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Disabling Silicosis Having Grave Prognosis- A Preventable Ancient Malady Still Prevalent in Modern Era - A Case Report & Review of the Subject

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SUMMARY

This case describes a middle-aged man who developed an advanced disabling and irreversible fibrotic lung disease after occupational over-exposure to fine respirable crystalline silica in a poorly ventilated enclosure for nearly two decades. A few years ago, his brother died of a similar illness at a younger age with same occupational exposure. Unfortunately, no correlation between dusty occupation, clinical features and radiological findings was considered until irreversible interstitial pulmonary fibrosis had set in. We report this case to underscore the importance of a relevant work place exposures history in the genesis of a vigilant and early diagnosis for improved patient outcomes.

Key words: Interstitial fibrosis, pneumoconiosis, silicosis.

INTRODUCTION

Silicosis is a fibrotic lung disease due to the inhalation of crystalline silica, usually in the form of quartz/rock dust/free silica (SiO_2 crystals) present in respirable size.[1] A number of industries and occupations are at risk including but not limited to mining, quarrying, stone works, foundries, abrasives, sand blasting, ceramics, concrete reconstruction, and glass making. Frequently the exposure is forgotten, missed or in the remote past and the true prevalence is difficult to estimate; reported cases worldwide represents the tip of the iceberg as only less than one third of the total cases are reported.[2] The diagnosis of silicosis is considered in a patient having positive occupational exposure to silica in the presence of compatible radiological features, while no specific treatment exists and prevention remains the preferred strategy.[1,2]

CASE REPORT

A 41 year old gentleman was admitted under pulmonary service via accident and emergency department due to acute worsening of dyspnea. He had a gradually progressive dyspnea for the past eighteen months that made him home-bound for one month and bed-bound for the past two weeks. It was associated with a dry, hacking cough and wheezes. There was no fever, sputum, chest pain, hemoptysis or upper respiratory symptoms. His respiratory problem started five years ago as an acute community acquired respiratory illness that responded poorly to antibiotics. Chest radiographs (Fig. 1) at that time lead to an empirical prescription of anti-TB treatment by his personal physician though his tuberculin skin test and sputa smears for AFB were negative. He had clinical improvement and could resume his daily activities until the next year when he developed a localized pleuritic anterior chest pain in association with worsening dyspnea. He consulted a senior respiratory physician who based on the radiographic features (Fig. 2) and no microbiological investigations, treated him as multi-drug resistant pulmonary tuberculosis with an under dosage regimen containing ethambutol, pyrazinamide, cycloserine and ethionamide. Patient could not adhere to this therapy and quit within 9 months, as he noticed further deterioration in

dyspnea and epigastric discomfort that made him home-bound, and he was lost to follow up. He This gentleman used to own and supervise a lucrative grinding plant where they used to crush rocks and sell off this material to ceramics industry. He used to smoke while being in this poorly ventilated workspace for up to 10 hours a day. There was no history of use of exhaust ventilation or any protective masks or equipment reflecting meager awareness of the potential hazards of fine occupational dusts exposure. His brother, who had similar occupational exposure as the index case, was also diagnosed as having chronic silicosis but he died due to a non-respiratory illness at the age of 36 years.

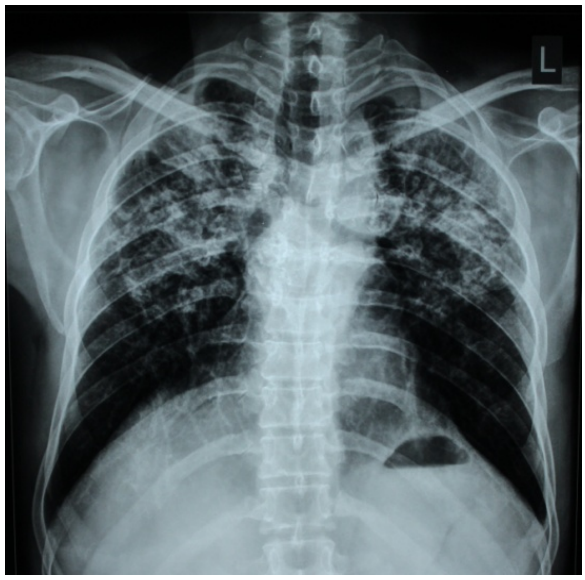

Fig. 1: Bilateral symmetrical nodular shadows involving the upper radiological zones. Mediastinal and hilar lymph node enlargement showing egg-shell calcification.

