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CASE REPORT

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Erasmus syndrome: A rare case report of silicosis and systemic sclerosis

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Abstract

People with silicosis may develop Erasmus syndrome, a condition characterized by the emergence of systemic sclerosis (SSc) after silica exposure. This case study emphasizes the significance of understanding the connection between occupational silica exposure, silicosis, and SSc. A 24-year-old male stonecutter got silicosis and a form of SSc following 8 years on his job as a stonecutter. The signs and symptoms the patient experienced were Raynaud's phenomenon, cutaneous fibrosis, arthralgia, digital pitting, and respiratory distress. High-resolution computed tomography (HRCT) revealed interstitial lung disease and calcified mediastinal lymph nodes. This case study demonstrates the clinical importance of the relationship between occupational silica exposure, silicosis, SSc, and Erasmus syndrome. Healthcare providers need to be aware of the possible difficulties and issues that may result from silica exposure. They should prioritize quick detection and efficient treatment plans for those who have been exposed to silica while on the job.

KEYWORDS

occupational lung disease, scleroderma, silicosis, systemic sclerosis

1 | INTRODUCTION

Erasmus syndrome is a rare condition characterized by the development of systemic sclerosis (SSc) following exposure to silica, with or without the presence of silicosis. The term was coined after the doctor who initially described several cases of SSc in gold mine workers in South Africa in 1957.[1] The significant risk of developing SSc in people exposed to silica has been suggested in the meta-analysis done by McCormic et al.[2] Prolonged contact with silica generates an inflammatory response that leads to polyclonal activation of T cells, its pathophysiology basis. In the long term, this stimulation produces self-reactive T-lymphocytes that are resistant to apoptosis. This chain of events eventually promotes the emergence and spread of autoimmune disorders such as SSc.[3]

Although Silicosis is not needed for developing Erasmus Syndrome, its presence increases the risk 24 times for developing SSc compared with the general population.[4] The prevalence of Erasmus syndrome among SSc patients is estimated to be only 0.9% based on a recent retrospective study of a large Brazilian cohort.[5] Inhaling crystalline silica particles found in stone, rock, sand, and clay may cause silicosis, a fibrotic lung disease.[6] Stone quarrying, mining, and

sandblasting are among the occupations linked to an elevated risk of silicosis.[7] Other professionals, including dental technicians, drillers, people who worked with quartz, and those exposed to abrasive powders, have also been reported to have Erasmus syndrome.[8] In addition to the well-known silicosis, silica exposure has been related to various autoimmune diseases. Miller et al.[9] comprehensively analyzed the literature and discovered epidemiological evidence linking silica inhalation to the beginning of several autoimmune disorders. Systemic lupus erythematosus, rheumatoid arthritis, primary systemic vasculitis, Wegener's granulomatosis, and SSc are examples of these disorders.[9] Anti-topoisomerase I antibodies are the most common autoantibodies in silica-induced SSc.[10] SSc has been associated with silica exposure, and a study of 14 people with the illness discovered that the majority (9 out of 14) had specific antibodies targeting topoisomerase I, whereas just one patient had anti-centromere antibodies. In a study done by Rustin et al. Erasmus syndrome is clinically, serologically, and immunologically not distinguishable from idiopathic SSc.[11] Understanding the relationship between silica exposure, silicosis, and SSc tremendously aids in diagnosing and treating individuals in the workplace.

In this study, we use the case of a male Bangladeshi stonecutter who worked for a local company for 8 years. SSc is a progressively worsening condition that can be debilitating over time. To the best of our knowledge, this might be the first recorded instance of Erasmus syndrome in Bangladesh.

2 | CASE REPORT

A 24-year-old stonecutter from Uttar Dinajpur, Bangladesh, presented to Rangpur Medical University's Internal Medicine Clinic. The patient's medical history included 3 months of occasional blue discoloration of the fingers and toes when exposed to low temperatures. Figure 1 shows the patient's left middle and little finger gangrenous ulcerations developed during the previous year due to progressive cutaneous constriction. The patient additionally reported increasing dyspnea with exercise, a mild nonproductive cough, polyarthralgia, and extensive cutaneous fibrosis. He had worked as a stonecutter for the previous 8 years, exposing him to significant quantities of silica dust on the job. He had no family history of such complaints and had no past encounters with tobacco, alcohol, high blood pressure, or diabetes.

