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CORRESPONDENCE

A case of refractory hypertrophic lupus erythematosus on the face whose irreversible skin fibrosis was treated by local full-thickness skin graft under disease control with a combined use of topical and systemic immunosuppressants, and hydroxychloroquine

Hypertrophic lupus erythematosus (HLE) is a rare subtype of discoid lupus erythematosus (DLE), characterized by verrucous and hyperkeratotic nodulo-plaques with tendency of affecting the sun-exposed skin.[1] It represents a fluctuating clinical course and often intractable to conservative treatment.[2] The long-standing lesion may cause cosmetic/functional impairment with excess fibrosis and develop squamous cell carcinoma (SCC),[1,3,4] with unknown frequency. No recommended treatment has been currently available, but surgical management may provide somewhat benefit in the selected clinical course, particularly in cases where the skin lesions developed irreversible and excess fibrosis.

A 60-year-old Japanese man with a 3-decade history of repeated skin erosions on his nose and lips had been diagnosed with DLE (Figure 1A). The skin lesion had persisted and gradually elevated. Examination showed a red-yellowish irregular-shaped mass on the dorsal nose with indurative erythema on the nose wing and upper lip (Figures 1B,C). Skin pathology of the dorsal nose revealed parakeratotic hyperkeratosis, follicular plugging, and focal vacuolar degeneration of basal layer with marked elongation of the rete ridges in the epidermis (Figure 1D). There were intense inflammatory infiltrates, mainly consisting of lymphocytes, in the upper dermis. Direct immunofluorescence was positive for IgG at the dermo-epidermal junction (data not shown). Otherwise, he had neither systemic symptoms and organ involvements nor evidence of systemic lupus. Based on the clinical course and pathology, he was diagnosed as HLE arising from the preexisting DLE, and given a potent topical corticosteroid and oral hydroxychloroquine with gradual exacerbation. Concomitant oral prednisolone (20 mg/day) was effective for progression of the skin mass and inflammation, but failed to improve the fibrotic mass. Because of the minimized disease activity and complication suspicious for SCC, we performed excisional biopsy, followed by reconstruction using full-thickness skin graft from the supraclavicular area (Figure 1E). Histopathology of the whole skin mass displayed obvious dermal fibrosis with findings similar to the previous biopsy, albeit much lesser inflammation (Figure 1F). After healing of post-operative wound, topical tacrolimus was challenged to the residual DLE lesions with successful tapering of oral prednisolone up to 2 mg/day, being continued because of resistance to further dose reduction. The grafted skin blended gradually with color and texture differences in the surrounding skin by 15 months after operation (Figure 1G).

Hypertrophic lupus erythematosus remains a diagnostic and treatment challenge. The long-standing lesions may often evolve into disfiguring and cribriform scar, requiring consideration of alternative treatment for the cosmetic and functional impairment,[5,6] like our case. Besides, the treatment-resistant HLE needs to deny the possibility of malignancy when suspected. In addition to histological confirmation of malignant transformation, the surgical excision of the persisted HLE skin may offer the advantage in co-removal of cosmetically impaired, fibrotic mass and underlying sun-damaged skin to prevent the relapse, particularly facial involvement. Moreover, the full-thickness skin graft from nonexposed skin not only resurfaces the affected skin, but also provides a match in tension and texture to the surrounding skin. Surgical treatment at a proper timing may thus be one of the optional choices for irreversible fibrotic HLE, whose disease activity is controlled under conservative treatment.

