SCLERODERMA SECONDARY TO SILICA EXPOSURE - A CASE REPORT

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ABSTRACT

In recent years there have been many reports of connective tissue diseases especially scleroderma following exposure to silica and silicone. We report a 51-year-old Chinese who developed a scleroderma-like disease and pulmonary silicosis eight years after exposure to silica. To our knowledge, this is the first case to be reported in Malaysia.

Keywords: *scleroderma, pulmonary silicosis, silica*

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INTRODUCTION

Progressive systemic sclerosis is a multisystem disease characterised by inflammation and degeneration of the integument, heart, lungs, kidneys, gastrointestinal tract and synovia. Its aetiology has not been clearly elucidated but the outstanding feature of the disease is the overproduction of qualitatively normal collagen in the affected organs. Occupational exposures to vinyl chloride monomer, silica dust, epoxy resin benzene and chlorinated and aliphatic solvents have been implicated as potential causes of the disease[1]. We describe here a 51-year-old Chinese man who developed a scleroderma-like disease and pulmonary silicosis eight years after exposure to silica.

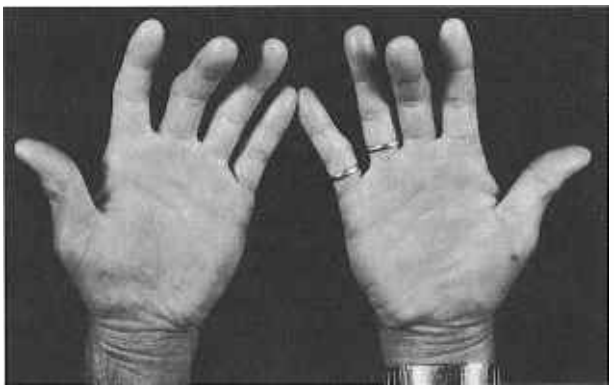
CASE REPORT

A 51-year-old Chinese man was referred to us for progressive tightening of the skin of his hands, face and perioral region over the last 5 years. He also had mild difficulty in swallowing both solids and liquids. On direct questioning it was found that he also suffered from a dry cough and decreased effort tolerance on exertion. He denied any history of loss of weight or appetite.

He had worked in a limestone quarry from the age of 18. Five years later he was employed a labourer in a silica factory. He was there for 10 years. For the last 5 years he had worked as a welder in a machinery factory prior to his retirement 3 years ago. There was no significant past medical and family illness. He was a smoker.

On examination he was well built with mild kyphosis. He had microstomia. The skin around the forearm and hands appeared shiny and thickened (Fig 1). There was flexion deformity of the proximal interphalangeal joint of the middle fingers and sclerodactyly was seen in both hands. He also had inducible Raynaud's phenomenon. There was no evidence of calcinosis, telangiectasia or vasculitis.

Fig 1 – Photograph of hands showing tight waxy skin with deformities



Examination of the chest showed decreased breath sounds over the left without any adventitious sounds. Chest radiograph showed evidence of pleural thickening and nodular shadows at the bases with egg shell calcification of the hilar lymph nodes consistent with pulmonary silicosis (Fig 2). Skin biopsy showed scanty subcutaneous tissue with marked collagenization of the dermis and atrophic skin appendages consistent with scleroderma (Fig 3).

A barium swallow examination showed hypomotility of the oesophagus and the presence of a sliding hiatus hernia. Pulmonary function tests revealed a restrictive pattern. Collagen screen including antinuclear and rheumatoid factors were both negative. Urinalysis, renal and liver profiles were normal.

DISCUSSION

Systemic sclerosis is characterised by thickening and fibrosis of the skin (scleroderma) and by distinctive forms of internal organ involvement. The prevalence rate is between 4 to 12 individuals per million population per year[2]. The pathogenesis of the disease remains unknown but dermal fibroblasts from involved skin accumulate Type I, III and IV collagen, fibronectin and glycosaminoglycan at an increased rate. Elevated levels of interleukin-2 and soluble interleukin-2 receptors are present in systemic sclerosis and are linked to clinical progression of the disease suggesting that lymphokines either stimulate collagen production or other cells such as monocytes and mast cells to release factors that in turn induce collagen biosynthesis in fibroblasts[3].