Figure 2 depicts the patient's face skin, both hands, forearms, chest, and legs presenting cutaneous symptoms during the physical examination, characterized by tight and adherent skin. According to the new Rodnan score (21 out of 51), the patient exhibited considerable cutaneous symptoms, indicating serious skin involvement. Sclerodactyly,

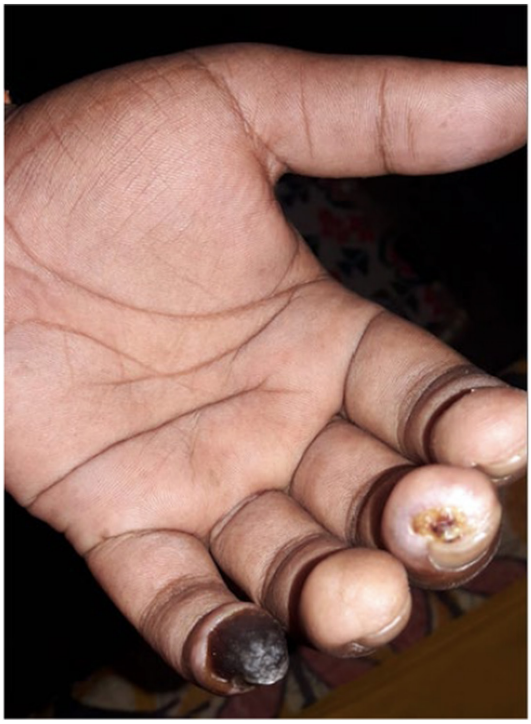

FIGURE 1 Showing ulcerative change in left middle and little finger.

a condition in which the skin of the fingers thickens and tightens, was seen in the patient on both hands. Raynaud's phenomenon was also present, as were pitting scars on the patient's fingers and longitudinal nail curvature. Furthermore, there were no furrows on the forehead.

Laboratory tests showed a hemoglobin level of 9.6 g/dL, an elevated erythrocyte sedimentation rate of 55 mm in the first hour, normal liver and renal function, and negative results for HIV, HBV, and HCV serology. The ANA profile revealed a high titer positive for topoisomerase I out of 17 antigens tested. Echocardiography revealed normal LV function but with Severe TR and pulmonary artery hypertension. Differential diagnosis of pulmonary tuberculosis (TB) was considered, and tests for TB were performed using GeneXpert and Interferon Gamma Release Assay, which yielded negative results. Spirometry showed moderately reduced diffusion capacity for carbon monoxide. On the chest CT scan, most of the lung segments, notably the basal portions, revealed increased density or sclerosis, as well as reticulonodular alterations and thicker interstitium (Figure 3). The patient's silicosis was designated as “p,” according to the International Labor Office standardized radiographic classification for silicosis grading. There were several calcified lymph nodes in the mediastinum. This study found radiological evidence for silicosis and clinical and serological evidence that points to SSc, which indicates a diagnosis of Erasmus syndrome.

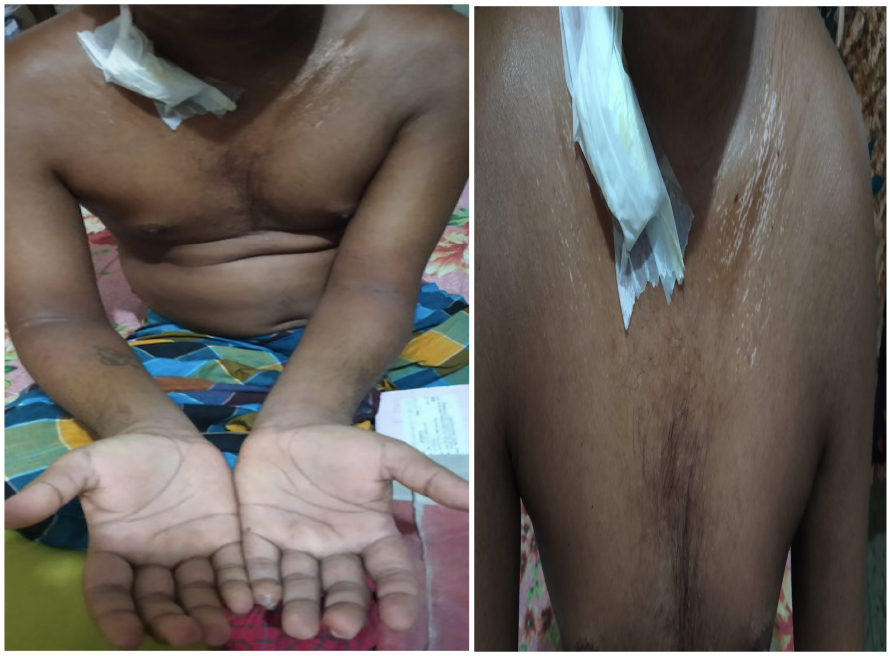

FIGURE 2 Showing skin tightening in the chest and limbs.

