Annular elastolytic giant cell granuloma after a cardiac pacemaker implantation

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INTRODUCTION

Annular elastolytic giant cell granuloma (AEGCG) is an uncommon condition belonging to the family of elastolytic granuloma, which also includes actinic granuloma, atypical necrobiosis lipoidica of the face and scalp, and Miescher's granuloma.[1] Classification of AEGCG as a distinct entity has been controversial. Although some consider it synonymous with actinic granuloma, a term often used interchangeably with AEGCG in the literature, others regard elastolytic granulomas as a variant of granuloma annulare.[2,3] These controversies exist largely because the etiology and pathogenesis of AEGCG is still unknown. Here we describe an unusual case of AEGCG that developed after a cardiac pacemaker implantation. The circumstances that surrounded the eruption of our patient's rash may suggest that trauma is a possible trigger for AEGCG.

Abbreviation used:

AEGCG: annular elastolytic giant cell granuloma

CASE REPORT

Our patient was a 76-year-old white man who presented with a widespread rash on his chest and upper back (Figs 1 and 2). He reported that within days after a cardiac pacemaker implantation, an isolated lesion erupted on the skin directly over the pacemaker. Over a 2-year period, without any spontaneous regression or color changes, this asymptomatic lesion slowly spread across the chest and upper back. The patient presented to us 2 years after the onset of the rash, and we performed a biopsy. The histopathology findings showed zonal interstitial giant cell–rich histiocytic infiltrate of the mid-dermis (Fig 3, *A* and *B*). Multinucleate giant cells were seen phagocytizing elastic fibers with subsequent variable central loss of elastic tissue. There were no significant deposits of mucin or necrobiosis nor presence of palisading histiocytes. Based on the clinical and histologic presentation, AEGCG was diagnosed.

The patient's rash resolved completely without residual atrophy or dyspigmentation after 1 month of twice-daily application of 0.1% triamcinolone cream. The lesions persisted in areas of his back where he could not reach to apply the medication.

DISCUSSION

AEGCGs can present initially as 1 or more papules that enlarge into annular/polycyclic plaques. Despite its name, AEGCG can also present simply as diffuse papules. There is a predilection for the trunk and neck. The macroscopic zonal differences have corresponding histologic features: biopsy of the center is devoid of elastic fibers, whereas the rim is marked by a granulomatous reaction consisting of histiocytes confined to the mid-dermis. A hallmark of AEGCG is the phagocytosis of elastotic material by histiocytes, often in the presence of giant cells, termed

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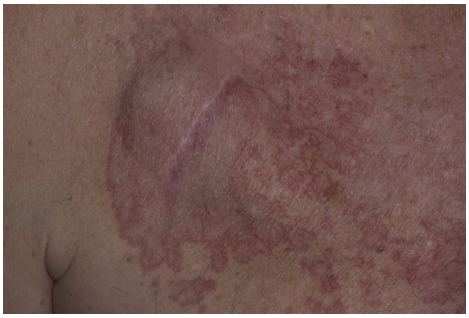

Fig 1. Sentinel lesion on the right upper chest, which was a well-demarcated annular-to-polycyclic plaque superimposed on an implanted cardiac pacemaker.

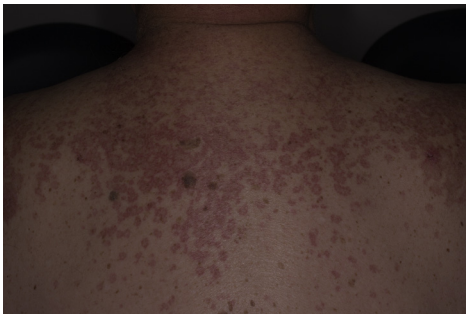

Fig 2. Multiple small erythematous papules and annular plaques, some of which have coalesced, on the upper back.

elastophagocytosis. The absence of palisading histiocytes, necrobiosis, and mucin is a histologic feature of AEGCG that distinguishes it from granuloma annulare.

