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Case Report

The Typical Cutaneous Presentation of Scleroderma

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ABSTRACT

Introduction

Scleroderma or systemic sclerosis (SSc) is an autoimmune disease characterized by autoimmunologic processes, vascular damage, inflammation, and widespread activation of fibroblasts, in which the skin, esophagus, lung, heart, and kidneys are most likely to be impacted as the disease progresses. The pathophysiology and etiology of SSc are intricated and multifaceted. Numerous cell types, including fibroblasts, lymphocytic cells, endothelial cells, and epithelial cells, are typically involved.

Case Presentation

In this context, we present a case report of a 52-year-old Cambodian male patient who presented with the typical cutaneous presentation of scleroderma. Laboratory investigations revealed the presence of anti-Scl-70 antibodies, which are associated with severe systemic sclerosis. A potent topical steroid, antihypertensive medication, and anti-inflammatory agent were prescribed to control his symptoms. A three-month follow-up is mandatory to monitor the extent of organ involvement. This typical cutaneous presentation can serve as a crucial hallmark for physicians, allowing for an instant diagnosis through visual inspection.

Conclusion

Scleroderma is an uncommon autoimmune condition affecting more females than males. Although early signs of scleroderma are rare, they should not be overlooked. Systemic organ involvement should be strictly monitored so that management can be more effective. There has not yet been a curative treatment for this disease. However, the management of organ complications has made significant progress.

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Introduction

Scleroderma or systemic sclerosis (SSc) is an autoimmune disease characterized by autoimmunologic processes, vascular damage, inflammation, and widespread activation of fibroblasts, in which the skin, esophagus, lungs, heart, and kidneys are most likely to be impacted as the disease progresses [1]. This disease tends to be common in women, with a female-to-male ratio between 3:1 and 14:1, whereas male black patients tend to have an earlier onset than white patients do [2]. The age of onset of the disease is between 30 and 50 years in the United States [3]. Scleroderma is an uncommon disease that affects 0.6--16 people per million people and increases in prevalence from 2--233 cases per million people annually [2–5].

Skin involvement can be distinguished into two clinical subtypes, limited cutaneous systemic sclerosis (LcSSc) and diffuse cutaneous systemic sclerosis (dcSSc), according to Bolognia et al. [6]. LcSSc is characterized by fibrotic skin alterations that are restricted to the fingers, hands, and face. In contrast, dcSSc is characterized by global cutaneous sclerosis that often starts in the fingers and hands and gradually spreads to the forearms, arms, face, trunk, and lower extremities [6]. The purpose of this report is to draw attention to specific skin signs that can provide clues for the clinical diagnosis of scleroderma.

Case presentation

A 52-year-old male Cambodian patient presented with multiple hypo- and hyperpigmented patches on his forehead and chest wall for one year. Moreover, he also reported the feeling of tenderness and movement restrictions upon his daily activities and mild dyspnea upon exertion. Neither dysphagia nor joint pain was reported, yet pallor of several digits during cold baths, called the Raynaud phenomenon (RP), was mentioned.The cutaneous findings included generalized skin hardening, thickening, shiny skin, forehead dyschromatosis, a mask-like facial appearance **(Figure 1)**, sclerodactyly, and a salt and pepper appearance on his chest wall **(Figure 2)**.

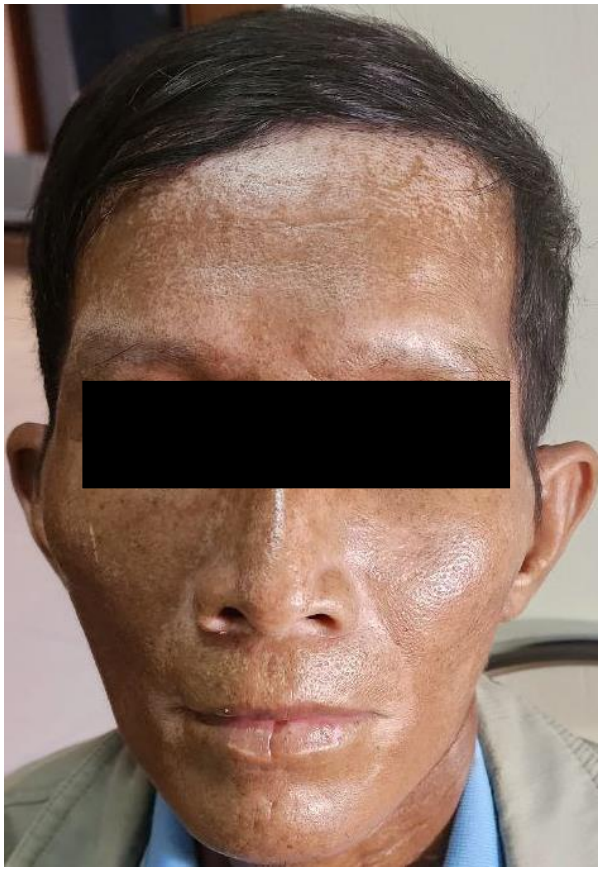

Figure 1. *Forehead dyschromatosis and a mask-like facial appearance are typical cutaneous presentations in scleroderma patients.*

