A 21-year-old man with cough and skin thickening

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CASE SUMMARY

A 21-year-old male presented with complaints of dry cough and shortness of breath for 5 years. The patient also noticed diffuse depigmentation and hyperpigmentation of the skin for 3 years associated with tightness of skin involving the face, hands, and legs which progressed to involve proximal limbs and trunk for the last 1 year. There was a history of Raynaud's phenomenon and difficulty in swallowing. He also complained of polyarthralgias involving the small joints of both hands for the last 1 year. There was no history of oral ulcers and proximal muscle weakness. He had been a lifetime never smoker. The patient had worked as stone cutter for the last 7 years and had significant exposure to silica dust. There was no past history of tuberculosis and no family history of autoimmune disease.

On examination, diffuse cutaneous thickening, sclerodactyly, and "salt and pepper" pigmentary changes were present [Figure 1]. Chest examination revealed bilateral end-inspiratory fine crepitations. Computed tomography scan of the thorax demonstrated mediastinal lymphadenopathy with eggshell calcification and bilateral upper lobe fibrosis [Figure 2]. Pulmonary function test revealed a restrictive defect with forced vital capacity of 56% predicted. Six-minute walk distance was 430 m, and saturation dropped from 92% to 86% at the end of the test. Antinuclear antibody titers were positive.

QUESTION

What is your diagnosis?

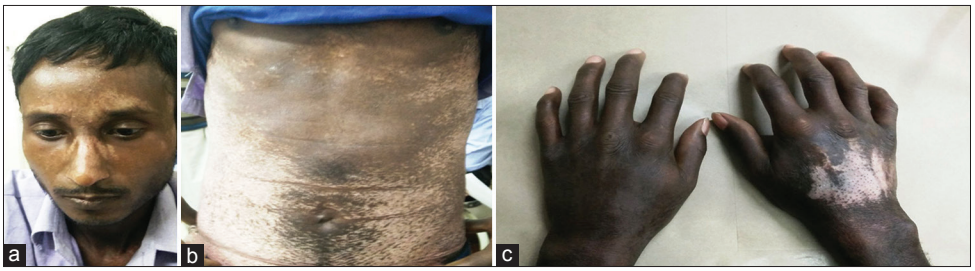

Figure 1: Clinical photograph demonstrating diffuse pigmentary changes (salt and pepper type) and thickening of skin of the face (a), trunk (b), and hands along with sclerodactyly (c)

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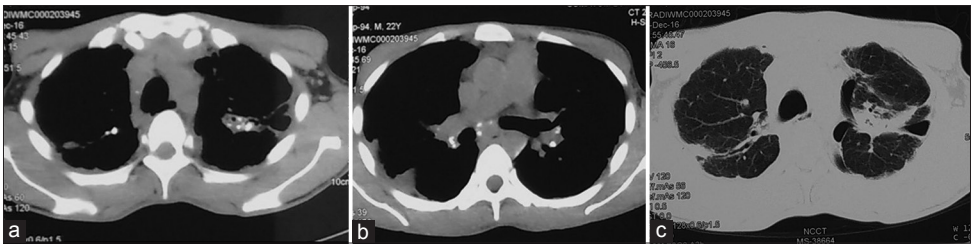

Figure 2: Computed tomography scan of the thorax demonstrating bilateral areas of upper lobe parenchymal calcification (a) with mediastinal lymph node calcification (b). Extensive fibrotic changes in both upper lobes are also noted (c)

ANSWER

Erasmus syndrome.

In view of significant silica exposure, skin changes suggestive of systemic sclerosis, and radiological findings consistent with silicosis, the diagnosis is Erasmus syndrome.

Systemic sclerosis is a multisystem disorder of unknown etiology. It is characterized by three main manifestations: organ-specific autoantibodies, end-organ fibrosis, and small vessel vasculopathy. Environmental and occupational exposures such as vinyl chloride, epoxy benzene, and silica have been associated with causation of systemic sclerosis. Erasmus syndrome describes the occurrence of systemic sclerosis in individuals with silica exposure with or without silicosis.[1] Silica exposure is one of the strongest recognized risk factors for the development of systemic sclerosis (relative risk 3.2).[2] Erasmus had described a high prevalence of systemic sclerosis in Witwatersrand gold miners exposed to dust containing free silica in 1957.[3] The first case of Erasmus syndrome from India was reported by Khanna *et al*. in 1997.[4] Since then, there have been a few case reports published about the disease.[5,6] Coexistence of pulmonary tuberculosis and Erasmus syndrome has also been reported.[7]

The mechanism of enhanced autoimmune response in patients with silica exposure remains unclear. There is depressed cellular immunity, and adjuvant effect of silica particles on antibody production has been postulated to cause autoimmune reaction. Silica exposure leads to increased lymphokine production by pulmonary macrophages leading to increased collagen production and chronic inflammation. The risk of disease is more in males exposed to silica as compared to females.[2] The risk is higher in miners as compared to nonminers with silica exposure, likely due to less level of exposure.[2] The clinical features of silica-associated systemic sclerosis are similar to idiopathic systemic sclerosis, and the diagnosis is based on exposure history with or without radiological findings consistent with silicosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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