Case 12

A 62-year-old Thai man from Nakonsritammarat

Present history:

Chief complaint: Itchy rash for 1 month

Present illness: The patient noticed pruritic faintly erythematous papules and plaques on upper chest & back, extensor side of both forearms and legs for 1 month. These rashes gradually increased in number over time.

Past history:

Diabetes mellitus, hypertension, gout

History of DRESS from allopurinol

Family history:

Nil

Physical examination:

Multiple confluent erythematous annular papules and plaques on upper back, upper chest and extensor surfaces of both forearms and legs (Fig 12.1, 12.2)

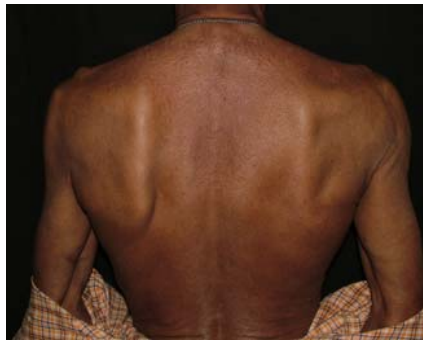

Fig 12.1

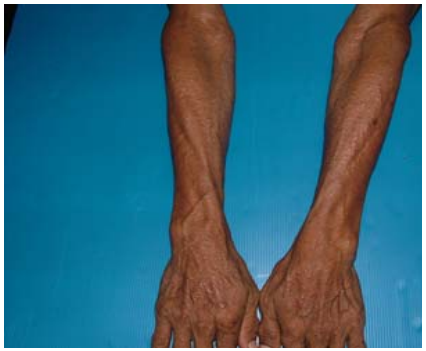

Fig 12.2

Histopathology : (S09-1554)

- Superficial and deep perivascular and interstitial infiltrate of lymphocytes and histiocytes in the dermis
- Lymphocytes around blood vessels and scattered histiocytes and multinucleated giant cells between collagen bundles
- Increase number of thick elastotic material and some within the giant cells
- Absent elastotic fibers in the central zone
- Mucin deposit between collagen bundles in some foci

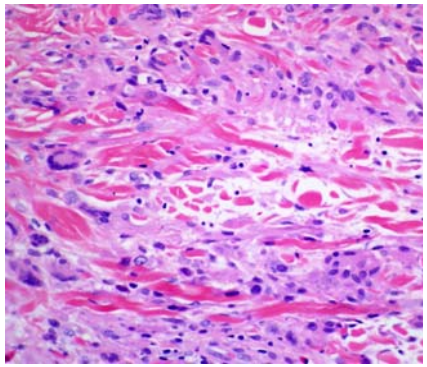

Fig. 12.3

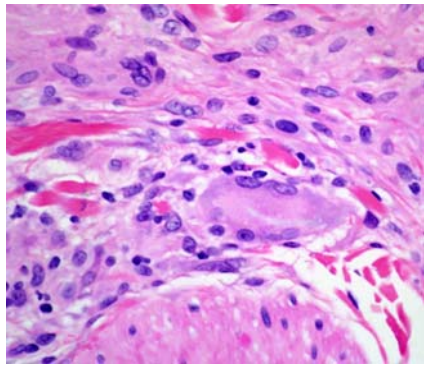

Fig. 12.4

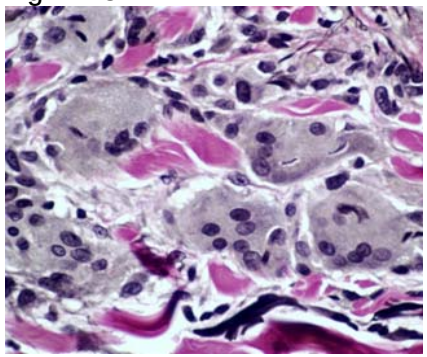

Fig. 12.5

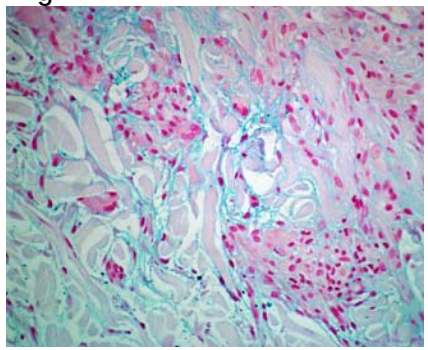

Fig. 12.6

Diagnosis : Annular elastolytic giant cell granuloma

Presenter : Chayada Kokpol
Consultants : Kumutnart Chanprapaph
Discussion:

