Quiz

Annular and serpiginous plaques in an old man

An 80-year-old male farmer presented with multiple reddish raised lesions on the abdomen and limbs, of 4 months duration. Past history was unremarkable. On examination, the patient had multiple discrete erythematous and skin colored annular, serpiginous, arciform and polycyclic plaques distributed on the abdomen, thighs and upper limbs [Figure 1]. Systemic examination was within normal limit. Routine investigations were normal. Skin biopsy showed multinucleate giant cells with palisading arrangement along with scanty elastic fibers in the upper and mid dermis [Figure 2]. Verhoeff von Gieson stained section showed elastic fiber degeneration and elastophagocytosis by giant cells granuloma [Figure 3].

WHAT IS YOUR DIAGNOSIS?

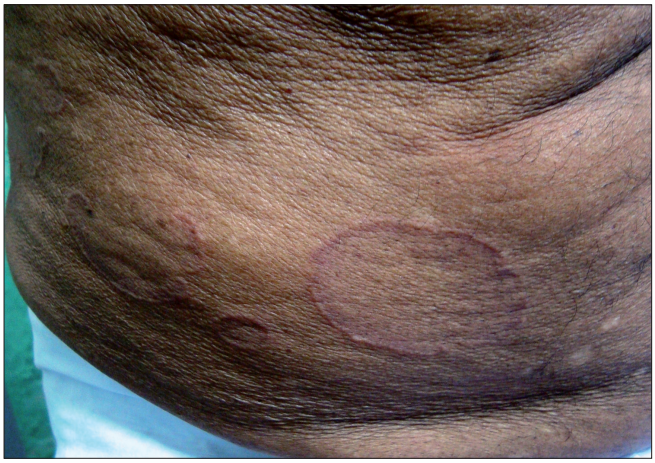

Figure 1: Annular, serpiginous and polycyclic plaques on abdomen

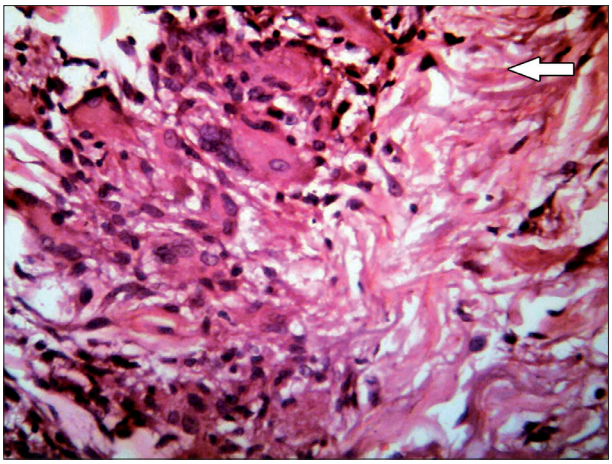

Figure 2: Palisading of giant cells with horizontal arrangement of collagen fibers (arrow) (H and E, ×400)

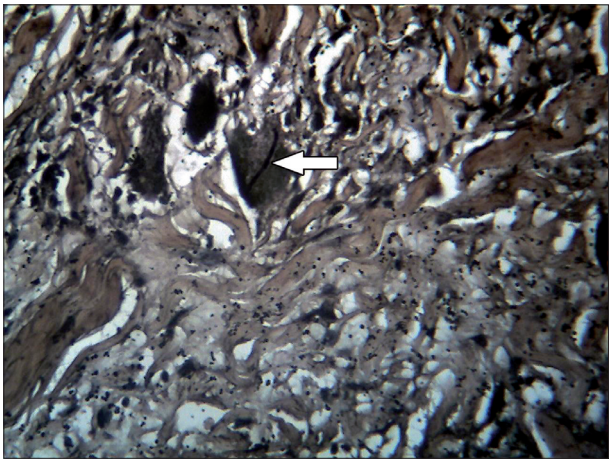

Figure 3: Scanty broken elastic fibers with elastphagocytosis (arrow) (Verhoeff van Geison, ×400)

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ANSWER

Annular elastolytic giant cell granuloma

DISCUSSION

Annular elastolytic giant cell granuloma (AEGCG) is a recently described entity with unknown etiology. It was first coined by Hanke *et al.*, for patients presenting with annular erythematous plaques on the sun exposed areas and classified under the noninfectious granulomatous group of disorders.[1] The disease usually occurs in middle-aged females, even though males are also affected.[2] It is grouped as a noninfectious granuloma along with actinic granuloma, atypical necrobiosis lipoidica, Meissner's granuloma and granuloma multiforme. However, some consider it as an entity with distinct clinical and histopathologic features. Actinic induced damage to the elastic fibers is now considered to be the hallmark of the disease even though lesions can occur in the sun protected areas also.[3]

The clinical presentation of AEGCG may resemble granuloma annulare, actinic granuloma or granuloma multiforme. The patient presents with annular, serpiginous, arciform and polycyclic plaques, usually on the sun exposed areas. Lesions on the sun protected areas are rare even though they have been reported in literature.[3] Presentation with annular plaques on the sun exposed areas encounters a wide variety of dermatoses. However, the histopathology of AEGCG is diagnostic. Presence of scanty elastic fibers in the area of the granulomatous infiltrate, palisading by multinucleate giant cells and elastophagocytosis by giant cells [Figure 3] are the histopathologic hallmarks of AEGCG. Elastophagocytosis by giant cells with abundant distribution of giant cells in the periphery is a characteristic and unique feature of AEGCG.[4] Presence of horizontally arranged collagen fibers resembling scar tissue [Figure 3] is another important feature of AEGCG.[5] Presence of mucin and collagen necrobiosis in the dermis distinguishes granuloma annulare from AEGCG. Actinic granuloma may clinically and histopathologically resemble AEGCG, but the characteristic elastophagocytosis is absent. Granuloma multiforme may mimic clinically AEGCG, but histopathology is characterized by necrobiosis which is absent in AEGCG. Chronic actinic damage of elastic fibers is considered to be the triggering factor for AEGCG.[6] Studies have shown that actinically damaged elastic fibers become antigenic, and 67 kDa elastin receptors are expressed by the epitheloid cells and the giant cells in the granuloma along with Factor XIIIa + dendritic cells and CD 68+ macrophages.[2-4] AEGCG has been associated with acute myelogenous leukemia, CD4 T-cell lymphoma, adult T-cell leukemia, cutaneous amyloidosis and squamous cell carcinoma of the lung.[7,8] However, these associations are casual. There is no definite treatment for AEGCG. Systemic steroids, cyclosporine, dapsone, chloroquine, tranilast (hemostatic agent), fumaric acid esters and topical tacrolimus/pimecrolimus are the drugs effective, according to case reports.[9] However, there are no randomized controlled trials in literature. Reports of AEGCG in Indian literature are extremely rare.[10] AEGCG should be considered in the differential diagnosis of annular plaques.

C. Soni Das, S. Pradeep Nair, V. Sreedevan, Lissy Skaria, Rani Mathew, Rony Mathew

Department of Dermatology and Venereology, T.D. Medical College, Alleppey, Kerala, India

Address for correspondence: Dr. S. Pradeep Nair, Department of Dermatology and Venereology, T.D. Medical College, Vandanam, Alleppey - 688 005, Kerala, India.
E-mail: dvmchtvm@yahoo.co.in

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