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Case Report

Erasmus syndrome: A rare case report

Chandan Kumar Sheet[1]*, Sudarsan Pothal[2], Rekha Manjhi[3], Pravati Dutta[4], Subhankar Chatterjee[5]

[1]*Dept. of Pulmonary Medicine and Critical Care, Calcutta Heart Clinic & Hospital, Kolkata, West Bengal, India*
[2]*Sri Jagannath Medical College and Hospital, Puri, Odisha, India*
[3]*Veer Surendra Sai Institute of Medical Sciences and Research, Sambalpur, Odisha, India*
[4]*KIMS Hospital, Bhubaneswar, Odisha, India*
[5]*Dept. of Internal Medicine and Critical Care, Calcutta Heart Clinic & Hospital, Kolkata, West Bengal, India*

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ABSTRACT

Erasmus syndrome is a rare clinical entity in which systemic sclerosis develops following exposure to silica with or without development of silicosis. There are very few cases reported in literature from India. Here we report a case of Erasmus syndrome in a 52 years male who is a stone crusher by occupation for last 22 years presented with dry cough, low grade irregular fever, progressive shortness of breath, slowly progressive sclerodactyly and features suggestive of Raynaud's phenomenon. Radiological evaluations revealed multiple subpleural and centrilobular nodules in bilateral upper, lower lobes and right middle lobe, conglomerated nodular opacities (progressive massive fibrosis- PMF). There was pleural thickening and calcification, bronchial dilatation and fibrosis, calcified right hilar and mediastinal lymphadenopathy. Serological markers like Anti Scl-70 antibody came out to be positive (3+) Based on clinical features diagnosis of systemic sclerosis was made. Based on exposure history and radiology silicosis was diagnosed. Association of silicosis and systemic sclerosis establishes the diagnosis of Erasmus syndrome. Treatment was started with Prednisone, cyclophosphamide and Nifedipine. Strict avoidance to cold and regular follow-up was advised.

1. Introduction

Inhalation of silica leads to multiple occupational and environmental illness.[1] Among which silicosis is most common, apart from silicosis it may manifest autoimmune disease like systemic sclerosis.[2] Erasmus syndrome is a very rare progressive occupational disease caused by inhalation of crystalline silica where there is development of progressive systemic sclerosis.[3] Here we present a case of 52 years stone crusher who was misdiagnosed as tuberculosis but subsequently diagnosed as Erasmus syndrome.

2. Case Presentation

A 52 years old male patient presented with increasing breathlessness on physical exertion, and tightening of skin over the fingers and forearms (for last 12 months), sporadic bluish discoloration of fingers on exposure to cold (suggestive of Raynaud's phenomenon), as well as low grade irregular fever and unproductive cough (for last eight months). There was no history of arthralgia, chest pain, dysphagia, haemoptysis, and loss of appetite or weight loss or reflux disease. He was non-smoker and had no history of hypertension or diabetes mellitus.

* *Corresponding author*.
E-mail address: chandansheet1986@gmail.com (C. K. Sheet).

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Figure 1: Loss of forehead wrinkling, patchy depigmented and pigmented areas suggestive of Salt and pepper dermopathy

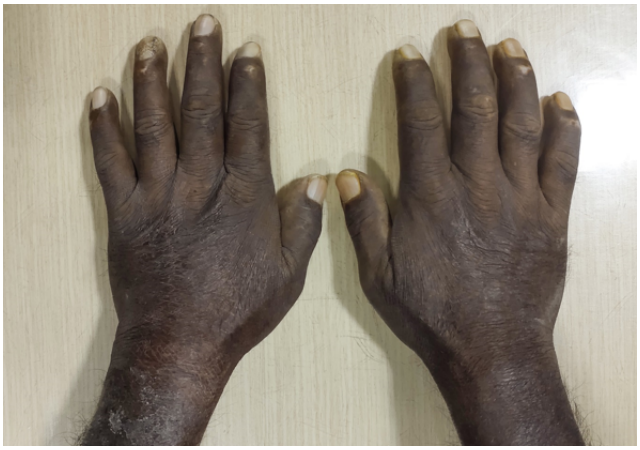

Figure 2: Skin fixity of hands and digits with apparent clawing and patchy pigmented and hyperpigmented areas over hands

