INTRODUCTION

Systemic sclerosis (SS) has been defined as a generalised disorder of the small arteries, microvessels and the diffuse connective tissue, characterized by scarring and vascular obliteration in skin, gastrointestinal tract, lung, heart, and kidney. The aetiology of SS is unknown, but it is probably caused by both endogenous and exogenous factors. Among the exogenous factors, several type of occupational exposure have been suggested as potential cause of SS including silica dust, vinyl chloride, epoxy resign, bleomycin, aliphatic and aromatic hydrocarbon, and toxic oil. Except for silica, all other substances induce changes that only mimic scleroderma and are partially reversible if the exposure is discontinued. Silica dust exposure is a well recognized risk factor for developing systemic sclerosis. Crystalline silica (quartz) particles less than 1 micrometer are the most pathogenic in silicosis. Crystalline silica particle are inert and remain in the tissue. They are taken up by macrophages to which silica is toxic. The macrophages die and silica is released allowing ongoing exposure. The hypothesis exists that silica can mediate cell activation and induces ICAM-1 on endothelial cells. This is though aid monocyte adhesion and migration to the dermis where an inflammatory response is set up. Silica can activate monocyte to release cytokine IL-1, and TNF which are fibroblast proliferative factor which increase production of collagen leading to cutaneous sclerosis, vascular occupation and pulmonary fibrosis.

CASE REPORT

A 32 years old male who was the manual stone cutter was presented with three and half years history of dyspnoea on exertion and non-productive cough, one and half years history of arthralgia, stiffening of small and large joint with diffuse muscle pain and one year history of skin thickening over dorsum of hand, forearm, face, leg and over abdomen. Patient also had complains of nausea, dysphagia, decreased appetite and weight loss. He also had features of Raynaud’s phenomenon in fingers and toes. There was no history of fever and photosensitivity. He has been worked as manual quartz stone cutter for about 17 years. He did not recall any similar complaints in his occupational friends and family member. He was taking anti tuberculous treatment since eight months before admission. On general physical examination there

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Silica Associated Systemic Sclerosis

DISCUSSION

Compared to other connective tissue disorder, silica associated systemic sclerosis (SA-SS) is relatively rare, has worldwide distribution and affects all races, but there is no epidemiological data available in Asia especially in India regarding silica-associated systemic sclerosis (SA-SS) to the best of our knowledge while it is a one of the rare complication of silicosis. In 1914 Bramwell noted an association between systemic sclerosis and silica but he was failed to link casually SS and silica exposure. In 1957 Erasmus reported the high prevalence of SS (32%) in gold miner exposed to dust containing a high percentage of free silica. These finding were confirmed by Rodman and colleagues who reported prolonged and heavy exposure to silica dust in 60 (42%) SS men, Sluis-Cremer and colleagues conducted a case-controlled study in which “cumulative lifetime silica exposure” was shown to be significant higher in cases (13%) compared with control. The risk of developing SA-SS after exposure to silica dust is up to four times greater than that for general population. Sometimes, the development of SSc may be consequence of the intensity rather than the duration of silica exposure. SA-SS appear after a mean of 16.2 year (range 4-36 yrs) of silica exposure. In majority of cases (72%), silicosis occurs before the onset of SA-SS. The lungs are almost invariably involved in SA-SS, with pulmonary fibrosis in the basal part. This index case had been worked for 17 years before he developed symptoms of SS and radiographic pattern of silicosis was not detected in routine chest radiograph. The role of genetic factor in the pathogenesis of SA-SS is partly clear in view of the fact that every person exposed to silica dust does not develop systemic sclerosis. But it is important to remember that these abnormalities occur in the context of necessary genetic & environmental background. Haustein et al supported the genetic association (HLA-A-DR3 alleles) with SA-SS but the review of dates & ages of onset, suggested that the onset of SA-SS is more likely to have an environmental trigger and encoded genetically. It is well established that crystalline silica particle < 1 micron are the most pathogenic in silicosis. These particles are phagocytised by...
FIGURES AND LEGENDS

Figure 1:
X-ray of both hand showing resorption of tip of terminal phalanges (acro-osteoysis) of index finger of both hands. (a) magnifying view (b)

Figure 2:
Barium swallow examination showing persistence of barium in oesophagus suggestive of decreased motility

Figure 3:
Chest x-ray (p a view) showing prominent linear markings in both lower zones

Figure 4:
CECT of the thorax mediastinal window, showing enlarged pretracheal, subcarinal and right paratracheal lymph node (a) CECT lung window showing small peribronchial, interstitial and pleural base nodules (b) interlobular septa thickening (c) irregular pleura-pulmonary interface (d)

We have reviewed the literatures to know how silica dust can cause or responsible for systemic effects of systemic sclerosis in addition to pulmonary fibrosis. The fact that same (IL1, IL2, IL 6 & TNF) and possibly other cytokines induces the maturation and activation of dendritic cells in the regional lymph nodes which is the key connecting element. It follows a polyclonal activation & proliferation of native T cells. Such T cells are thought to operate to cause vascular occlusion & cutaneous sclerosis.

It is established now that patient with SA-SS have clinical, immunological and serological pictures which are indistinguishable from idiopathic SS, although as a group the SA-SS patients have higher percentage of pulmonary involvement (bibasilar fibrosis) and the anti-Scl-70 antibody. In our patient clinico-radiological, immunologically and serologically parameters of SA-SS were indistinguishable from those of idiopathic SS but...
patient had history of exposure to silica dust since last 17 years which gives clue to the diagnosis of SA-SS.

REFERENCES