“Ring”-Shaped Lesion Over Neck

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 × 9 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.

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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. 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.

Management of these cases is challenging and controversial with reports of spontaneous resolution over months to years leaving normal skin or mottled pigmentation. Various local treatment modalities with topical and intralesional steroids, topical calcineurin inhibitors, retinoids, and c 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.

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Conflicts of interest
There are no conflicts of interest.

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