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CASE REPORT

Erasmus Syndrome in a 35-Year-Old Male: A Rare Case Report

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Abstract:

Silicosis is an inflammatory disease of the lung that develops from prolonged pulmonary inhalation and retention of crystalline silica and an immune reaction characterized by irreversible lung fibrosis. It mainly occurs in people involved in stone-quarrying, mining, and sand blasting. Erasmus syndrome is a rare condition where systemic sclerosis develops following exposure to silica with or without silicosis. Only a few case reports are available in the literature. We report here a case of Erasmus syndrome in a 35-year-old manual laborer who presented with arthralgia, Raynaud's phenomenon, skin tightening and microstomia along with features of Interstitial Lung Disease (ILD) and pulmonary arterial hypertension. Serological markers of systemic sclerosis were strongly positive. After a diagnosis of Erasmus syndrome was made, a combination of drugs including Prednisone, Cyclophosphamide, and Nifedipine was instituted. This led to improvement in his symptoms over 6 months.

Keywords: *Interstitial lung disease, Pulmonary arterial hypertension, Silicosis, Systemic sclerosis]*

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Case Report:

A 42-year-old non-diabetic and non-hypertensive male patient presented to our facility with a history of progressively worsening shortness of breath and persistent dry cough for 2 years. For this he was given CAT 1 anti tubercular therapy 5 months after his initial symptoms without significant improvement. After 2-3 months he noticed both hypo ad hyper pigmentation on different part of the body including over the scalp, upper part of the neck, chest and leg [figure 1]. Subsequently he also noticed gradual thickening and tightening of the skin of hand [fig 2], face, feet and trunk. As it was progressive, he felt difficulty in gripping as well as difficulty in opening of mouth. There was also typical color change on exposure to cold. He also gave history of multiple joint swelling and pain involving small joints of hand and feet, both wrist and knee joints without stiffness and was improved with rest but required analgesic at times. He had repeated episodes of heart burn without any difficulty in deglutition. Occupational history revealed that he worked as a stone crusher for nearly 10 years in a stone quarry but left his job 2 years ago. No family history of similar complaints was noted.

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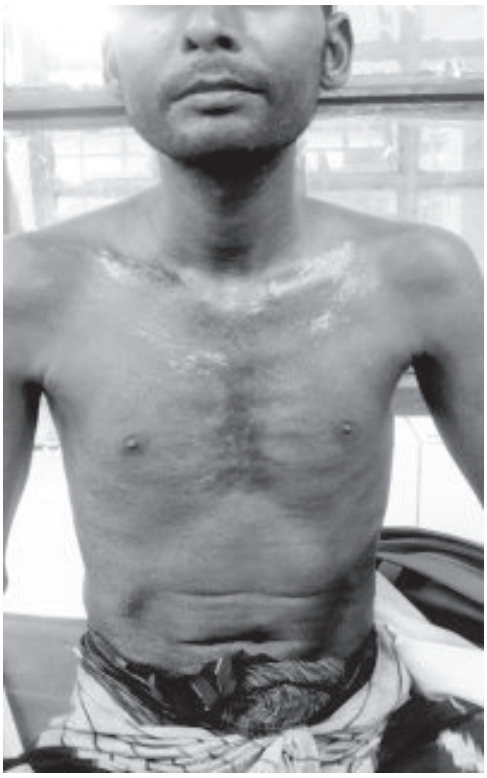

Fig.-1: skin pigmentation over body.

