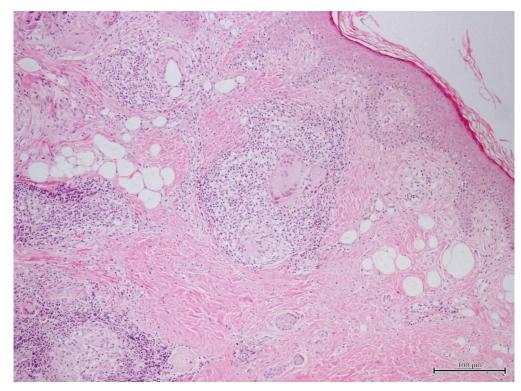


Fig 2: Granuloma formation accompanied by multinucleated giant cell histiocytes that do not form palisade within the collagen fibers of epidermis and dermis (H&E, X100)

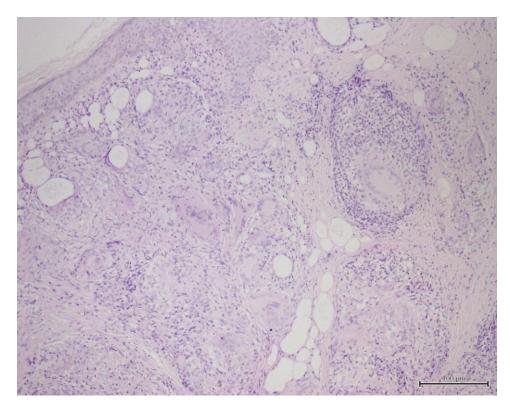


Fig 3: PAS staining showed no sign of mucinous material (PAS, X100)

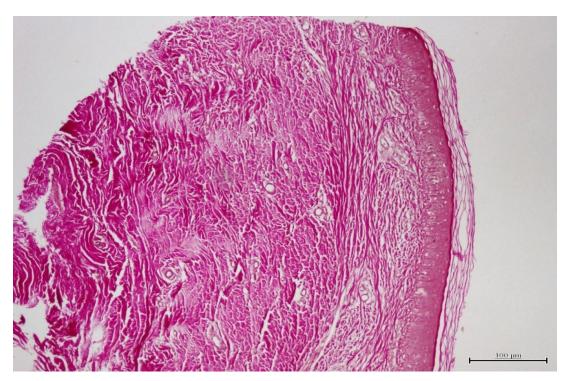


Fig 4: Loss of elastic fibers in dermis (Elaston Von Gieson, X100)