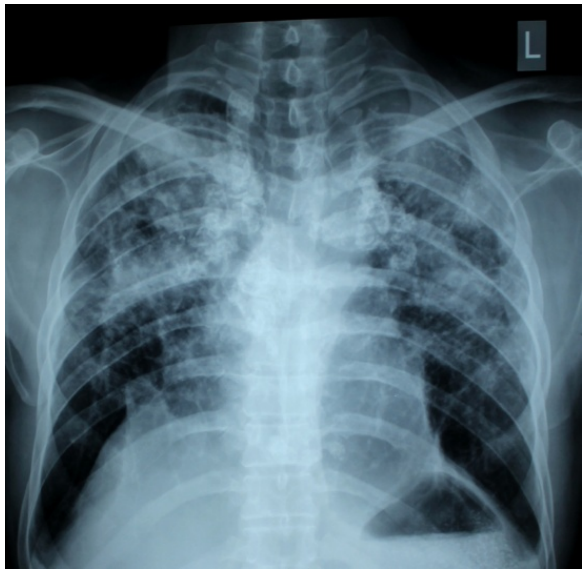

Fig. 2: Bilateral loss of lung volumes and tenting of diaphragms due to progressive massive upper lobes fibrosis.

On physical examination, he was a thin lean person weighing 59 kg. His blood pressure, temperature and pulse were normal while he was breathing at a rate of 28/min, maintaining oxygen saturation at 91% while breathing room air. There was generalized reduction in chest expansion, vesicular breathing and crackles with wheezes all over but reduced movements and hyperresonant percussion and reduced breath sounds intensity was appreciated on the left side. His second heart sound (P2) was loud with tachycardia while abdominal, musculoskeletal and neurological examinations were unremarkable. Investigations included CBC (haemoglobin 15.7 g/dL, WCC10.8 x 10^9, platelets 316 x 10^9 and ESR was 05 mm at 1^{st} hour. Biochemical profile revealed slight elevation of liver enzymes (bilirubin 0.2 mg/dL, ALT 74 IU/L, AST 147 IU/L, alkaline phosphatase 203 IU/L). Renal functions were normal. Serum IgE was 363 IU, indicating sensitization to occupational dusts. Spirometry illustrated severe obstruction without any reversibility and blood gas analysis showed a respiratory alkalosis with arterial hypoxemia (pO2 71 mmHg). On 6 minutes walk test, he could cover a distance of 195 m while breathing room air, and there were no desaturations below 90%.

Chest radiograph (Fig. 3) showed bilateral almost symmetrical confluent nodular shadows involving the upper radiological zones, along with enlargement and egg shell calcification of the hilar and mediastinal lymph nodes. A hyperlucent area without lung markings was seen in the left upper zone, raising suspicion of a localized, secondary spontaneous pneumothorax that was confirmed on HRCT chest (Fig. 5A and B) that also delineated the presence of egg shell calcified enlarged hilar, mediastinal, para-aortic and subaortic lymph nodes.

Normal lung parenchyma in upper lobes was replaced by fibrotic masses with few air bronchograms evident on the right side. Lung bases were characteristically spared and extrapulmonary calcified silicotic nodules were also evident in the

