

Case Report

Generalized morphea: A Case Report

Kittisak Payapvipapong MD*,
Kobkul Aunhachoke MD*

Department of Dermatology, Phramongkutklao Hospital, College of Medicine, Bangkok, Thailand

Generalized morphea is a subtype of localized scleroderma that lacks systemic manifestations. It is a rare condition in which idiopathic sclerosis of the skin occurs in a widespread manner. We report a 37-year old Thai female who presented with multiple hypopigmented lesions on right leg and right arm with a skin biopsy showing the typical sclerotic change. The patient responded to treatment with topical calcipotriol.

Keywords: Generalized morphea, Localized scleroderma, Topical calcipotriol

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Scleroderma, a chronic disease of unknown etiology, is characterized by skin fibrosis and divided into two clinical entities: localized scleroderma and systemic sclerosis (SSc). The lesions of localized scleroderma are usually limited to the skin and subcutaneous tissue. Localized scleroderma differs from SSc by the lack of Raynaud's phenomenon, acrosclerosis, arthritis and internal organs involvement. Moreover, it carries a good prognosis. Localized scleroderma is classified into three types *i.e* 1) morphea en plaque, the most common form, 2) linear scleroderma and 3) generalized morphea⁽¹⁾.

We report a case of generalized morphea, a rare idiopathic sclerosis of the skin that occurs in a widespread manner.

Case Report

A 37-year old Thai female presented with a 9-year-history of asymptomatic whitish patches on her right leg. The lesions were gradually enlarged, becoming more indurate and painful. She had applied clobetasol propionate prescribed by a doctor and noted that the lesions became slightly softened and hyperpigmented. 4 years later, multiple hypopigmented lesions occurred on her right arm. She denied the history of Raynaud's phenomenon, dysphagia, dyspnea and photo-sensitivity.

Physical examination revealed multiple ill-

defined and well-defined hypo- and hyperpigmented sclerotic patches and plaques on her right arm, left arm and right leg. Sclerodactyly and periungual telangiectasia were not observed (Fig. 1, 2). A skin biopsy revealed flattening of rete ridges with sclerosis of collagen fibers in the upper dermis (Fig. 3).



Fig. 1 A 37-year old Thai female presented with well-defined hypo-hyperpigmented sclerotic patches and plaques on her right leg.

Correspondence to:

Payapvipapong K, Department of Dermatology, Phramongkutklao Hospital, College of Medicine, Bangkok 10400, Thailand.
Phone: 08-3881-1666
E-mail: kittisak66@yahoo.com



Fig. 2 Multiple ill-defined and well-defined hypohyperpigmented sclerotic patches and plaques on both arms.

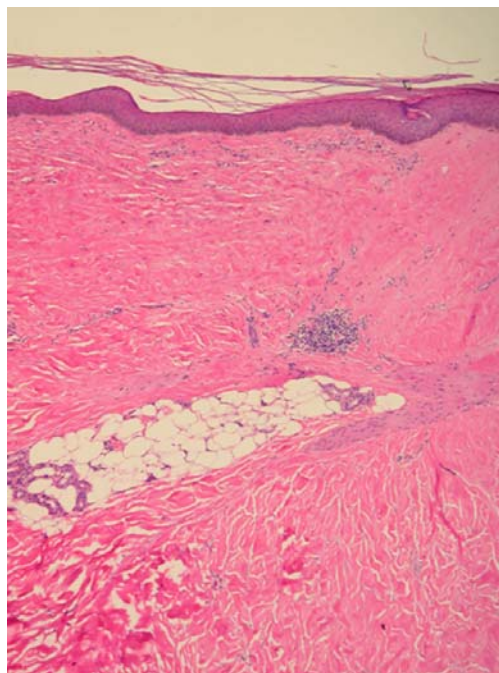


Fig. 3 The skin biopsy showed flattening of rete ridge of epidermis with sclerosis of collagen fiber in upper dermis

Laboratory investigation revealed normal complete blood count, liver function test, and blood chemistry. Anti-nuclear antibody (ANA) test showed homogeneous pattern at 1:80 titer. Anti-ds DNA, anti-Sm, anti-RNP/Sm, anti-Jo-1, anti-Ro, anti-Scl 70, anti-SSA and anti-SSB are all negative.