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Fig 2 - Chest radiograph showing pleural thickening and nodular shadows at the bases with egg shell calcification of the hilar nodes

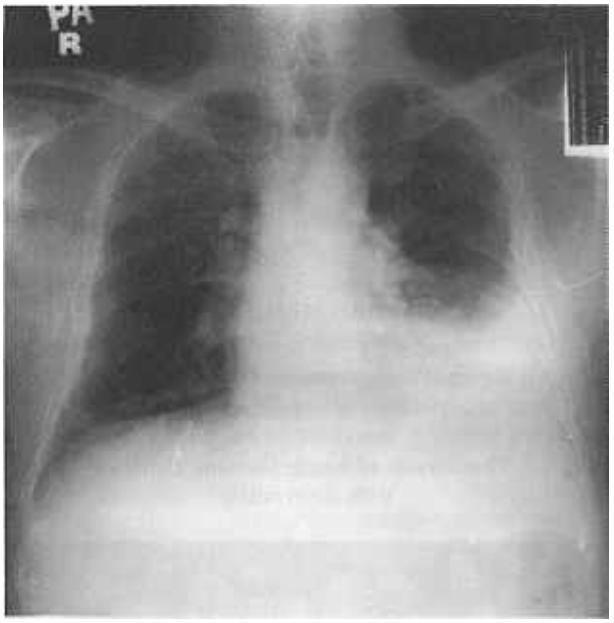
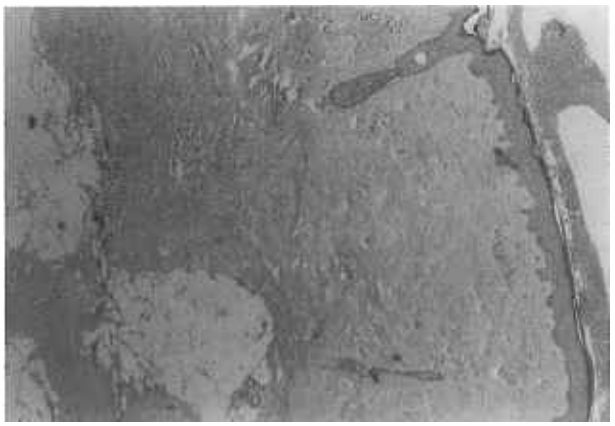


Fig 3 - Skin biopsy showing marked collagenization of dermis with atrophic skin appendages



Silicosis is the fibrotic disease in the lung due to prolonged inhalation of silica dust (silicone dioxide) and has been recognised as an occupational hazard in quarrying, masonry, mining and sand blasting. Lung macrophages exposed to silica elaborate factors that cause chronic inflammation and fibroblastic proliferation resulting in progressive fibrosis. Exposure to silica has also been reported to cause scleroderma-like disease[4,5]. The mean exposure time was 14.5 years (range 4-33 years) and the interval between the beginning of exposure and the onset of scleroderma averaged 24.4 years (range 4-45 years). Our patient was exposed to silica for at least 15 years, working in a limestone quarry and a silica factory. The role of silica in the origin and pathogenesis of scleroderma remains controversial. Silica is known to be a highly immunogenic substance and can induce an autoimmune response leading to the development of connective tissue diseases[6]. Silicone (the reduced form of silica reacted with methylchloride and polymerised to stable polydimethylsiloxane) has been used in reconstructive surgery and augmentation mammoplasty for years because it is considered to be chemically inert. Unfortunately, recent studies have shown that women with silicone breast implants have developed a wide range of rheumatic diseases especially scleroderma[7-9], although others have questioned such an association[10,11]. In view of the large numbers of women with silicone breast implants, it is important to remind clinicians to inquire of every female patient with scleroderma (and other connective tissue diseases) whether mammoplasty has been performed. Removal of the implants may lead to resolution of the disease.

Silica induced scleroderma cannot be distinguished from the idiopathic form by epidemiological, clinical, immunological or by parameters referring to the blood vessels or collagen metabolism. Rustin et al[12] have studied 17 coal miners exposed to silica dust who subsequently developed a scleroderma-like disease. They concluded that these patients have clinical, immunological and serological features that were indistinguishable from the idiopathic form of the disease although as a whole, the former group had a higher prevalence of pulmonary involvement and anti Scl-70 antibody. Antinuclear antibodies (typically nucleolar pattern) were detected in both groups in around 90%, rheumatoid factor in 30%, polyclonal hypergammaglobinaemia in 20% and cryoglobinaemia in 40%. The prognosis in both forms of disease were similar.

Treatment of scleroderma includes measure to improve the peripheral blood flow and to reduce inflammation and fibrosis in the affected organs. Our patient was advised to protect his hands and feet against cold exposure and to avoid smoking. Antifibrotic drugs like penicillamine and colchicine have been used but their efficacies remain unproven.

In view of the clear association of silica to disease, careful history taking of previous exposure is important in any patient suffering from connective tissue disease especially scleroderma.

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