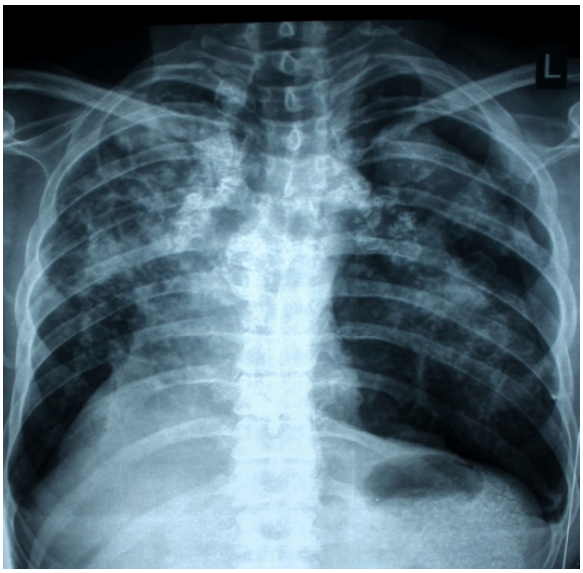

Fig. 3: Pnemothorax in left upper radiological zone, contralateral shift of mediastinum and trachea to the right and calcified nodular shadows in the spleen.

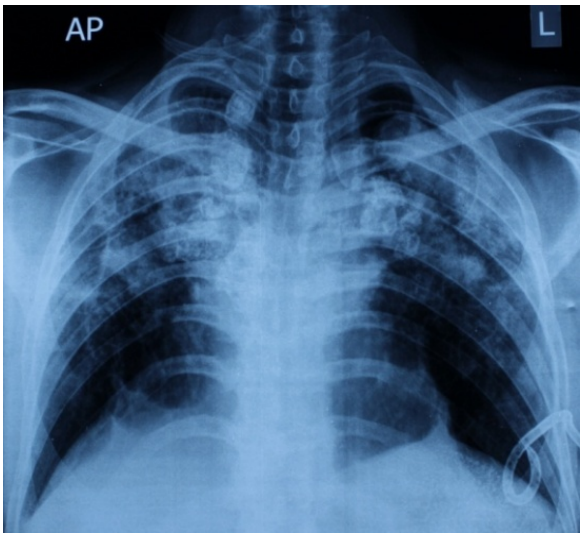

Fig. 4: Pigtail catheter in left pleural space with full lung expansion and central mediastinum.

spleen (Fig. 5 C). Keeping in view the strong history of occupational exposure, clinical features and

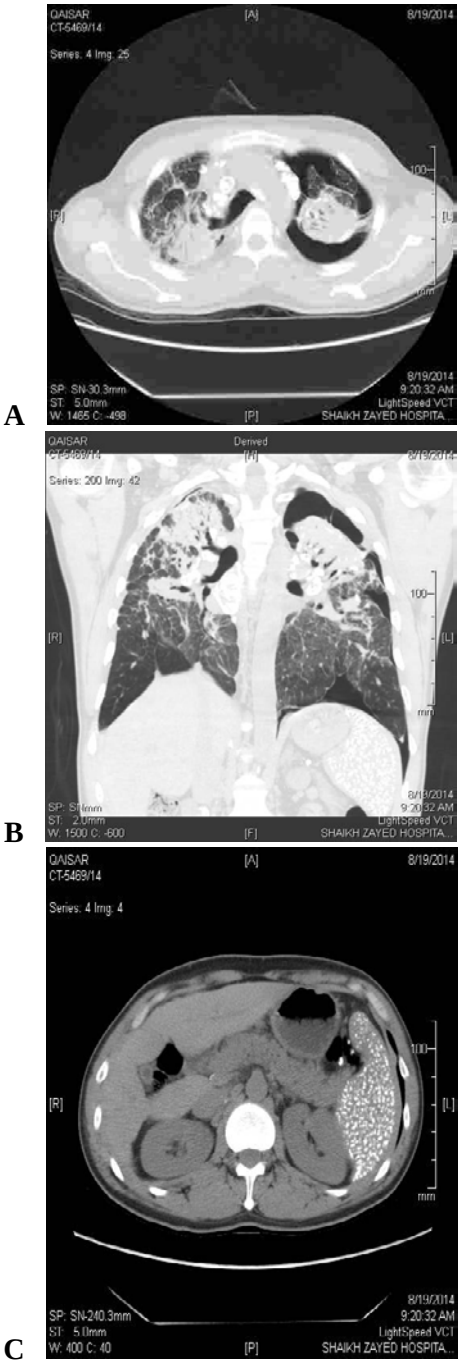

Fig. 5: HRCT chest images showing an organized chronic pneumothorax on the left side, bilateral upper lobe fibrotic masses with relative sparing of the lung bases (A and B). Nodular calcification is seen in the spleen (C).