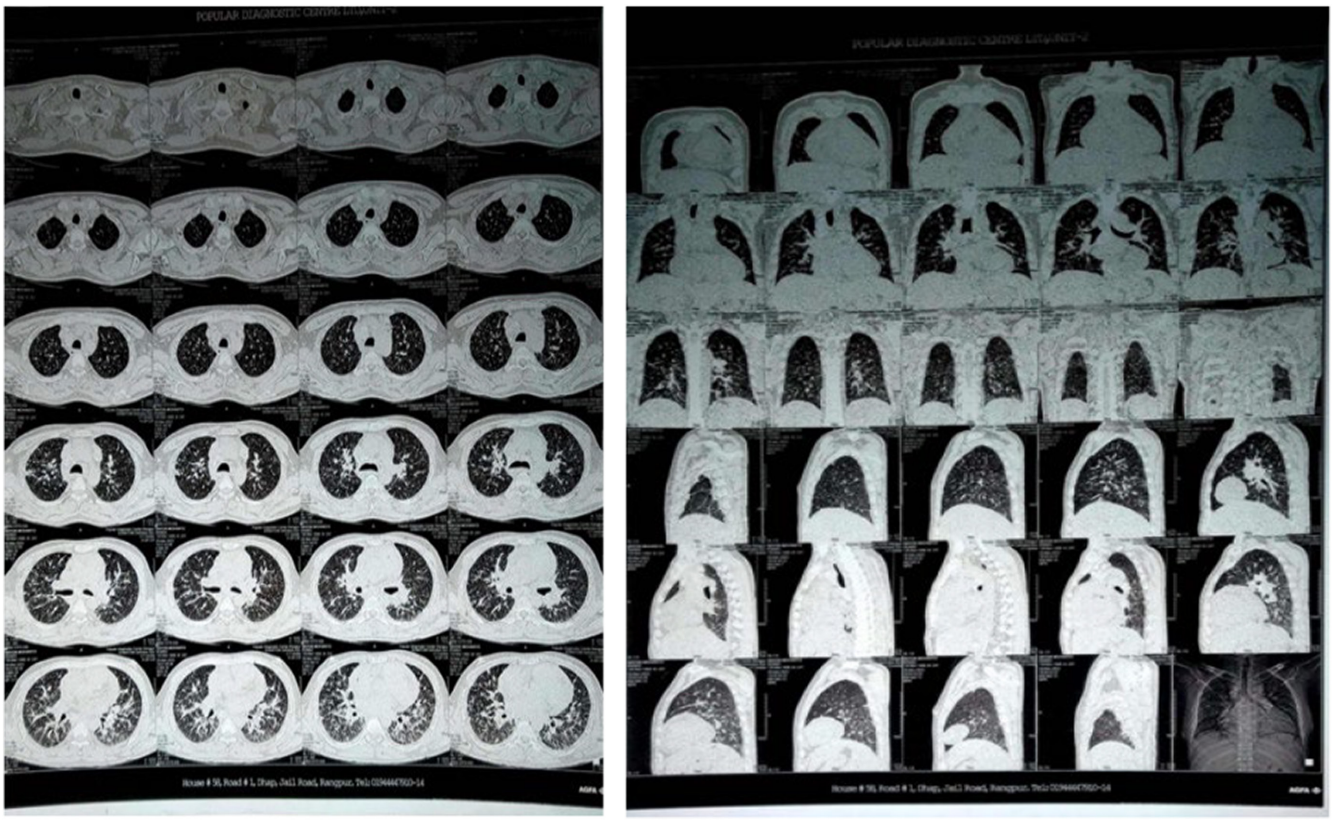

FIGURE 3 Computed tomography (CT) scan of the chest showing diffuse areas of increased density/sclerosis with reticulonodular changes and thickening of the interstitium.

He received monthly intravenous cyclophosphamide and oral prednisolone for 6 months for SSc. He was prescribed oral tadalafil 10 mg twice a day to address pulmonary artery hypertension. He also received care from a respiratory therapist and was referred for rehabilitation.

3 | DISCUSSION

SSc, silicosis, and Erasmus syndrome are all separate medical disorders with unique traits and effects. However, considering their potential linkages can display how these

elements interact and how this impacts people's health. SSc and other autoimmune illnesses have been linked to silica exposure as potential triggers.[12] The immune system can be stimulated by silica particles, which might contribute to an unusual immunological response and possibly cause the onset or worsening of autoimmune diseases. However, the precise processes and causal connections are still being researched.

