CASE REPORT

Erasmus Syndrome—A Rare Cause of Diffuse Cutaneous Systemic Sclerosis due to Silica Exposure: A Case Report

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Abstract

Erasmus syndrome is a rare disease characterized by the development of systemic sclerosis (SSc) in the background of silica exposure or silicosis. Here we report a case of a 42-year-old male who presented with skin tightening for 2 years and progressive dyspnea for 1 year. Patient worked as a sandblaster for 6 months. On examination, there was sclerodactyly, fixed flexion deformity of the lower limbs, diffuse skin tightening and salt and pepper rashes on the chest wall and upper back and crepitations on the chest auscultation. Chest imaging and pulmonary function tests were suggestive of interstitial lung disease. The autoimmune profile showed Scl-70 +++. The diagnosis of erasmus syndrome was made based on specific clinical presentations and characteristic chest imaging findings in a male patient with a history of silica exposure.

Keywords: Case report, Erasmus syndrome, Interstitial, Salt and pepper rash, Sclerodactyly, Silicosis, Systemic sclerosis.

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Introduction

Silicosis, falling under the category of occupational lung diseases, is a preventable fibrosis lung disease resulting from the inhalation of fine crystalline silica particles.[1,2]

One of the most prevalent minerals in the crust of the planet is silica, which can be found in nature in both crystalline and amorphous forms.[3] Silica, when inhaled in its crystalline form, is the most common form linked to the development of occupational lung diseases.[2–4] The chance of developing silicosis is significantly increased by any employment that disturbs the earth's crust, such as processing rock that contains silica. Sandblasting, packing silica flour, mining, stone-cutting, and granite quarrying are among the main environmental exposures.[5,6] As silica-containing dust particles are inhaled, these foreign micro particles in the lungs cause a series of inflammatory events, including the activation of macrophages, the release of inflammatory cytokines, the production of free radicals, and the up regulation of cell-signaling pathways. These events culminate in the emergence of symptoms like cough, dyspnea, and easy fatigability.[7]

Several systemic autoimmune disorders, in addition to silicosis, have been attributed to or found to have an association with silica exposure.[3] According to a review of the literature by Miller et al., there is epidemiologic evidence linking silica inhalation to the onset of Wegener's granulomatosis, rheumatoid arthritis, systemic lupus erythematosus, primary systemic vasculitis, and systemic sclerosis (SSc).[8]

Erasmus Syndrome is a rare disease phenomenon identified by the onset of SSc in the context of silica exposure or silicosis.

Case Description

A 42-year-old euglycemic, normotensive, hypothyroid, non-smoker and habitual tobacco chewer male presented with a history of skin tightening, sclerodactyly, and fixed flexion deformity of the both lower limb for the past 2 years, with symptoms aggravating over the last 6 months. He also complained of gastroesophageal reflux disease (GERD) symptoms, dyspnea on exertion, and a dry cough for the last one year. No Raynaud's phenomenon was reported. He had no history of hemoptysis, fever, or chest pain. The past history of TB was denied. He has been a farmer by profession for the last 12 years, and he had also worked as a sandblaster for 6 months, some 12 years ago. He left the job when his brother succumbed to silicosis after working as a sand blaster for a significant period of time (6 years).

On examination, he had diffuse skin tightening, microstomia, sclerodactyly, and clubbing. Facies was masked with taut and shiny skin, loss of facial expression, paucity of normal skin wrinkling in the face and forehead, and areas of hyperpigmentation

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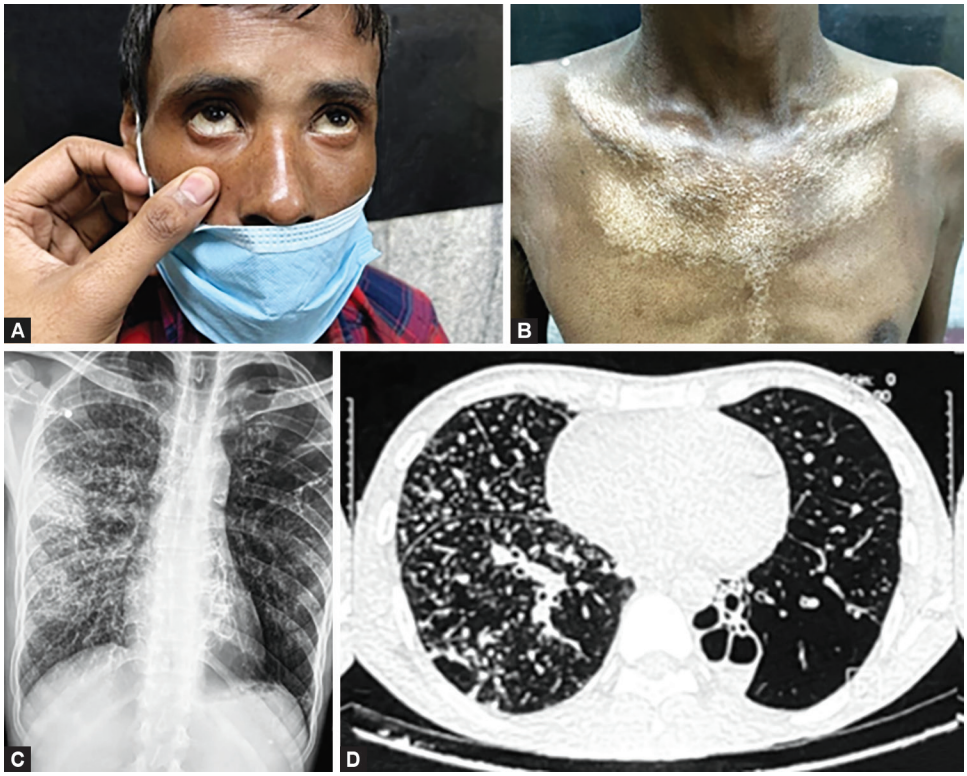
Patient consent statement: The authors have obtained written informed consent from the patient for publication of the case report details and related images.