Discussion

Generalized morphea is a rare condition in which idiopathic sclerosis of the skin occurs in a widespread manner. The most frequent age of onset is between 30 and 40 years and the lesions start between the age of 11 and 50 years in 80% of patients. It displays widespread, well-circumscribed, indurated plaques. The plaques are usually ivory-colored and may show violaceous borders, especially when the disease is active. Other plaques may be hyperpigmented. Patients with generalized morphea do not have features of systemic sclerosis, but the disease may result in severe scars and functional disability⁽²⁾.

Diagnostic criteria of generalized morphea consists of 1) four or more lesions of morphea or linear type, 2) involvement of two or more areas of the body out of seven areas *i.e.*, head and neck, the right upper extremity, the left upper extremity, anterior trunk,

posterior trunk, right lower extremity and left lower extremity⁽¹⁾.

The cause of localized scleroderma is unknown but may involve an autoimmune etiology. Generalized morphea has been associated with polymyositis and sick sinus syndrome⁽⁹⁾ or necrotizing vasculitis⁽¹⁰⁾. Antinuclear antibodies, antihistone antibodies, and rheumatoid factor may be present. Furthermore, antibodies to single-stranded DNA (ssDNA) are seen in over 50 percent of cases. Eosinophilia may occur in localized scleroderma and indicates active illness⁽³⁾. *Borrelia burgdorferi*, a tick-borne spirochete, has been associated with both morphea and lichen sclerosis in Europe and Japan⁽⁴⁾. Unlike localized morphea, lesions of generalized morphea show less frequent spontaneous involution. Our case fits the criteria as followed, 1) 6 lesions of morphea, 2) involvement of 3 body areas, 3) laboratory result are negative.

The treatment of generalized morphea is challenging. High-potency topical glucocorticoids may be applied locally, with their effect augmented by intralesional injections of triamcinolone. Systemic glucocorticoids, antimalarials, colchicine, and azathioprine are usually ineffective. D-penicillamine (2.0-

5.0 mg/kg) may halt the formation of new lesions and induce the softening of the older lesions⁽⁵⁾. Oral calcitriol (0.50-0.75 mg daily) may improve joint mobility and skin extensibility in adult patients with generalized morphea⁽⁶⁾. UVA1 (340-450 nm) phototherapy may also be helpful⁽⁷⁾. Oral methotrexate (15-25 mg per week) may provide some benefit to a subset of patients⁽⁷⁾. Finally, infliximab showed significant reduction in cutaneous sclerosis and dyschromia⁽¹⁾.

Some improvement is usually seen in the course of 3-5 years, but the disease may last for many years. In most cases, the skin slowly softens and the pigmentation decreases. In time, the tendency to ulcerate with trauma and blistering decreases. Although some patients may be severely disabled by the immobility associated with the sclerotic changes and contractures, others with widespread sclerosis may remain surprisingly active. Patients usually remain in good health⁽¹¹⁾.

Our patient is concerned about the progressive disability and cosmetic problem, She has excellent response with topical calcipotriol. Patient education is also very important.

In conclusion, we report a rare and typical case of generalized morphea that presented with progressive multiple hypopigmented lesions on the extremities.

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รายงานผู้ป่วย Generalized morphea

กิตติศักดิ์ พยัฆาพงศ์, กอบกุล อุณหโชค

Generalized morphea จัดอยู่ในกลุ่ม *localized scleroderma* ซึ่งเป็นโรคที่มีรอยโรคผิวหนังแข็งที่กระจายไปตามที่ต่างส่วนต่าง ๆ ของร่างกายโดยไม่พบความผิดปกติของระบบอื่น ๆ ผู้เขียนรายงานผู้ป่วยหญิงอายุ 37 ปี ซึ่งมีรอยโรคสีขาวซึ่งเป็นปื้นแข็งบริเวณ แขนขวา และขาขวาโดยผลตรวจทางพยาธิวิทยายืนยันการวินิจฉัยโรคหนังแข็ง และผู้ป่วยตอบสนองดีต่อการใช้ยาทา calcipotriol