SSc is a chronic autoimmune multisystem disease characterized by progressive fibrosis and widespread vascular dysfunction. Raynaud's phenomenon, telangiectasias, subcutaneous calcium deposits, cutaneous fibrosis, arthralgia, esophageal dysmotility, pulmonary hypertension, and interstitial lung disease are among the common signs.[13] Several environmental agents have been implicated in SSc based on case clustering or formal epidemiological studies. One of the earliest and strongest such associations with silica dust was discovered because of the high frequency of SSc among stone masons.[14] Gold miners in South Africa and coal miners in the United States have both been found to cluster similarly.[15] The case described here involves prolonged exposure to silica dust, as indicated by the individual's occupational history. Based on the current classification criteria, the diagnosis of SSc was confirmed.[16] Consequently, this case aligns precisely with the definition of Erasmus syndrome.

There is not a proven cure for Erasmus syndrome. Most treatments are symptomatic, such as nifedipine for Raynaud's, antiulcer medications for gastric reflux, and anti-inflammatory medications for arthralgia. The involvement of the lung, however, should be considered when treating Erasmus syndrome because it would necessitate immunosuppressive therapy such as cyclophosphamide, methotrexate, or mycophenolate mofetil.[5] Immunosuppressive medication started early on is thought to postpone lung damage and reduce mortality.[8]

Silicosis is an irreversible occupational lung disease caused by exposure to silica dust. Its grave and long-lasting consequences threaten workers in various industries, with symptoms often manifesting years after exposure.[17] Silicosis is induced by inhaling and subsequent pulmonary retention of crystalline silica. Individuals employed in mining, construction, sandblasting, and stone quarrying occupations exhibit a heightened susceptibility to the development of silicosis. Inhalation of minute crystalline particles induces inflammatory responses and fibrotic changes within the pulmonary tissue. The occurrence of inflammation can potentially cause lung fibrosis, resulting in a decline in pulmonary function. Silicosis has been associated with TB, immunological disorders, and chronic obstructive pulmonary disease (COPD).[18]

The prioritization of prevention strategies for silicosis and Erasmus syndrome is imperative. Occupational health and safety practices are critical for limiting silica exposure and preserving workers' health.[19] Employers are suggested to utilize engineering methods to decrease silica dust production and distribution, such as building effective ventilation systems and using wet dust suppression technology. Employees would benefit from personal protective equipment, such as respirators, as an extra layer of safety. To reduce silicosis instances, careful adherence to established standards for regularly monitoring occupational silica dust concentrations and the provision of substantial training programs focusing on hazard awareness and adopting safe work techniques may be considered.

One of the most essential components in resolving the silicosis issue would be enacting and enforcing rules and regulations. To maintain worker safety, governments, and regulatory agencies are advised to implement rigorous regulations on enterprises about allowed silica exposure levels in the workplace.[20] Implementing occupational health surveillance systems to monitor worker health and detect silicosis early might be critical. Access to good healthcare, particularly yearly medical exams, in a susceptible population may be beneficial for early detection and treatment of silicosis and associated illnesses such as Erasmus syndrome.

4 | CONCLUSION

In conclusion, this case report highlights the significance of acknowledging the connection between occupational silica exposure, silicosis, SSc, and Erasmus syndrome in clinical settings. Healthcare providers should be mindful of the potential complexities and complications arising from silica exposure and consider prompt diagnosis and effective management strategies for individuals exposed to silica in their work environments. SSc-like symptoms in silicosis should be treated promptly since lung involvement affects prognosis and requires the start of immunosuppressive medication. Further research is necessary to gain deeper insights into the underlying mechanisms and to develop more tailored therapeutic approaches for this distinctive syndrome.

AUTHOR CONTRIBUTIONS

Somee Rauniyar: Conceptualization; formal analysis; investigation; supervision; validation; writing – original draft; writing – review and editing. **Biyas Thapa:** Conceptualization; formal analysis; investigation; supervision; validation; writing – original draft; writing – review and editing. **Prakash Gupta:** Conceptualization; formal analysis; investigation; project administration; supervision; visualization; writing – original draft; writing – review and editing. **Rupak Subedi:** Conceptualization; formal

analysis; validation; writing – original draft; writing – review and editing. **Bishwodip Baral:** Conceptualization; formal analysis; validation; writing – original draft; writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

The authors have no conflict of interest to declare.

DATA AVAILABILITY STATEMENT

The data supporting this article's findings are available from the corresponding author upon reasonable request.

CONSENT

The written consent was obtained from the patient.

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