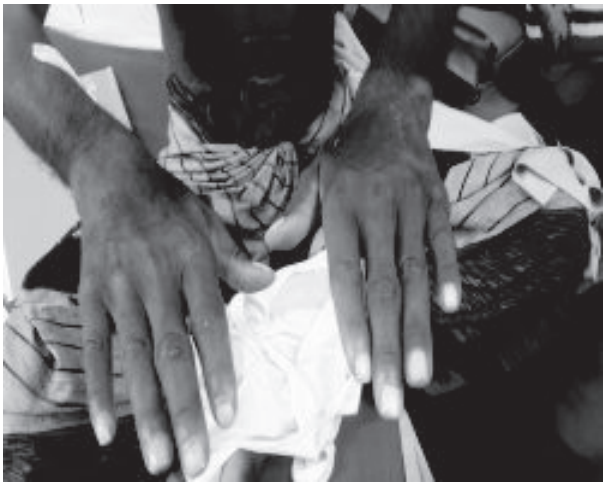

Fig.-2: Sclerodactyly and pseudoclubbing

On examination he was anxious and ill-looking. Face is smooth, shiny, tight, immobile with hypo and hyper pigmented area. There was loss of wrinkling of forehead. Nose was pinched up and tapered. There was puckering of skin around the lip with difficulty in opening the mouth. The skin of the both hands is smooth, shiny, tight, thick, edematous, with pigmented and hypo pigmented area. Skin of the other parts of the body was also tight and thick. Respiratory system examination revealed bilateral end-inspiratory crepitations in upper and mid-chest. Cardiological evaluation showed normal heart rate and rhythm, S1- normal and P2- loud with no added sounds. CBC, urinalysis, LFT and electrolytes were within normal limit. HIV, HBV and HCV serology was negative. Interestingly chest x-ray was unremarkable [fig 3]. High resolution Computed Tomography (HRCT) of the chest revealed diffuse fibrosis, nodular lesions, occasional ground glass changes involving mainly the upper and middle lobes suggestive of Interstitial Lung Disease (ILD) [fig 4]. In Pulmonary function test spirometry revealed moderate restrictive defect, 6MWT revealed desaturation with moderate limitation of walking distance and DLCO consistent with restrictive ventilatory defect. Anti nuclear antibody by indirect fluorescent (IF) was positive and Anti Scl-70 antibody was also strongly positive. Echocardiogram showed left ventricular ejection fraction of 66% and raised pulmonary artery systolic pressure (PASP=55 and PAMP=40 mm Hg).

Considering the presence of telltale clinical manifestations like arthralgia, Raynaud's phenomenon, skin tightening over the face and extremities, microstomia, and Interstitial Lung Disease (ILD), along with supporting laboratory evidence including significant pulmonary arterial hypertension and strongly positive anti-Scl 70, a serological marker, clinic-pathological diagnosis of systemic sclerosis was safely made.

Again, in light of the significant occupational exposure to silica and the absence of any chronic drugs that induce lung fibrosis, such as bleomycin, methysergide, cyclophosphamide, and others, as well as any significant family history, a final etiological diagnosis of Erasmus syndrome (diffuse cutaneous systemic sclerosis associated with silica exposure) was made in the absence of any other possibility. Treatment was started with prednisone and cyclophosphamide for ILD and nifedipine for pulmonary hypertension and Raynaud's with avoidance of cold exposure. There was improvement in skin tightening, arthralgia, Raynaud's phenomenon, and cough, although marginal improvement in dyspnoea on follow up over the phone as the patient could not attend

