Flagellate erythema in systemic sclerosis: A case report

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INTRODUCTION

Flagellate erythema is characterized by linear erythematous streaks with hyperpigmentation. The condition is a characteristic side effect of bleomycin, an antineoplastic sulphur-containing antibiotic developed in the 1960s, and might also occur after consumption of raw or undercooked shiitake mushrooms.[1] Moreover, it is less frequently associated with dermatomyositis or adult-onset Still disease.[2] Precise mechanisms for the formation of these lesions are unknown and seem to vary by etiology. We report the first described case of flagellate erythema associated with systemic sclerosis.

CASE REPORT

A 49-year-old man had extensive bilateral dermal sclerosis of his fingers and hands (Fig 1) associated with severe Raynaud phenomenon, which led to trophic disorders of both hands, and a dry cough for about 5 months. In addition to his acral lesions, he had nonpruritic, linear maculopapular erythematous plaques on the chest, upper back, shoulders, and arms (Figs 2 and 3). Old lesions progressively developed hyperpigmentation as new ones continued to appear. He denied consuming any new foods or medications, and his sun exposure did not increase. A computed tomography scan revealed early interstitial lung disease without fibrosis. Pulmonary function tests and a cardiac ultrasound showed restrictive lung disease and moderate pulmonary hypertension. The laboratory investigations revealed the presence of antinuclear antibodies (titer 1:1280) and Scl70 (topoisomerase 1-specific) antibodies, and kidney failure was not evident.

Skin biopsies of an old hyperpigmented lesion on the back and early erythematous lesions of the right shoulder demonstrated thickened collagen bundles with discrete lymphocytic perivascular infiltrates in the dermis and collagen fibrosis in the hypodermis (Fig 4). A few melanophages were present in the pigmented lesions.

These findings supported the diagnosis of systemic sclerosis and therapeutic management consisted in oral corticosteroids (prednisone 7.5 mg/day) and intravenous cyclophosphamide (700 mg/m^2/month). Lung disease was still progressing after 6 courses of cyclophosphamide.

DISCUSSION

Flagellate erythema is linear, whip-like erythema without an external cause. It is a characteristic but rare side effect of bleomycin therapy, and a number of cases have been described.[3] Flagellate erythema also occurs after consumption of raw or undercooked shiitake (*Lentinus edodes*), a type of mushroom initially used in Japanese and Chinese cuisine and in traditional Asian medicine.[1] This type of rash is classically named flagellate dermatitis or shiitake dermatitis and is increasingly being reported as shiitake consumption is expanding worldwide. Besides these 2 classic etiologies, flagellate erythema is also associated with dermatomyositis (described as

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Fig 1. Bilateral dermal sclerosis of fingers and hands of patient with flagellate erythema.

Fig 2. Linear erythematous and hyperpigmented lesions on back of patient with flagellate erythema.

zebra-like stripe eruption or centripetal flagellate erythema) and adult-onset Still disease.[1,2] Finally, a few cases have been reported in association with parvovirus B19 infection, systemic lupus erythematosus, and various types of chemotherapy.[4,5] To the best of our knowledge, this is the first case described in association with systemic sclerosis.

Exact mechanisms of flagellate erythema are unknown. Minor trauma is often used to explain the shape and distribution of the lesions. For example, in bleomycin-associated cases, scratching is supposed to lead to extravasation of the drug to the skin but as in our case pruritus might be absent.[3] Hyperpigmentation is described in bleomycin-associated cases; because bleomycin is supposed to reduce the turnover of the epidermis, prolonged contact between keratinocytes and melanocytes is induced, potentially causing the hyperpigmented lesions. In our case, histologic findings seem to indicate a postinflammatory hyperpigmentation.

In addition to flagellate erythema, bleomycin can also cause lung and skin fibrosis.[6,7] Yamamoto et al, upon histologic evaluation of a case of bleomycin-induced flagellate erythema, indicated the presence of dermal sclerosis.[8] Tumor growth factor ß and T_H2 cytokines, such as interleukin 4 and 6, appear to be involved in the pathogenesis of bleomycin-induced sclerosis, systemic sclerosis, and localized scleroderma in murine models and in humans.[7,9,10] These

Fig 3. Linear erythematous lesions on right shoulder of patient with flagellate erythema.

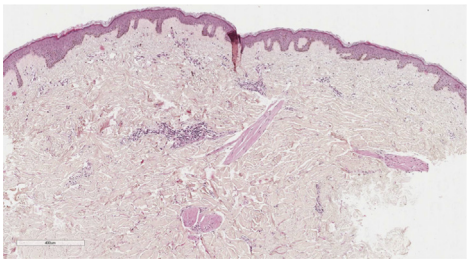

Fig 4. Thickened collagen bundles and collagen fibrosis in hypodermis of skin lesion tissue of patient with flagellate erythema. (Hematoxylin-eosin saffron stain; original magnification: ×25.)

biomolecules play an important role, among others, in the stimulation, activation, and extracellular-matrix production of the fibroblasts.

In conclusion, our case strengthens the proposed association between flagellate erythema and dermal sclerosis; however, additional research is needed to define the precise roles of these profibrotic cytokines in the peculiar shape of this skin eruption.

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