Scleroderma: A Case Report

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ABSTRACT
The concept of scleroderma as progressive systemic sclerosis was first described in detail in 1945. This article reports a case of systemic sclerosis. A 33-year-old woman complained of hand joints stiffness and skin lesion since 3 years ago. She also experienced tightened mouth with difficulty to swallow (dysphagia). Physical examination revealed a diffuse fibrosis of skin and internal organ (lung, esophagus). ANA test and CRP were positive. The patient was given symptomatic and supportive treatments focusing on the organ systems involved.

Key words: scleroderma, systemic sclerosis, dysphagia

INTRODUCTION
Scleroderma is derived from the Greek words skleros (hard or indurated) and derma (skin). Hippocrates first described this condition as thickened skin, scleroderma. Scleroderma is a rare disease; it usually presents between the ages of 35 and 55, with an up to 8-fold female excess. The prevalence of scleroderma is estimated to be between 30 and 130/million; the wide variation is due to the lack of population studies, as the disease is rare. There is some evidence suggesting that the disease has a higher incidence in black African populations. So far only a weak association between HLA and scleroderma has been found; stronger links have been found with specific autoantibodies (anti-topoisomerase and anti-centromere antibodies). A number of environmental triggers are thought to be risk factors for the disease. Exposure to silica dust (stone masons and gold miners) has been linked with the disease.1,2

In 1945, Robert H. Goetz first described in detail the concept of scleroderma as a systemic disease; he introduced the term progressive systemic sclerosis. The term systemic sclerosis is used to describe a systemic disease characterized by skin induration and thickening accompanied by various degrees of tissue fibrosis and chronic inflammatory infiltration in numerous visceral organs, prominent fibroproliferative vasculopathy, and humoral and cellular immune alterations.1

Scleroderma in childhood is rare and heterogeneous; and subtypes are determined by the type and number of lesions, the area of involvement and serological abnormalities. Localized scleroderma is the most common and can present at any age, with appearance of a patch of abnormal skin; if untreated, generally follows a course of active expanding disease, fibrosis and eventual softening with some "remission". The functional and cosmetic impact can be profound, as the lesions may interfere with growth of a limb and subcutaneous tissues (of the face or a limb).3

Systemic scleroderma includes progressive diffuse fibrous changes of the skin and fibrous changes involving internal organs — most commonly lungs, gastrointestinal tract, heart and kidneys — with significant mortality. Systemic scleroderma is slowly progressive, has a guarded prognosis and requires potent immunosuppression (corticosteroid and methotrexate) to control disease and limit severe disfigurement and disability, although clinical trials are lacking to guide practice.3

The American College of Rheumatology (ACR) criteria for the classification of systemic sclerosis require one major criterion or two minor criteria4,4.

Major criteria:
Proximal scleroderma characterized by symmetric thickening, tightening, and induration of the skin of fingers and that is proximal to the metacarpophalangeal or metatarsophalangeal joints. These changes may affect the entire extremity, face, neck, and trunk (thorax and abdomen). Skin in the face tightened, with a characteristic beak-like facies and pautcity of wrinkles. Sclerodactyly with digital ulceration, loss of skin creases, joint contractions, and sparse hair.
Figure 1 Masklike facies with stretched, shiny skin and loss of normal facial lines giving a younger appearance than actual age; hair and eyebrows are dyed black, and the nose are sharp, beak-like. Thinning of lips and perioral sclerosis result in a small mouth (microstomia), which is asymmetric; creating a snarling appearance. Sclerosis (whitish, glistening areas) and multiple telangiectases (not visible at this magnification) are also present.

Figure 2 Hypopigmentation caused by diffuse cutaneous scleroderma. Widespread thickening of skin, including truncal involvement, with areas of increased pigmentation and depigmentation.

Figure 3 Hand contractures in severe, long-standing diffuse systemic sclerosis. Both hyperpigmentation and hypopigmentation secondary to scleroderma. The “tanned” skin is actually hyperpigmentation secondary to scleroderma. The hypopigmentation over the metacarpophalangeal joints is also a result of skin inflammation.

Minor criteria:
º Sclerodactyly characterized by thickening, induration, and tightening of the skin, limited to only the fingers.
º Digital pitting scars or loss of substance from the finger pad. Depressed areas of the fingertips or a loss of digital pad tissue occurs as a result of ischemia.
º Bi-basilar pulmonary fibrosis includes a bilateral reticular pattern of linear or lineonodular densities, most pronounced in basilar portions of the lungs on standard chest roentgenography. These densities may assume the appearance of diffuse mottling or a honeycomb lung and are not attributable to primary lung disease.

CASE REPORT
A 33-year old woman came to the Outpatient Internal Department on April 19, 2011 with chief complaint of stiffness of hand joints and skin lesions for 3 years. She experienced whitish skin lesions that gradually blackened and became tightened. She also experienced tightened of her mouth causing difficulty to swallow. There were no history of corrosive liquid consumption and vomitus after eating.

On physical examination, the patient was fully alert with normal blood pressure and no sign of fever. Abnormalities were found in her face, neck, chest and superior extremities (figs. 1,2,3). Complete blood count, liver function test and renal function test were within normal limits, ANA test: 153 (<20), CRP: positive. Radiological examination of her hand revealed deformities of medial phalangeal and contracture of distal phalangeal of digit I-V. Density and trabeculation of the bone were...
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Scleroderma (systemic sclerosis), a rare chronic disorder characterized by diffuse fibrosis of the skin and internal organs. Causes of scleroderma are not known, but autoimmunity, endothelial cell damage, and increased production of extracellular matrix appear to play a key role in the pathogenesis of scleroderma.

Treatment is symptomatic and supportive, focusing on the organ systems involved. There is no effective therapy for the underlying disease process. However, management of specific organ manifestations of this disease have improved substantially.

REFERENCES