ABSTRACT

Aim: Silicosis also known as potters rot is the most common occupational lung disease. People employed in occupations like sandblasting, surface drilling, tunneling, silica flour milling, ceramic making are predisposed to developing silicosis. We report a case of progressive massive fibrosis secondary to silicosis in a stone quarry worker.

Case Report: A forty five year old stone quarry worker presented with chronic dry cough and breathlessness. His chest CT showed presence of multiple calcified mediastinal lymphnodes with irregular mass like areas. Based on the occupational exposure and radiographic images, a diagnosis of progressive massive fibrosis due to silicosis was made.

Discussion: Pneumoconiosis is group of lung diseases related to occupational exposure to inhaled dust. The most common among pneumoconiosis is silicosis. Based on the amount and duration of exposure the clinical and radiological features of silicosis vary. Progressive massive fibrosis is a potentially fatal stage in complicated silicosis. In a majority of cases, a positive occupational history and radiological features are sufficient to make a diagnosis.

Conclusion: There is no specific treatment for silicosis. Avoidance of further exposure, using personal protective measures, periodic medical checkup and strict legislations to protect employees and a system to check compliance should be ensued.

Key Words: Progressive massive fibrosis, Silica dust

INTRODUCTION

Progressive Massive Fibrosis (PMF) is a potentially fatal and debilitating occupational hazard occurring in persons working in respirable dust industries. This condition most commonly occurs in association with occupational lung diseases like coal workers pneumoconiosis and silicosis. We report a patient with progressive massive fibrosis secondary to silicosis. The diagnosis was established by occupational history and radiological features.

CASE DESCRIPTION

A forty five year old man presented with symptoms of dry cough and breathlessness on exertion of six months duration. He was not a smoker and had no previous history of tuberculosis. He had worked in a stone quarry for twenty years and had no co morbid illness. On examination his vitals were stable. On Respiratory examination there were bilateral scattered crepts. His chest x ray (fig 1) showed diffuse reticulonodular opacities and multiple mass-like symmetrical lesions with irregular margins in the hilar region bilaterally. High resolution computed tomography (HRCT) of chest revealed presence of multiple calcified mediastinal lymphnodes with irregular mass like areas in the bilateral upper and left lower lobe predominately in the hilar and peri hilar region. Few specks of calcification were noted within the lesion. There were also multiple nodules of varying sizes with centrilobular and perilymphatic distribution. (fig2,3) Imaging, supported by occupational history was suggestive of progressive massive fibrosis (PMF) due to silicosis. The patient is currently on symptomatic management and has not worsened till the last contact.

DISCUSSION

Silicosis also known as “potters rot” is an important occupational lung disease caused by inhalation of crystalline silica.
Silicon is abundant in nature as free silica in combination with oxygen (quartz) or as silicates in combination with oxygen and other elements. Silicon contributes to about 28% of the earth’s crust and a major part exists in quartz form. The sandstone industry, stone quarrying and dressing, granite industry, grinding of metals, sand blasting, iron and steel foundries, silica milling, flint crushing and manufacture of abrasive soaps are some of the occupations related to silica exposure. Slate pencil industry and agate grinding industry which is peculiar to India carry a high risk of silicosis. Crystalline silica is classified as a group 1 substance by the International Agency for Research on Cancer.

Silicosis has affected humans since ages. In 1705, Ramazzini cited Diembrock’s description of the lungs of stonecutters who possibly had silicosis. The term silicosis was coined by Visconti in 1870. Though occurring since ages, due to industrialization and mechanized mining, the prevalence of silicosis has increased alarmingly in the twentieth century. In 2007, the U.S. Occupational Safety and Health Administration (OSHA) estimated that more than two million employees are exposed to silica in general industry, construction, and maritime industry. Epidemiological studies on silicosis in India has shown varied prevalence ranging from 3.5% in ordnance factory to 54.6% in slate pencil industry.

The primary pathology in silicosis is the formation of silicotic nodules. The inhaled silica particle is engulfed by the alveolar macrophages. However they are unable to digest the material. The silica particle in the macrophages damages the lysosomal membranes of the alveoli which triggers the release of proteolytic enzymes into the cytoplasm leading to death of the macrophage. Continued exposure results in an alteration of the macrophage function. Release of inflammatory cytokines like interleukin 1, free radicals and growth factors follows which stimulates collagen synthesis and production of antibodies against collagen. These anticollagen antibodies stimulate fibroblasts to then produce more collagen which eventually leads to silicotic nodule formation.