The pathogenesis of AEGCG remains a mystery. O'Brien and Regan[3] coined the term *actinic granuloma*, a related condition to AEGCG, and hypothesized that ultraviolet-induced damage and antigenization of elastin fibers are responsible for the granulomatous process. This theory is supported by a report of actinic granuloma occurring in the setting of prolonged doxycycline-induced phototoxicity.[4] In general, the diagnosis of actinic granulomas is reserved for AEGCG lesions confined to sun-exposed regions.[1] Recent molecular studies found an up-regulation of metalloproteinase-12 in AEGCG lesions, postulating its role in the degradation of elastic fibers.[5] Elastic fibers are essential to the development of AEGCG because scars devoid of such fibers are found to be spared of AEGCG.[6] The literature contains mixed reports of AEGCG responding to immunotherapy. This finding suggests that the immune system is involved in the pathogenesis of AEGCG, but the exact mechanism remains unknown.

To our knowledge, there are no reports of AEGCG occurring after a cardiac pacemaker implantation. We considered foreign body granulomas as a pathogenesis for our patient, as foreign body granulomatous reactions have been reported with cardiac pacemaker wires.[7] In these cases, however, elastolysis was absent, and removal of the wires was curative. More importantly, a foreign body granulomatous process would not explain our patient's widespread rash, as foreign body granulomas are generally confined to the area surrounding the foreign body. We suspect that trauma during the implantation may have caused lysis of elastic fibers that initiated the granulomatous cascade, which then developed into a sentinel lesion on the skin directly over the pacemaker before spreading. Pestoni et al[8] described a similar case in which a young man had AEGCG over an area of his leg that had been repeatedly traumatized from the heat of an exhaust pipe and was further exacerbated by mechanical trauma. Nonetheless, pacemaker implantation is a common procedure, yet AEGCG has not been widely reported. Therefore, we cannot exclude the possibility that it was a coincidence our patient had AEGCG after the implantation of the pacemaker.

The literature reports mixed outcomes for the treatment of AEGCG. Some investigators report significant resolutions, albeit with residual atrophy or dyschromia, using topical pimecrolimus,[9] systemic minocycline,[2] and hydroxychloroquine.[10] These authors cautioned that the possibility of a spontaneous resolution could not be ruled out. In our patient, the lesion persisted in areas where medication could not be applied, which strongly suggests that the rash was responding to topical triamcinolone. Although AEGCG is not fatal, it may cause significant psychosocial morbidity to the patient, especially if it occurs in cosmetically sensitive areas. Clinicians who are aware of its presentation, especially under unusual circumstances such as a pacemaker implantation, are best equipped to diagnose and treat the condition.

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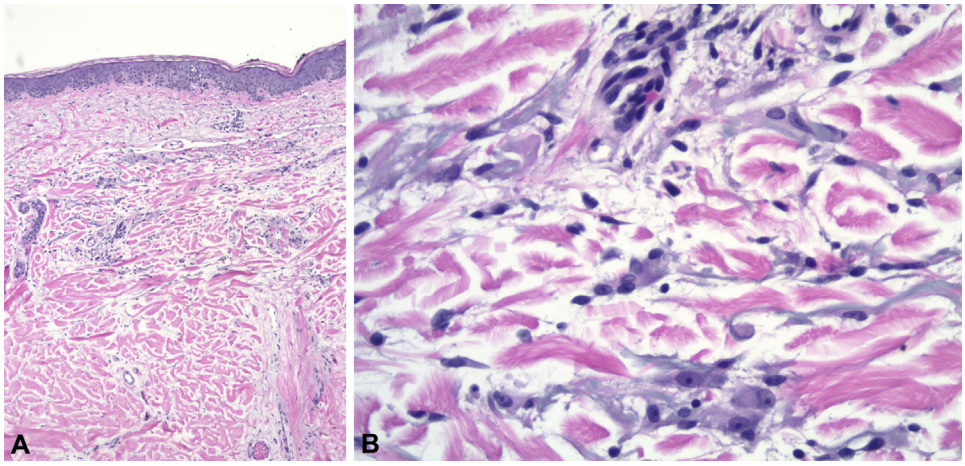

Fig 3. **A**, Histopathologic image of a biopsy section taken from a lesion on the upper back, depicting mid-dermal interstitial infiltrate of histiocytes and giant cells, without necrobiosis, mucin, or palisading histiocytes. **B**, Biopsy specimen depicts elastic tissue fragments, elastophagocytosis, and multinucleate giant cells. (**A** and **B**, Hematoxylin-eosin stain; original magnifications: **A**, ×4; **B**, ×20.)

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