The patient was further sent for laboratory investigations, including chest X-ray, fluorescent anti-nuclear antibody (FANA), anti-centromere antibody, anti-Scl-70 antibody, complete blood count, liver function test, blood urea nitrogen, and creatinine tests. FANA was positive, with a 1:2560 homogenous titer. Additionally, anti-Scl-70 antibodies were also positive, indicating severe systemic sclerosis with an increased risk of developing interstitial lung disease. The negativity of anti-centromere antibodies suggests that the patient does not belong to the LcSSc subtype. Overall, the physical examination and existing laboratory results were consistent with the cutaneous diffuse form of scleroderma.

Furthermore, the patient was advised to undergo physical therapy and exercise to improve joint mobility and muscle strength. A potent topical steroid (Clobetasol propionate 0.05%) was prescribed daily to treat skin hardening. Amlodipine 10 mg was administered daily to control RP. The patient was also advised to avoid smoking and cold temperatures to prevent worsening RP. Finally, 0.6 mg of colchicine was given daily to reduce inflammation and fibrous tissue. We will perform follow-up every three months during the first year of treatment.

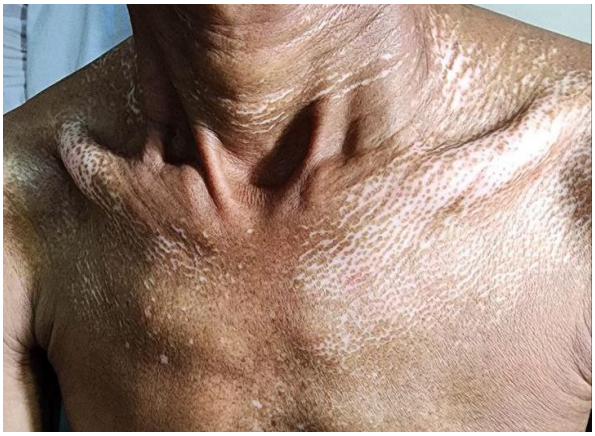

Figure 2. The appearance of salt and pepper is another typical cutaneous presentation in scleroderma patients

Discussion

The patient presented with the context of unspecified contact dermatitis, yet on typical cutaneous examination, we found generalized skin hardening, dyschromatosis, and a salt and pepper appearance, suggesting scleroderma. The positivity of FANA and anti-Scl-70 antibodies helped us confirm the diagnosis, which is consistent with the literature. Although no formal diagnostic criteria have been developed, the determination of the correct subset of this disease, including SSc overlap syndrome, is crucial [1].

The age group of SSc patients in Thailand commonly ranges from 50--69 years, which is consistent with our case report in Cambodia [7]. The complications of scleroderma include interstitial lung disease, pulmonary artery hypertension, renal crisis, hypertension, fibrosis, cardiomyopathy, heart failure secondary to pulmonary artery hypertension (PAH), and esophageal dysmotility [6].

The differential diagnosis of scleroderma-like syndrome should not be overlooked. These include circumscription scleroderma, eosinophilic fasciitis, sclerodermiform, genodermatoses, nephrogenic fibrosing dermopathy, and scleroderma-like lesions in malignancies [1].

The limitation of this case is that further investigations are needed to evaluate the patient's systemic organ involvement. Histopathological analysis of this patient has not yet been performed because of the limited availability of laboratory resources. A high-resolution CT scan is required for this case with the purpose of evaluating whether the patient has pulmonary fibrosis. Echocardiography is also needed to determine the patient's cardiac involvement. Genetic susceptibility tests might play a role in determining disease associations.

Conclusion

Scleroderma is an uncommon autoimmune condition affecting more females than males. Although early signs of scleroderma are rare, they should not be overlooked. Systemic organ involvement should be strictly monitored so that management can be more effective. There has not yet been a curative treatment for this disease. However, the management of organ complications has made significant progress. The ten-year survival rate is approximately 70%; therefore, health and psychological education are indispensable for patients. Encouragement is strongly needed so that the patient can adhere to the treatment regimen.

Acknowledgment

We greatly appreciate Dr. Savoeurn Pok for collecting documents and arranging the appointments for this case report.

Contribution

Dr. Chendavatey Pok and Dr. Savoeurn Pok were responsible for the writing of this manuscript.

Declaration

There are no conflicts of interest. Informed consent was obtained from the patient.

Abbreviations

SSc: Systemic sclerosis, LcSSc: limited cutaneous systemic sclerosis, dcSSc: diffused cutaneous systemic sclerosis, FANA: fluorescence antinuclear antibodies, PAH: pulmonary artery hypertension, RP: Raynaud phenomenon.

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