Based on the amount, duration of exposure and onset of symptoms, silicosis can be classified as acute silicosis, chronic silicosis and accelerated silicosis. “Chronic simple silicosis”, the commonest form, develops after long-term exposure usually 10-30 years to smaller amounts of silica dust. “Accelerated silicosis” develops 5-10 years after exposure and progresses rapidly with a higher risk of complicated disease. “Acute silicosis” also called Silicoproteinosis develops a few weeks to 5 years after exposure to high concentration of silica dust. Acute silicosis progresses rapidly to respiratory failure and death. When severe scarring leads to confluence of small nodules into a larger lesion with more severe symptoms and respiratory impairment, it is termed as “Complicated silicosis”. It is more common in accelerated type than with the chronic variety.

Majority of the patients present in chronic silicotic stage. Even in the presence of advanced radiological lesions, patients are asymptomatic. The most common symptom is dyspnoea on exertion. The severity of dyspnoea increases with progress of the disease. Cough is associated with minimal expectoration in the initial stages and the productivity increases as the disease advances. Chest pain and haemoptysis indicate the possibility of complication like tuberculosis. In late stages cor pulmonale results. Silicosis may be complicated with other lung diseases including lung cancer and autoimmune diseases. Patients with silicosis are more susceptible to developing pulmonary tuberculosis, “silicotuberculosis”. Patients with silicosis are also prone to develop repeated Infections, chronic obstructive pulmonary disease and tracheobronchial compression by enlarged mediastinal lymphnodes. Pleural involvement in silicosis is rare. Few instances of bilateral pneumothorax have been reported.

Radiological lesions vary in different types of silicosis. On HRCT thorax, multiple bilateral centrilobular opacities, multifocal patchy ground-glass opacities, and consolidation with occasional crazy paving characterize acute silicosis. Chest x ray of patients with chronic simple silicosis shows the presence of multiple small nodules, 2-5 mm in diameter with associated calcifications. These nodules have a upper lobe predominance. HRCT in chronic simple silicosis shows perilymphatic distribution of nodules in centrilobular, paraseptal, and subpleural regions. Hilar and mediastinal lymphadenopathy may precede the parenchymal lesions. Eggshell pattern calcification of lymph nodes is common. Complicated silicosis, also known as progressive massive fibrosis, develops with confluence of individual silicotic nodules. On CT scan, PMF appears as focal soft-tissue masses, usually measuring more than 1 cm in diameter, with irregular margins, calcification, and involving apical and posterior segments of the upper lobes, surrounded by areas of emphysematous changes. With advancing fibrosis, these opacities migrate towards hila with resultant paracatricial emphysema. In silicosis associated with tuberculosis (silicotuberculosis) the radiological picture includes asymmetric nodules or consolidation, cavitation, and a rapid disease progression.

Silicosis is usually diagnosed by eliciting a positive occupational exposure history. A positive occupational history with radiological features of silicosis is sufficient to make a diagnosis of silicosis. Lung biopsy is rarely required. Lung biopsy may show the presence of silicotic nodules. A typical silicotic nodule has the following characteristics; a central zone with whorls of dense, hyalinized fibrous tissue, a mid zone with concentrically arranged collagen fibers and an outer zone with randomly orientated collagen fibers mixed with dust-loaded macrophages and lymphoid cells. Patients of silicosis should be screened for tuberculosis.
There is no specific treatment for silicosis. Avoidance of further exposure is the first step in treatment. Only a few treatment options are available and they are mostly for symptomatic management. Bronchodilators and corticosteroid therapy may be useful\textsuperscript{13, 14}. N-Acetyl cysteine has shown reduction in lung fibrosis in silica exposed rats\textsuperscript{15} Lung transplant has been an option for end-stage disease treatment. However, logistic reasons prevent advising lung transplant for all patients. It should be remembered that silicosis is a completely preventable disease. Education to workers, use of personal protective equipment and periodic medical screening should be made compulsorily. If PMF is diagnosed the person should immediately cease work in the industry. Strict legislations to protect employees and a system to check compliance should be ensued.

**CONCLUSION**

Silicosis is a common occupational disease which can present even after cessation of exposure. Majority of the patients can be diagnosed by eliciting a proper occupational exposure history. There is no specific treatment. Strict legislations with regard to using personal protective equipment in the occupational area and a periodic health check up for the employees is the need of the hour. In countries with high incidence of Tuberculosis, possibility of silico tuberculosis should always be considered.

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**Figure 1:** Chest X-ray showing mass-like symmetrical lesions with irregular margins in the hilar region bilaterally.

**Figure 2:** Bilateral multiple centrilobular and perilymphatic nodules with irregular mass like areas in left lower lobe

**Figure 3:** Calcified mediastinal lymphnodes with irregular mass likes areas in the bilateral upper lobes