Annular elastolytic giant cell granuloma, or actinic granuloma, is an uncommon, idiopathic disorder of middle-aged adults, which occurs more commonly in women than in men. Patients have been reported from several areas of the world, including Australia, UK, USA, the Caribbean, and Africa.[1,2]

The pathogenesis of the disease is obscure but actinic hypothesis is mostly accepted. The postulated mechanism behind the actinic damage assumption is that solar-damaged elastic fiber are weakly antigenic and result in a cell-mediated immune response with a predominance of CD4 lymphocytes. This immune response attempted to repair the actinically damaged skin resulted in granulomatous inflammation instead. Support for this hypothesis comes from the sun-exposed distribution of the disease, the presence of CD4cells, and the presence of giant cells that appear to be ingesting the elastic fibers. This evidence, however, is merely correlative and does not address whether the granuloma are the consequence of or are actually the cause of the damaged elastic fiber.[1,3,5]

Annular elastolytic gaint cell granuloma presents with large annular plaques, raised erythematous borders (3-5 mm) and slightly atrophic, hypopigemnted central regions that are distributed mainly in sun-exposed areas. The sites of predilection include the neck, face, chest and arms. Scale is rarely observed. Individual lesions measure 1-10 cm in diameter, and the total number of lesions is generally less than ten. Patients are usually asymptomatic.[2,3]

The histologic features are best appreciated in a radial biopsy that contains the central zone, the elevated border, and the skin peripheral to the ring.

The central zones shows the hallmark of the disease, that is, near or total absence of elastic fibers, best appreciated with an elastic tissue stain (e.g., Verhöeff-van Gieson). The collagen in this zone may show horizontally oriented fibers producing a slight scar like appearance. By contrast, the zone peripheral to the annulus shows an increased amount of thick elastotic material with the staining properties of elastic tissue.

Sections through the raised border show a granulomatous infiltrate with either of these patterns seen in granulomatous annulare, that is, sarcoidal granulomatous type, interstitial granulomatous type or, less commonly, in a palisade granulomatous type. Multinucleated histiocytes are conspicucous, usually being large and containing as many as dozen nuclei, mostly in haphazard arrangement, but sometimes in ringed array. Elastic fibers are identified adjacent to and within the giant cells, giving rise to the term elastophagocytosis.[5,6]

The principal differential diagnosis is granuloma annulare because engulfment of abnormal elastic fibers can also occur in granuloma annulare as well as in other granlomatous processes.[6]

Although some elastolysis has also been described in granuloma annulare, it is the complete loss of elastic tissue in the central zone that has been used as the primary basis for separating the diseases. Other features that have been evoked for distinguish them are the presence of larger and more numerous giant cells, the absence of mucin in annular elastolytic giant cell granuloma, and sparing of areas that lack elastic tissue, such as scars. [4,6]

Treatment of anuular elastolytic giant cell granuloma is difficult. The lesions tend to persist and respond little, if at all, to topical or intralesional glucocorticoids. There is anedoctal evidence for beneficial to acitretin, isotretinoin, cyclosporine, chloroquine, pentoxifylline, cryotherapy, PUVA, and methotrexate. Excision of lesion followed by partial-thickness skin grafting has been performed with no recurrence at 15 months of follow-up, but this would rarely be required. Some patients experience spontaneous resolution of the lesions. Patients are generally advised to avoid sun exposure and to wear sunscreen to prevent development of new lesions.[1,2]

References

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