

Annular Elastolytic Giant Cell Granuloma

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annular infiltrative plaques over the dorsum of his arms and forehead for six years. Mild central atrophy was noted in each lesion. There was no loss of sensation or enlargement of the peripheral nerves. There was no similar skin lesion in his family members. No significant drug history was noted. He came to Hong Kong for four years and worked as a security guard for 12 years with 12-hour outdoor patrol each day.

CASE SUMMARY

History and physical examination

A 55-year-old-Nepalese-male complained of multiple asymptomatic dusky red annular patches with raised edge around his knees (Figures 1 and 2) and lower part of his trunk for eight months and

Concerning his past health, he is a chronic alcoholic; drinking three cans of beer per day for about 25 years. His liver is cirrhotic with esophageal varices. He also got syphilis in 1974 and was treated in Nepal with penicillin injection.

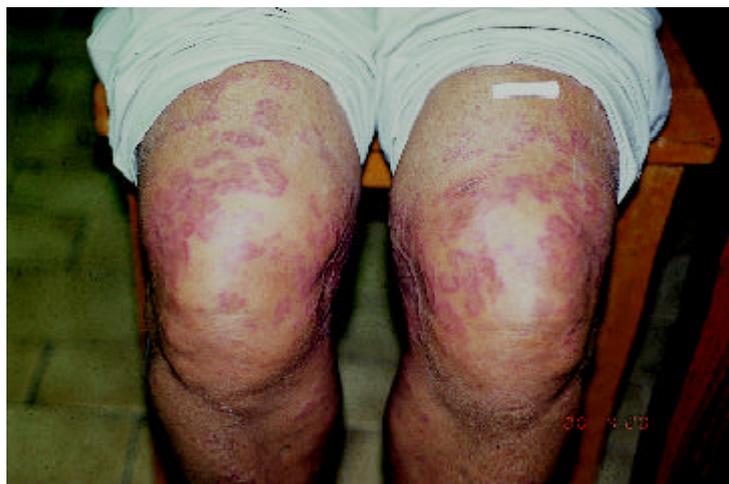


Figure 1: Multiple asymptomatic dusky red annular patches with raised edge around his knees



Figure 2: Close up of lesion at Figure 1. Note the mild central atrophy

Differential diagnoses

These included tuberculoid leprosy, granuloma annulare, necrobiosis lipoidica diabetorum, cutaneous sarcoidosis, and deep fungal infection.

Investigations

His liver function was grossly impaired and complete blood picture showed with thrombocytopenia. The chest X-ray was normal and calcium level was within normal limit. The antinuclear factor was slightly raised but the anti-DNA and anti-ENA were both negative. His blood for VDRL was non-reactive and FTA, TPHA were reactive. Tissue smear and culture showed no mycobacteria tuberculosis and other mycobacteria isolated. Microscopic examination showed focal degenerative collagen and elastic fibers associated with multinucleated giant cells and palisading histiocytes in background of solar elastosis (Figures 3 and 4). The overlying epidermis showed mild increase in thickness. The skin appendages were unremarkable. There was no increase in mucin. The features were consistent with that of elastolytic granuloma.

Diagnosis

The diagnosis is annular elastolytic giant cell granuloma.

Management

The patient was advised to avoid sun-exposure and application of high SPF sunscreen. He was reassured about the benign nature of the disease. Since he will leave Hong Kong soon, no further trial of systemic treatment was given.

REVIEW OF ANNULAR ELASTOLYTIC GIANT CELL GRANULOMA

Definition

Annular elastolytic giant cell granuloma (AEGCG) is a rare granulomatous skin disease described by Hanke et al. in 1979. It is characterized by presence of multinucleated giant cells containing elastic fibres at the lesion edge, and the absence of elastic fibres at the

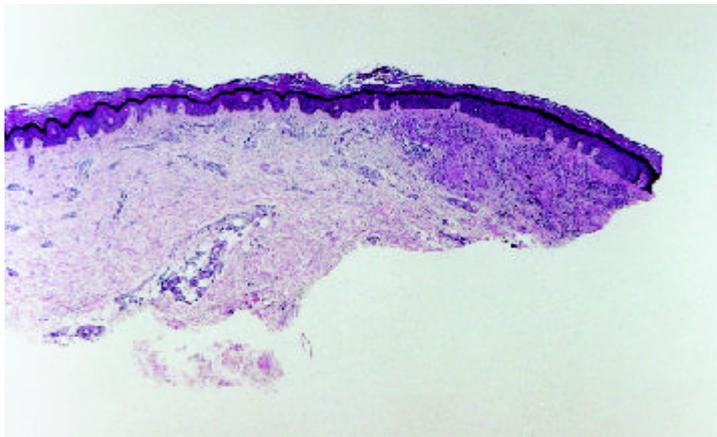


Figure 3: This biopsy includes the edge of the annular lesion wherein are granulomatous aggregate of histiocytes, some engulfing dermal elastic fibres (H&E) (By courtesy of Dr. W. Y. Lam, Department of Pathology, TMH)