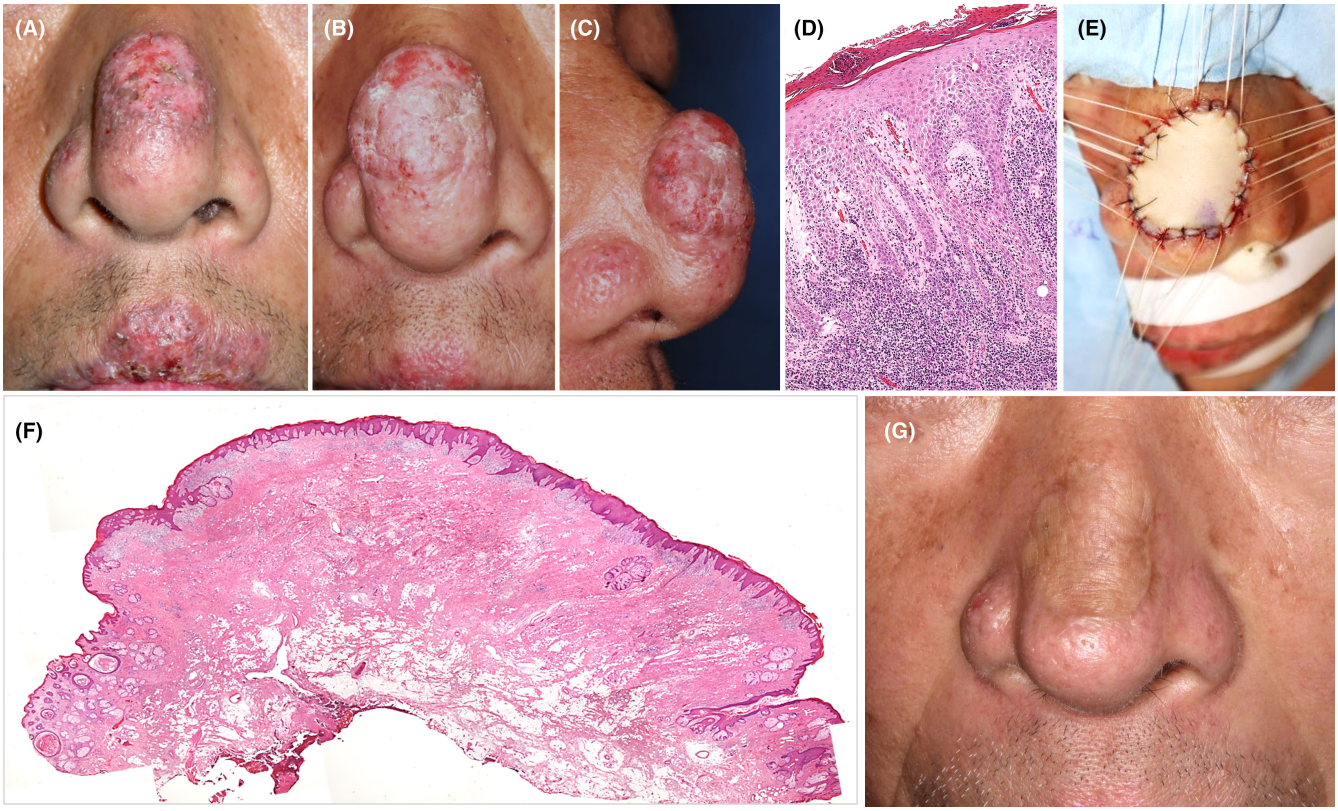

FIGURE 1 Clinical and pathological findings. (A, B) Scaly indurative erythema with erosions on the dorsal nose, nose wings, and upper lip, being given a diagnosis of DLE three decades ago. (C) A red-yellowish papillomatous mass on the dorsal nose. (D) The pathology of the biopsied dorsal nose skin showing parakeratotic hyperkeratosis, focal vacuolar degeneration of basal layer, dense lymphocytic infiltration, and marked elongation of the epidermal rete ridges, suggestive of HLE (×100, HE). (E) The whole skin mass in the dorsal nose was excised and reconstructed by full-thickness skin grafting from the clavicle after histological confirmation. (F) The pathology of the excised dorsal nose skin lesion displaying conspicuous fibrosis of the entire dermis with mild inflammatory cell infiltrates localized to the superficial dermis (x2, HE). (G) A 15-month post-operative clinical image demonstrating recurrence-free with relatively color and texture matching to the surrounding skin

CONFLICT OF INTEREST

The authors declare no conflicts of interest.

DECLARATION SECTION

Approval of the research protocol: No human participant was involved in this study.

Informed Consent: N/A.

Registry and the Registration No. of the study/trial: N/A.

Animal Studies: N/A.

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