

Disseminated Papular Interstitial Elastolytic Giant Cell Granuloma

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Sir,

Elastolytic giant cell granuloma (EGCG) is a rare granulomatous skin disease, the main pathological findings being the formation of non-palisading granuloma and phagocytosis of elastic fibres by histiocytes and/or multinucleated giant cells (1–5). Size, shape and the number of lesions vary greatly. In general, the lesions are localized on sun-exposed areas and show an annular configuration (1). We present a patient with numerous small papules with no annular lesions developing on either sun-exposed or non-sun-exposed areas, which histopathologically showed interstitial EGCG.

CASE REPORT

A 52-year-old Japanese man visited us with a 4-month history of an asymptomatic skin eruption on the trunk and limbs. He had been treated by a local dermatologist with oral and topical steroids to no avail before his visit to our clinic. Physical examination revealed multiple red-brown, elastic hard small papules disseminated on his trunk, shoulders, forearms and legs (Fig. 1). At some places there was coalescence between neighbouring papules forming irregular, erythematous patches. The laboratory study of his peripheral blood, which was performed because of suspected granulomatous diseases or lymphomas, showed no abnormal changes in differential blood cell counts, electrolytes or biochemical data, including serum levels of angiotensin-converting enzyme and aldolase. No abnormal lesions were detected in the lung, heart or eyes. The histopathological picture of one of the papules showed normal epidermis and a perivascular lymphocytic infiltrate associated with interstitial granulomatous changes in the upper half of the reticular dermis. The granulomatous lesion was accompanied by multinucleated giant cells (Fig. 2). The presence of naked granuloma, which is one of the characteristic features of sarcoidosis, could be found at only one small site among several histological sections. Elastica-Masson stain showed a total loss of elastic fibres in the area with granulomatous changes (Fig. 3). There was no evidence indicating collagen destruction or mucin deposition, i.e. the characteristic features of granuloma annulare (GA) (data not shown).

The patient was started on treatment with a topical application of 0.05% clobetazole propionate ointment and most lesions had almost cleared by 4 weeks.

DISCUSSION

We present a unique case of numerous papules disseminated mainly over the trunk, and that histologically showed interstitial EGCG. Several skin diseases must be considered for the differential diagnosis, including annular elastolytic giant cell granuloma (AEGCG), sarcoidosis, GA and other skin diseases showing interstitial granuloma.

AEGCG is a rare granulomatous skin disease first described by Hanke et al. in 1979 (11). Over 30 cases have been reported in the literature (1), most showing solitary or multiple annular or ring-shaped patches 1–6 cm in diameter with elevated borders. The formation of non-palisading granuloma and phagocytosis of elastic fibres by multinucleated giant cells are the main pathological findings. AEGCG can be ruled out in this case because its infiltration pattern is similar to that of sarcoidosis, i.e. it is diffuse rather than interstitial.



Fig. 1. Numerous papules disseminated over the trunk.

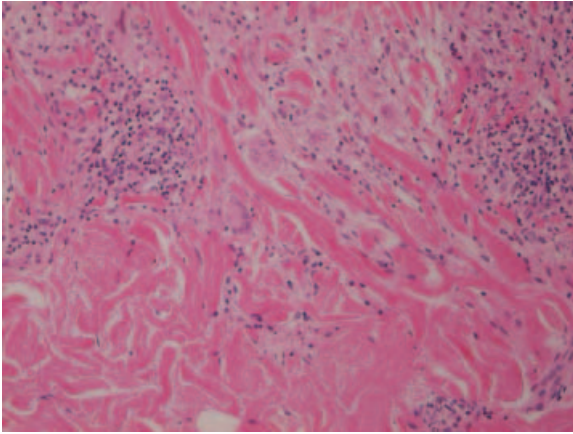


Fig. 2. Interstitial infiltration of histiocytes with some giant cells was seen in the upper reticular dermis. Lymphocytes were found at the perivascular portions (H&E stain. $\times 200$ the original magnification).

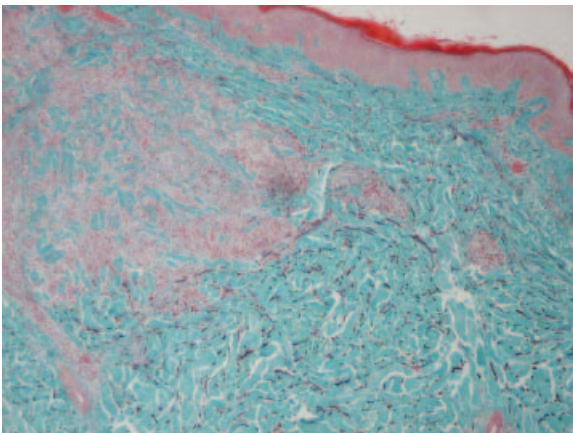


Fig. 3. Elastic fibres had disappeared inside the granulomatous lesion (Elastic-Masson stain. $\times 200$ the original magnification).

Sarcoidosis is known to show several different clinical types, among which we have recently reported a rare annular elastolytic type (6). The histological findings are similar to those found in AEGCG owing to the existence of elastolytic change in the granulomatous lesion. However, sarcoidosis can be ruled out in the present case from the results of laboratory examination and the existence of interstitial granulomatous changes rather than the characteristic naked granuloma (12).

GA also resembles AEGCG clinically (1). The histopathological characteristics of GA are the presence of palisading granuloma with a central necrobiotic change and mucin deposition. Mucin deposits are found in approximately 80% of patients with GA (6), but not in AEGCG (1). On the other hand, it has been pointed out that elastolytic changes, which are characteristic of AEGCG, were not found at all in 6 cases of GA

(7, 8). In our case, GA could be excluded because the granulomatous lesion histologically showed elastolytic changes with interstitial infiltration of histiocytes together with some giant cells but with neither necrobiotic change nor mucin deposition.

Interstitial granulomatous dermatitis (IGD) has recently been reported in association with rheumatoid arthritis, clinically showing a linear arrangement of erythema. In IGD, the histopathological findings show a band-like, but not interstitial, infiltration of histiocytes in the reticular dermis surrounding necrotic collagen bundles (9, 10). The presence of elastolytic changes has not been reported in IGD. Thus, IGD can be excluded in the present case by the differences in the clinical features and the absence of elastolytic changes in the dermis.

To the best of our knowledge, there has been no similar clinical case report in the English literature describing numerous small papules with no large annular lesions, and histologically interstitial EGCG. From these observations, we propose the descriptive term of “disseminated papular interstitial elastolytic giant cell granuloma” for the present case.

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