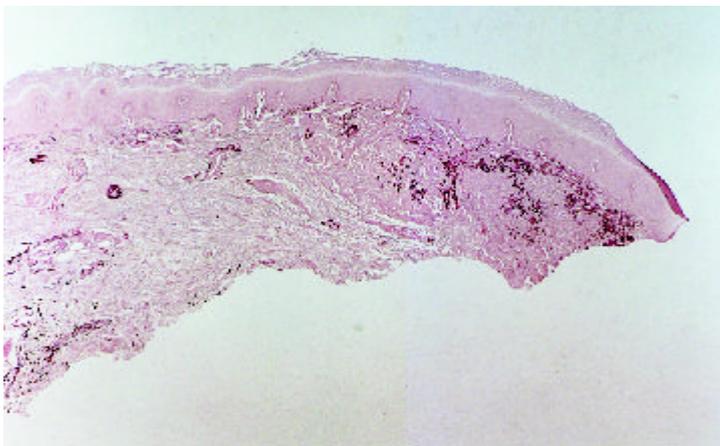


Figure 4: This elastic stain shows elastic fibres (brown streaks) phagocytosed within the granuloma of histiocytes, and the centre of the lesion shows absence of elastic fibres (Orcein) (By courtesy of Dr. W. Y. Lam, Department of Pathology, TMH)

lesion centre without necrobiosis or mucin and lipid deposition.¹

Clinical features

The clinical picture is quite variable with lesions varying in size, number and shape. Lesions are either solitary or grouped in annular patches with elevated borders and central atrophy as seen in our patient. They are mostly found in middle-aged white women, mainly localized on sun-exposed areas and rarely on covered areas. The lesions may be pruritic or asymptomatic. There are about 30 reported cases of AEGCG in the literature.^{1,6}

One case report mentioned that a 15-year-old Japanese girl with AEGCG had bilateral granulomatous uveitis and multiple elastolytic granuloma formation appeared in the cervical lymph nodes, parietal peritoneum, and mesentery. The patient also had seronegative arthritis of both knees, slight hepatosplenomegaly, iron deficiency anaemia, and leukocytopenia;² indicating that systematic involvement could occur. Hanke also reported two cases of AEGCG associated with necrobiosis lipoidica and sarcoidosis.¹ The course of AEGCG is chronic. Spontaneous remission have been reported in three patients within one year.¹

Histopathology

Generalized granuloma annulare (GA) is the main clinical and histopathological differential diagnosis. It is characterized by poor response to therapy and protracted course, which may also be seen in AEGCG. Thus the main differentiating point relies on the presence of a palisading granulomatous reaction with elastolysis and elastophagocytosis in AEGCG.¹ Although elastophagocytosis seems to be a non-specific phenomenon which is also found in some cases of generalized GA, the distinctive zone of elastophagocytosis with abundant distribution of giant cells at the periphery of the lesion in AEGCG are different from GA. Moreover, the absence of collagen necrobiosis and mucin deposition further differentiate it from GA.¹

Other differential diagnoses include sarcoidosis, tuberculoid leprosy, annular lichen planus, subacute lupus erythematosus and lichenoid photosensitivity reactions. The histopathology is the main differential feature. Despite the above distinctive features, Ragaz and Ackerman still considered AEGCG as GA occurring in sun-damaged skin.³

The pathogenesis of AEGCG is still unknown. It is postulated that ultraviolet radiation, heat or other unknown factors might change the antigenicity of elastic fibres which may trigger a cellular immune reaction directed to them. The immunohistochemical finding of CD4+ cells predominate over CD8+ cells in the inflammatory infiltrate seems to support this hypothesis.⁴ Granuloma formation and elastophagocytosis might be a reflection of the inflammatory process targeting the dermal elastic tissue. The final result is rarefaction or complete absence of elastic fibres and quiescence of the inflammatory reaction in the centre of the lesion.

Treatment

Among various treatments, clofazimine, chloroquine,¹⁵ dapsone, excision of a solitary lesion, intralesional or systemic corticosteroids,¹ retinoid-PUVA (RePUVA) and cyclosporin⁴ were reported to be effective in single-reported cases.

Cauterization, cryotherapy, PUVA,¹ topical corticosteroids¹ and methotrexate⁴ were reported to be ineffective in other single-reported cases. Because of the small number of cases described, there was no controlled study in the treatment of this disease.

Learning points:

It is still controversial whether annular elastolytic giant cell granuloma is a specialty of granuloma annulare occurring in sun-damaged skin, or a separate entity.

References

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