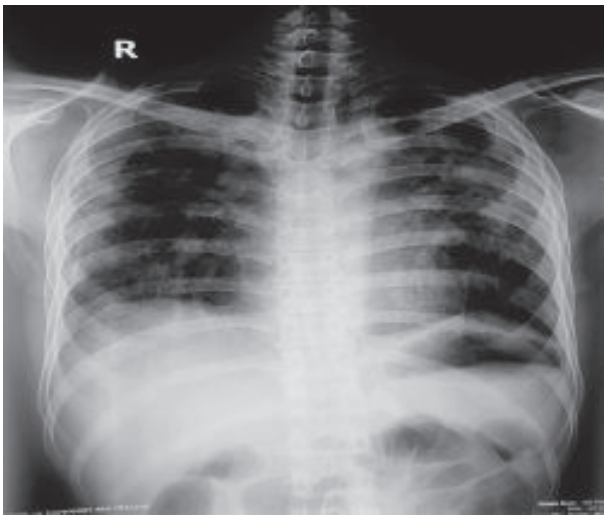

Fig.-3: chest x-ray

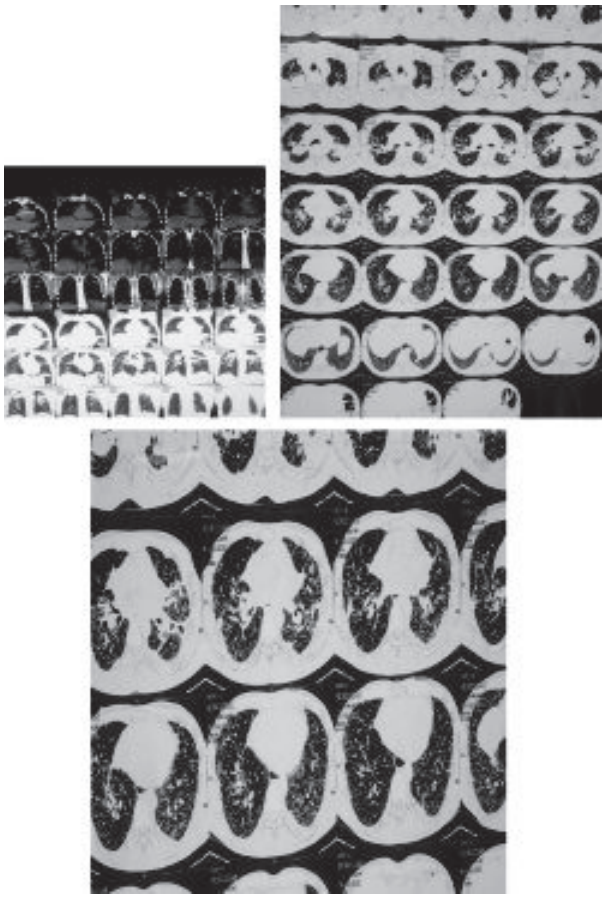

Fig.-4 (a,b,c): HRCT of chest.

physically due to the COVID pandemic situation. As his condition was static, he refused to come in our institute due to economic constraint.

Discussion:

Systemic sclerosis is an autoimmune inflammatory disease that causes vascular changes and degeneration with diffuse tissue fibrosis affecting the skin, lung, kidney, heart, gastrointestinal tract, and synovium[1]. Numerous occupational and other exposures, including vinyl chloride, epoxy benzene, organic solvents, silica environmental and occupational exposures have been implicated as potential causes of SSc[2].

Continuous inhalation of mineral dust containing silica leads to silicosis, probably the most common form of pneumoconiosis. Silicosis is an inflammatory disease of the lung that develops from prolonged pulmonary inhalation and retention of crystalline silica that causes an immune reaction mounted by the body to this extraneous chemical characterized by irreversible lung fibrosis. It mainly affects people involved in stone-quarrying, mining, and sand blasting as an occupational hazard[3]. Silicosis is often associated with other diseases of the lung, such as pulmonary tuberculosis, lung carcinoma, and less commonly, autoimmune diseases like systemic sclerosis (SSc), rheumatoid arthritis (RA), and systemic lupus erythematosus (SLE)[4]. Continuous exposure to silica is associated with abnormalities of humoral and cellular immunity as well as alterations in T-helper and T-suppressor lymphocytes, hypergammaglobulinemia, and antinuclear antibody and rheumatoid factor often become positive[5]. Based on these immunological aberrances, silica exposure has been incriminated as a cause of progressive systemic sclerosis.

Erasmus in 1957 first described the association [6] of exposure to silica and the development of systemic sclerosis, although credit for the first observation of this association goes to Bramwel in 1914. Patients with silica-associated systemic sclerosis (SA-SSc) are clinically, serologically, and immunologically indistinguishable from those with idiopathic systemic sclerosis (SSc), but SA-SSc patients have a higher prevalence and severe pulmonary involvement (bibasilar fibrosis)[7,8]. Although spontaneous remission in idiopathic SSc was reported, no such event was found in silica-induced SSc[8]. Anti-SCL-70, which is the predominant autoantibody present in SA-SSc, is usually associated with severe interstitial lung

disease. Perhaps there is increased production of the anti-Scl-70 antibody in genetically susceptible people who are exposed to silica[9]. Survival in SA-SSc patients was found to be less than in the control "idiopathic" SSc group[10].

So, it is clear that silicosis should be ruled out in every patient with SSc, especially in males, because lung involvement is an important aspect in the prognostic outcome and avoidance of exposure to toxic agents can stabilize the disease progression and even lead to improvement in some cases.

Conclusion:

An underlying systemic sclerosis, which contributes to dyspnoea by multiple mechanisms-pulmonary fibrosis, pulmonary artery hypertension, and localized thoracic skin disease, must be kept in mind whenever the degree of dyspnoea cannot be explained by the extent of silicosis induced lung fibrosis alone and especially if additional clinical pointers are present.

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