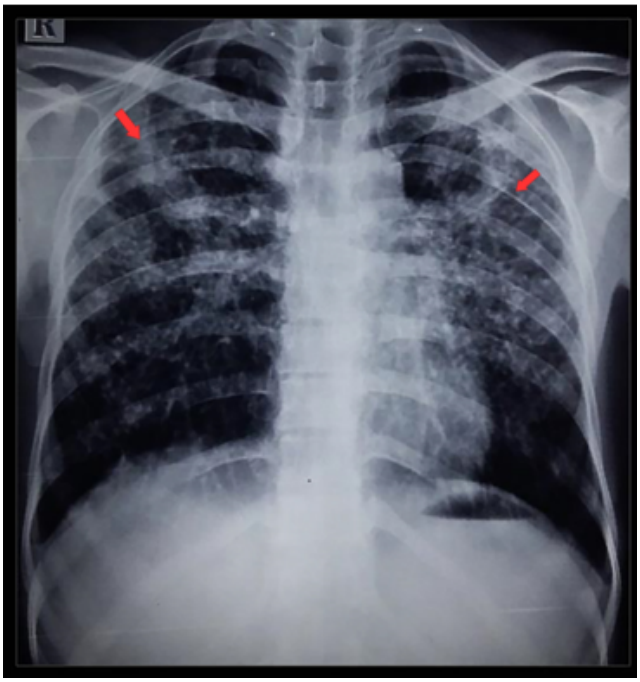

Figure 3: Chest X-ray showing bilateral upper andmidzones Micronodules (left &; right)

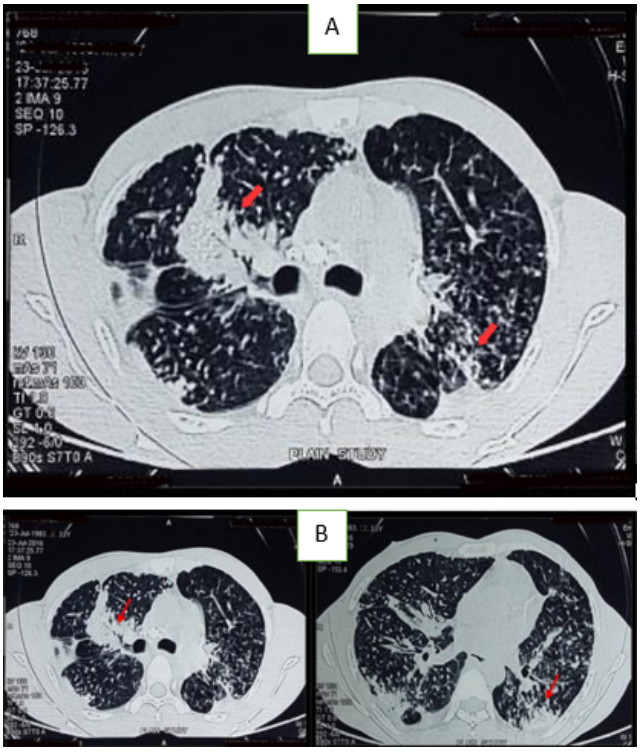

Figure 4: A&B: HRCT thorax showing multiple subpleural and centrilobular nodules in bilateral upper, lower lobes and right middle lobe and progressive massive fibrosis there is also bronchial dilatation and fibrosis

