**Title :**Erasmus Syndrome: A Rare Case Report of Silicosis and Systemic Sclerosis

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Abstract:

Introduction: People with silicosis may develop Erasmus syndrome, a condition characterized by the emergence of systemic sclerosis after silica exposure. This case study emphasizes the significance of understanding the connection between occupational silica exposure, silicosis, and systemic sclerosis.

Case Report: A 24-year-old male stonecutter got silicosis and a form of systemic sclerosis following 8 years on his job. Among the signs and symptoms that the patient experienced were cutaneous fibrosis, arthralgia, Raynaud's phenomenon, digital pitting, and respiratory distress. High-resolution computed tomography (HRCT) proved beneficial in detecting ILD and locating calcified mediastinal lymph nodes. This case study emphasizes the importance of understanding the connection between occupational silica exposure, silicosis, and systemic sclerosis.

Conclusion: This case study demonstrates the clinical importance of the relationship between occupational silica exposure, silicosis, systemic sclerosis, and Erasmus syndrome. The Erasmus syndrome (or silicosis), an occupational lung ailment, has been related to the emergence of scleroderma, particularly systemic sclerosis.

Keywords: Silicosis, scleroderma, systemic sclerosis, occupational lung disease.

Introduction:

When systemic sclerosis develops following interaction with silica, whether or not silicosis is present, Erasmus syndrome may be diagnosed. A physician coined the term "systemic sclerosis" in 1957 (1) to describe a disease he encountered often among South African gold mine workers. McCormic et al. (2) provide substantial evidence for a relationship between silica exposure and the start of systemic sclerosis in a meta-analysis. Prolonged contact with silica generates an inflammatory response that leads to polyclonal activation of T cells in the context of pathology. This stimulation, in the long term, leads to the production of self-reactive T-lymphocytes that are resistant to apoptosis. This chain of events eventually promotes the emergence and spread of autoimmune disorders such as systemic sclerosis (3).

Erasmus Syndrome may occur in the absence of silica, however exposure to silica raises the likelihood of developing systemic sclerosis by a factor of 24 when compared to the general population. Inhaling crystalline silica particles, which are found in stone, rock, sand, and clay, may cause silicosis, a fibrotic lung disease (5). Stone quarrying, mining, and sandblasting are among occupations that have been linked to an elevated risk of silicosis (6). In addition to the well-known silicosis, silica exposure has been related to a variety of autoimmune diseases. Miller et al. (2012) did a comprehensive analysis of the literature and discovered epidemiological evidence linking silica inhalation to the beginning of a number of autoimmune disorders. Systemic lupus erythematosus, rheumatoid arthritis, primary systemic vasculitis, Wegener's granulomatosis, and systemic sclerosis (SSc) are examples of these disorders (8). Anti-topoisomerase I antibodies are the most common autoantibodies in silica-induced systemic sclerosis (SSc). Systemic sclerosis (SSc) has been associated to 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. Understanding the relationship between silica exposure, silicosis, and systemic sclerosis 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 eight years. Systemic sclerosis, a terrible condition, developed progressively over time. As far as we know, this is the first recorded instance of Erasmus syndrome in Bangladesh.

**Case report:**

A 24-year-old stonecutter from Uttar Dinajpur, Bangladesh, attended Rangpur Medical University's Internal Medicine Clinic. The patient's medical history included three months of occasional blue staining of the fingers and toes when exposed to low temperatures. Figure 1 shows the patient's left middle and little finger ulcers and gangrenous alterations, which developed during the previous year as a result of progressive cutaneous constriction. The patient additionally reported increasing dyspnea with exercise, a mild nonproductive cough, polyarthralgia, and extensive cutaneous fibrosis. For the previous eight years, the individual had worked as a stonecutter, exposing them to significant quantities of silica dust in the job. The individual in question 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, which are 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, 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 brow.

The first hour lab tests showed a 55 mm erythrocyte sedimentation rate and 9.6 g/dL hemoglobin. HIV, HBV, and HCV serology and liver and kidney function tests were negative. All of the HIV, HBV, and HCV serology tests, as well as the liver and kidney function tests, came back negative. The existence of topoisomerase I was indicated by the presence of high titer positive in the ANA profile. 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). There were also several calcified lymph nodes in the mediastinum. This study found bilateral interstitial lung disease and calcified mediastinal lymph nodes, which might indicate the existence of silicosis and systemic sclerosis, commonly known as Erasmus syndrome.

**Discussion**:

Systemic sclerosis, silicosis, and Erasmus syndrome each have unique symptoms and results. The examination of the relationships between these components can provide insights into their impact on happiness. There may exist a potential association between systemic sclerosis and exposure to silica. (7). The presence of silica particles has the potential to stimulate the immune system and expedite or exacerbate the development of autoimmune diseases. Additional research is required in order to ascertain processes and establish causal relationships.

Systemic Sclerosis (scleroderma) is a chronic autoimmune illness that affects numerous physiological systems.Symptoms include fibrosis and vascular dysfunction.In addition to esophageal dysmotility, pulmonary hypertension, and interstitial lung disease, Raynaud's phenomenon causes telangiectasias, subcutaneous calcium deposits, skin fibrosis and thickening, myalgia, and other symptoms (10). A variety of environmental factors have been related to systemic sclerosis (SSc) via the finding of case clusters and thorough epidemiological studies. Exposure to silica dust has been related to increased rates of systemic sclerosis (SSc), and this relationship was initially observed in stone masons (11). According to a study, there is a similarity in the clustering behavior of South African gold workers and their counterparts in the United States coal mining industry (12).The present case pertains to an individual who has been subjected to prolonged exposure to silica dust in the workplace. Systemic sclerosis has been confirmed using the current diagnostic criteria (13). This particular case exhibits similarities to the diagnostic criteria of Erasmus syndrome.

Silicosis, an occupational respiratory ailment of considerable peril, poses a significant threat to the well-being of individuals employed across various industrial sectors. Silicosis is characterized by enduring consequences. The disease is permanent and often develops years after being exposed to silica dust (14). Silicosis is induced by the inhalation 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 results in the induction of inflammatory responses and fibrotic changes within the pulmonary tissue.The occurrence of inflammation has the potential to induce lung fibrosis, resulting in a decline in pulmonary function.Silicosis has been associated with tuberculosis (TB), immunological disorders, and chronic obstructive pulmonary disease (COPD) (15).

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 (16). Employers must utilize engineering methods to decrease silica dust production and distribution, such as the building of effective ventilation systems and the use of wet dust suppression technology. Employees should be given with 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, as well as the provision of substantial training programs focusing on hazard awareness and the adoption of safe work techniques, is required.

One of the most essential components in resolving the silicosis issue is enacting and enforcing rules and regulations. To maintain worker safety, governments and regulatory agencies must implement rigorous regulations on enterprises about allowed silica exposure levels in the workplace (17). It is critical to implement occupational health surveillance systems in order to monitor worker health and detect silicosis early. Access to good healthcare, particularly yearly medical exams, is crucial for early detection and treatment of silicosis and associated illnesses such as Erasmus syndrome.

**Conclusion:** it is imperative to establish a clinical connection between occupational silica exposure, silicosis, systemic sclerosis, and Erasmus syndrome. Healthcare professionals responsible for the care of patients who have experienced occupational silica exposure should possess knowledge regarding potential complications and prioritize prompt diagnosis and suitable treatment. Further investigation is warranted in order to comprehend the etiology of this condition and offer enhanced, tailored therapeutic interventions.

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