Figs 1A to D: (A) Masked facies with skin tightening; (B) Salt and pepper rash on anterior chest wall; (C) Chest X-ray PA view image; (D) CT thorax image

and depigmentation (Fig. 1A). Mouth opening was small, with difficulty opening the mouth fully. Salt-pepper appearance was seen on the anterior part of the chest and neck, and upper back. Raynaud's phenomenon was absent. Fingers were puffy with taut, shiny skin (Fig. 1B). There was a moderate pallor without any significant lymphadenopathy. Crepitations were heard over the right infra-scapular area. A clinical diagnosis of diffuse SSc was made. Autoimmune serology came out to be positive for ANA (3+, nucleolar) with the anti scl-70 antibody (+++) and negative for the anticentromere antibody (–). C3 level was 121 mg/dL, and C4 level was 24 mg/dL. A complete hemogram showed normocytic normochromic anemia. Chest X-ray showed patchy opacities in right middle and lower zones with surrounding reticulonodular opacities, left lower lobe collapse superimposed ring shadows, likely bronchiectatic changes and fibrotic bands in left lung field and computed tomography (CT) thorax showed patchy areas of subpleural consolidation in right upper and middle lobes with cavitation, multiple centrilobular nodules in right lung fields (predominantly in upper and middle lobes), cicatricial atelectasis with tractional bronchiectasis noted in left lower lobe with compensatory hyperinflation of left upper lobe and multiple densely calcified hilar lymph nodes noted bilaterally as well as in the pre and paratracheal and subcarinal groups (Figs 1C and D). Sputum AFB and CB-NAAT reports were negative. Functional assessment was done, and it showed severe restriction with FEV1/FVC 100, FVC 0.71 (48%), and FEV1 0.71 (31%). Echocardiography showed no pulmonary artery hypertension (PAH). Nail-fold capillaroscopic examination was suggestive an of active scleroderma pattern. As a part of ruling out active tuberculosis disease, bronchoalveolar lavage (BAL) fluid GeneXpert, tuberculin skin test (TST), and interferon gamma release assay (IGRA) test done and all reports were negative. In view of SSc in a male patient with a history of silica exposure and characteristic CT, a diagnosis of Erasmus syndrome was made.

Discussion

Silicosis is the most prevalent type of pneumoconiosis, resulting from crystalline silica particles being inhaled and retained in the lungs, leading to persistent inflammation and the eventual emergence of lung fibrosis.[3,9,10]

On the basis of the rate of progression of symptoms since exposure, silicosis can be categorized as acute, accelerated, or chronic.[2] Pleuritic chest discomfort, dry cough, exertional breathlessness, weight loss and easy fatigability are some of the symptoms. Based on its pathological features, silicosis can also be categorized as diffuse interstitial fibrosis, progressive massive fibrosis, silico-proteinosis, or nodular.[2] Since its respiratory symptoms and imaging findings closely resemble TB, silicosis can easily be misdiagnosed as TB, and hence it becomes imperative to rule out active pulmonary TB among patients with silica exposure. According to studies, exposure to silica may increase the risk of developing tuberculosis.[11–13]

Dr LD Erasmus, a South African physician, first documented the incidence of silica-associated SSc in 1957. According to Erasmus, 17 gold miners from Witwatersrand, South Africa, developed exertional dyspnea, skin changes, flexure contractures on the hands, Raynaud's phenomenon, and occasionally resorption of the terminal phalanges.[14] Of those seventeen miners, ten had no radiographic signs of silicosis, four had suspected silicosis, and three had definitive silicosis.[14] There have been more confirmed reports on the incidence of progressive SSc in people with substantial silica exposure since the release of Dr Erasmus' study. But it took ten years for silica exposure to be officially acknowledged as a predisposing

factor to SSc, which made it possible to eventually refer to this disorder as "erasmus syndrome".[15]

Eliminating silica dust exposure is the gold standard for managing erasmus syndrome. Treatment of silica-related SSc is identical to idiopathic forms of SSc. Organ-based symptomatic treatment is the standard method used to manage SSc.[16,17] For example, Raynaud's phenomenon is alleviated with phosphodiesterase-5 inhibitors and/or calcium-channel blockers, while short courses of non-steroidal anti-inflammatory medications are administered to people who have myalgia and/or arthralgia. Patients who have severe organ involvement and/or diffuse and escalating cutaneous involvement are treated with systemic immunosuppressive medications like methotrexate, mycophenolate mofetil, or cyclophosphamide.[17]

Rustin et al. found that there is no clinical, serological, or immunological difference between idiopathic SSc and Erasmus syndrome.[18] Recognizing the connection between silicosis, SSc, and silica exposure is extremely helpful for diagnosing and treating patients in the workplace.

Conclusion

Erasmus syndrome is a rare disease entity. Hence, patients with systemic signs of silicosis should undergo rigorous screening to rule out SSc. The key to diagnosing erasmus syndrome is keeping a high index of suspicion. For people who have been exposed to silica at work, prompt diagnosis and action can enhance their quality of life and avert potentially fatal complications. Therefore, it is very important to conduct an occupational hazard awareness program for workers and industrial governing authorities, and to encourage the use of personal protective equipment to prevent silica particle inhalation.

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Ethical Approval

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

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