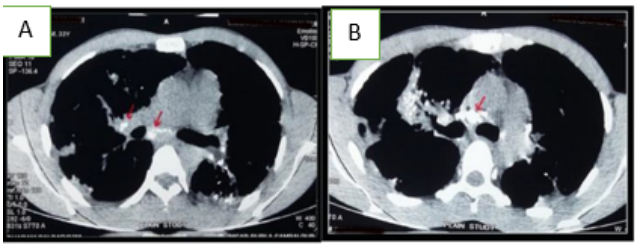

Figure 5: A & B: Mediastinal window of HRCT chest showing irregular pleural thickening with calcification, calcified right hilar and mediastinal lymphadenopathy

He worked as a stone crusher for nearly 22 years and was forced to leave that job nine months back due to excessive breathlessness. Treatment history revealed his treatment with anti-tubercular therapy four times at various local hospitals but he never completed a course due to gastrointestinal intolerance. Other than that, there was no other significant drug history. He had no family history of similar complaints.

General examination revealed no clubbing, pallor or cyanosis. There was loss of wrinkling over forehead, facial dry skin with irregular hypopigmentation and depigmentation suggestive of salt and pepper dermopathy (Figure 1). Digital examination revealed apparent clawing of fingers due to skin fixity. There was also thickening of skin over fingers and forearm and hypopigmented patchy areas over the dorsum of his hands (Figure 2). Joints of the upper and lower extremities were non-tender. There was no calcinosis cutis. Examination of respiratory system revealed vesicular breath sounds both side and inspiratory coarse crepitation over bilateral infraclavicular, mammary, suprascapular and infrascapular areas. No abnormalities were detected on cardiac and abdominal examinations.

2.1. Investigations

Full blood count, liver, renal function tests, serum electrolytes and urinalysis were within normal limits. Human Immunodeficiency Virus (HIV), Hepatitis B Virus (HBV), Hepatitis C Virus (HCV) serology was negative. Chest X-ray showed bilateral upper and midzones micronodules (Red arrows, Figure 3). High resolution Computed Tomography (HRCT) of the thorax showed multiple sub-pleural and centri-lobular nodules in bilateral upper, lower lobes and right middle lobe, conglomerated nodular opacities consistent with progressive massive fibrosis (PMF) (Red arrows in Figure 4a&b). Furthermore, there was pleural thickening and calcification, bronchial dilatation and fibrosis, calcified right hilar and mediastinal lymphadenopathy (Red arrows in Figure 5a&b). Spirometry revealed severe restrictive abnormality [Forced vital capacity (FVC)- 43%, Forced expiratory volume in the first second (FEV1)- 51% and FEV1/FVC 86%]. Sputum and Bronchoalveolar lavage (BAL) fluid were negative for Ziehl-neelsen stain (AFB). Fibreoptic bronchoscopy revealed normal tracheobronchial tree. BAL fluid gene xpert cartridge- based nucleic acid amplification test (CBNAAT) for mycobacterium tuberculosis and rifampicin (MTB/Rif) was not detected. BAL fluid cytology showed increased Polymorphonuclear (PMN), Lymphocyte and alveolar macrophages. Antinuclear antibodies (ANA) by Hep-2 method was found to be positive in high titres (1:640). In addition, nuclear speckled pattern and Anti Scl-70 antibody were also positive (3+). However, serum C reactive protein (CRP) (CRP 4mg/dl) and rheumatoid factor was negative. Echocardiography was normal and without any pulmonary arterial hypertension.

2.2. Diagnosis

Based on the presence of clinical features, Raynaud's phenomenon, sclerodactyly, skin tightening over the face and extremities, typical salt and pepper dermopathy, positive Anti Scl-70 antibody, a diagnosis of systemic sclerosis was made asper American college of rheumatology (ACR)/European league against rheumatism (EULAR) criteria for systemic sclerosis. Furthermore, chronic silicosis was diagnosed based on prolonged occupational exposure to silica (for 22 years) due to stone crushing, absence of any chronic drugs exposure or similar family history, presence of micronodules, progressive massive fibrosis (PMF), calcified mediastinal lymphadenopathy and fibrosis in chest radiology (HRCT chest). Finally, an etiological diagnosis of Erasmus syndrome (cutaneous systemic sclerosis associated with silica exposure) was proposed because of association of silicosis and systemic sclerosis.

