"Ring"-Shaped Lesion Over Neck

Sunmeet Sandhu, Deepak Vashisht, Prashant Sengupta[1], Vikas Pathania

Department of Dermatology, Armed Forces Medical College, [1]Department of Pathology, Command Hospital, Pune, Maharashtra, India

Case History

A 73 years old female presented with an insidious onset asymptomatic palm-sized ring-shaped lesion over the neck (right) which had been progressing gradually for the past one year. There was no history of itching, altered or loss of sensation, prolonged drug intake before the onset of the lesion, any comorbidities such as diabetes mellitus or any systemic involvement. She had routine outdoor exposure to sunlight.

Examination revealed multiple well-defined shiny erythematous papules coalescing to form an annular plaque with central clearing over the supraclavicular region (Right) measuring 13 cm × 09 cm in maximum dimensions [Figure 1a]. Also, there were discrete to coalescing erythematous non-follicular dome-shaped papules over the upper back [Figure 1b]. Sensations were preserved over the lesion and peripheral nerves were not thickened. Mucosae, palms, soles, scalp, hair, and nails revealed no abnormality. Complete blood counts, liver and renal function tests, and other biochemical tests including angiotensin convertase enzyme (ACE) levels were within the normal range. Venereal Disease Research Laboratory (VDRL) test was negative and the chest X-ray was noncontributory.

Biopsy from the edge of the annular lesion revealed keratinized thinned out epidermis. Papillary dermis showed a dense collection of histiocytes and numerous multinucleated giant cells seen engulfing elastotic fibers and moderate perivascular lymphohistiocytic infiltrate [Figure 2a and b]. There was no increase in dermal mucin, necrobiosis, or palisading granuloma. Masson trichome section showed disarray of collagen with occasional giant cells with engulfed intracytoplasmic collagen [Figure 3a and b]. Ziehl-Neelsen and fungal stains were negative.

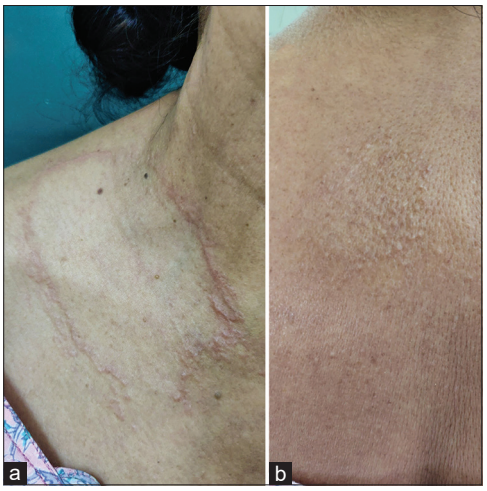

Figure 1: (a) Multiple shiny skin-colored to erythematous papules coalescing to formannular lesion with erythematous border and central clearing over right supraclavicular region mesuring 13 cm × 09 cm in maximum dimensions. (b) Discrete to coalescing erythematous nonfollicular dome-shaped papules seen over the upper back

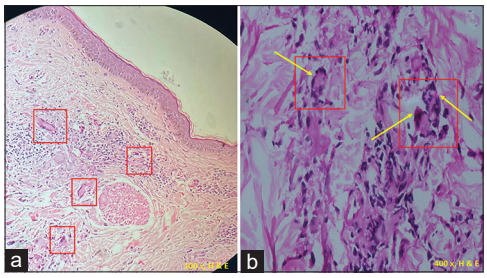

Figure 2: (a and b) Biopsy from the edge of the annular lesion revealed keratinized thinned out epidermis. Papillary dermis showed a dense collection of histiocytes and numerous multinucleated giant cells (red rectangles) seen engulfing elastotic fibers (yellow arrow) and moderate perivascular lymphohistiocytic infiltrate. (a: 100×;H and E and b: 400×; H and E)

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Address for correspondence:
Dr. Deepak Vashisht, Department of Dermatology, Armed Forces Medical College, Pune, Maharashtra - 411 040, India.
E-mail: deepak3975@gmail.com

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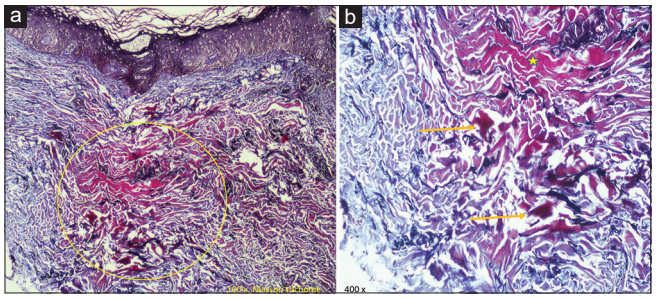

Figure 3: (a and b) Masson trichome section showed disarray of collagen (yellow circle and star) with occasional giant cells with engulfed intracytoplasmic collagen (orange arrow). (a 100×; Masson trichome b: 400×; Masson trichome)

Answer

Annular Elastolytic Giant Cell Granuloma (AEGCG).

Discussion

Hanke *et al*. first described Annular Elastolytic Giant Cell Granuloma (AEGCG) in 1979, a rare granulomatous clinical entity characterized by degradation of elastic fibers and its phagocytosis by multinucleated giant cells.[1] It usually manifests in the fourth to seventh decade with variable sex preponderance.[2] Predominant presentation is over sun-exposed areas as multiple small skin-colored to erythematous papules coalescing to form polycyclic annular lesion with raised borders to arciform lesions. There may be hypopigmentation to atrophic changes in the center. Clinical variants with only popular lesions, reticular, brown to livid, and partly atrophic lesions have been reported.[3]

Pathogenesis is not clear. Ultraviolet (UV) radiation mediates damage to elastic fibers via Matrix metalloproteinase (MMP) enzyme 12 resulting in antigenic stimulation by damaged elastic fibers inducing a cellular immune response. Sixty seven kDa elastin receptor is expressed on the epithelioid cells and giant cells in the granuloma.[4] Lesions can also occur over sun-protected areas as well, hinting at factors other than UV light in the pathogenesis. Casual association with other conditions such as diabetes mellitus and certain malignancies have been reported.

Clinical differentials include other annular and arciform skin conditions like granuloma annulare, granuloma multiforme, sarcoidosis, necrobiosis lipoidica, actinic granuloma, and Hansen's disease.

The hallmark histological feature is elastophagocytosis of dermal elastic fibers by the multinucleated giant cells with reduced or absent elastic fibers. The presence of horizontally oriented fragmented elastic fibers is characteristic with the absence of necrobiosis or mucin deposition distinguishing the entity from necrobiosis lipoidica and granuloma annulare.[3] Four histopathological variants including giant cell variant, necrobiotic variant, histiocytic variant, and the sarcoid variant have been described by O' Brien with giant cell variant being the commonest.[5,6]

Management of these cases is challenging and controversial with reports of spontaneous resolution over months to years leaving normal skin or mottled pigmentation.[2] Various local treatment modalities with topical and intralesional steroids, topical calcineurin inhibitors, retinoids, and cryotherapy are being used. Systemic therapy with steroids, hydroxychloroquine, acitretin, cyclosporine, clofazimine, dapsone, methotrexate, fumaric acid esters, tranilast, psoralen plus ultraviolet A therapy and narrowband ultraviolet B therapy are considered in resistant cases.[2,3]

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Conflicts of interest

There are no conflicts of interest.

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