2.3. Treatment and follow-up

Treatment was initiated with tapering dose of prednisolone (40 mg/day based on body weight 48 kg), cyclophosphamide (1 g/month for 6 months) and Nifedipine (started with 20 mg/day and with a plan to increase up to 120 mg/day over next 6 months and to continue lifelong). He was also advised to evade exposure to cold strictly and to attend follow-up clinics on a regular basis.

3. Discussion

Systemic sclerosis is a multisystem autoimmune disorder of idiopathic aetiology manifested vasculopathic changes, and diffuse tissue fibrosis in multiple organs such as lung, heart, kidney and gastrointestinal tract, as well as skin. It is predominantly found in females. However, males have a more severe expression of disease and higher mortality.[1] It may be caused by environmental and occupational exposures to vinyl chloride, organic solvent, and seldom by

silica.

Silicosis is a type of pneumoconiosis. It is an occupational disease characterised by irreversible lung fibrosis because of crystalline silica inhalation, retention, and pulmonary reaction among workers in mining, quarrying, masonry and sand blasting sectors. Silicosis is frequently linked with tuberculosis (silico-tuberculosis), lung carcinoma and occasionally related with systemic sclerosis (silica associated systemic sclerosis -SA-SS), systemic lupus erythematosus and rheumatoid arthritis.[2]

Exposure to silica affects both humoral and cellular immunity, hypergammaglobulinemia and results in increases in antinuclear antibody and positive rheumatoid factor, as well as T-helper and T-suppressor lymphocytes.[3] Moreover, continual exposure to silica results in changes in soluble interleukin-2 (IL-2) receptors. Elevated lymphokines can either trigger collagen synthesis directly or prompt other cells (like monocytes or mast cells) to release factors that further stimulate collagen production in fibroblasts. This assertion is corroborated by heightened levels of soluble IL-2 receptors observed in individuals with systemic sclerosis. It is postulated that silica could activate monocyte to release fibroblast proliferative factors (such as cytokine IL-1, 6, and TNF alpha), which in turn promotes collagen production, resulting in cutaneous sclerosis, vascular occlusion, and pulmonary fibrosis..[4]

In 1914 Bramwel first observed the association of previous exposure to silica and later development of systemic sclerosis.[5] Subsequently, Erasmus in 1957 demonstrated a notable occurrence of progressive systemic sclerosis (PSS) among gold miners exposed to dust containing a significant proportion of free silica (±30%). Later on, Devulder et al. in 1977 explored the link between silica and systemic sclerosis, suggesting the term "Erasmus syndrome" to describe the concurrent presence of silicosis and PSS.[6]

In India, the initial instance of "Erasmus syndrome" (silicosis-induced systemic sclerosis) was documented by Khanna et al. in 1997.[7] Following that, three additional cases were also documented.[8–10]

4. Conclusion

Erasmus syndrome is a sporadic progressive disease caused by inhalation of crystalline silica, where there is development of progressive systemic sclerosis, which leads to progressive dyspnoea and multiple symptoms. There might be associated interstitial lung disease or severe restrictive lung disease over silicotic lung. Sometimes, pulmonary hypertension could be developed which can complicate the morbidity of silicosis. Therefore, while dealing with the systemic sclerosis occupational exposure to silica to be searched and while dealing with silicosis features of systemic sclerosis to be looked for as well as serological testing must be ordered to detect early Erasmus syndrome.

5. Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his 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.

6. Source of Funding

None.

7. Conflicts of Interest

There are no conflicts of interest.

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Author biography

Chandan Kumar Sheet, Consultant https://orcid.org/0000-0002-4173-2377

Sudarsan Pothal, Professor and HOD https://orcid.org/0000-0002-9789-2855

Rekha Manjhi, Professor and Head

Pravati Dutta, Professor and Head

Subhankar Chatterjee, Consultant

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