characteristic radiological findings, a final diagnosis of 'Progressive Massive Fibrosis due to Chronic

Silicosis' was made. The pneumothorax was localized by the assistance of a bedside pleural ultrasound examination that showed replacement of normal granular appearance of lung parenchyma by linear white shadows (barcode sign) in the presence of air. Under aseptic measures, a small-bore (10 F) pigtail catheter was inserted in the left pleural space in the mid axillary line (Fig. 4). After lung was fully expanded, pleurodesis was performed using 8 grams of talc slurry. Patient was also managed with inhaled bronchodilators and anti-inflammatory drugs (salmeterol and fluticasone 25/250 mcg 2 actuations BD, salbutamol 2 actuations PRN and oral theophylline) along with mobility exercises, calcium and vitamin D supplements, pneumococcal vaccine and elaborate pulmonary rehabilitation. He was advised to avoid smoking and any kind of dust exposure and continue to follow up in out patients' pulmonology service.

DISCUSSION

Silica (silicon dioxide) is the most abundant mineral on earth and exists in both crystalline and amorphous forms.[1, 5] Amorphous forms, including vitreous silica and diatomite (formed from skeletons of prehistoric marine organisms), are relatively nontoxic.[3] Crystalline forms of silica include quartz, cristobalite, and tridymite. Quartz is the most common type, and is a major component of rocks including granite, slate, and sandstone. Granite contains about 30 percent free silica, slate about 40 percent, and sandstone is almost pure silica.[4] Silicosis, also known as grinder's disease or potter's rot is one of the ancient diseases encountered to mankind and has been described as early as the 18th century.[1] Silica exposure can not only result in silicosis, but a number of conditions including chronic bronchitis, emphysema, COPD, tuberculosis (2-30 fold increased risk), Caplan's syndrome, lung cancer, and some immune mediated diseases like systemic lupus erythematosis.[5,6]

Silicosis may present in many forms depending on the latency period including simple silicosis, acute silicoproteinosis (which resembles pulmonary alveolar proteinosis), accelerated silicosis and progressive massive fibrosis (complicated chronic type).[1,2,5] Chronic silicosis develops slowly, usually appearing 10 to 30 years after exposure to low dust concentrations and accelerated silicosis having similar features to chronic silicosis develops within 10 years and is associated with high-level exposures.[1,5] Chest radiology plays a crucial role in the diagnosis of various forms of silicosis. Radiographic features include small rounded opacities in simple or uncomplicated silicosis that are well defined 1 to 10 mm and uniform in shape and attenuation.[7,8] Silicotic nodules are usually symmetrically distributed and tend to occur first in the upper zones and later may involve other zones. Silicotic nodules, which are the pathologic hallmark of silicosis, may develop first in the hilar lymph nodes & may be confined to this area; they may become encased in egg shell calcification (although not pathognomonic, but is exclusively seen in silicosis) and may impinge or erode into the airways.[9] With disease progression in both chronic & accelerated silicosis, the nodules become confluent, over 1 cm leading to the development of progressive massive fibrosis (PMF), as was seen in the index case.[5] Acute silicosis is rare, and develops after overwhelming silica exposure resulting in symptoms within a few weeks to a few years.[10] There may be genetic factors behind the fact that some individuals develop acute silicosis while others develop accelerated form of silicosis after such heavy silica exposures in confined places like in denim jeans sand blasting.[11] The disease presents radiographically with a characteristic basilar alveolar filling pattern, *without* rounded opacities or lymph node calcifications.[10,11] In contrast, most individuals with extremely high silica exposures initially display radiographic features identical to those of simple silicosis, which progress rapidly to PMF over a period of as few as four to five years.[5,10]

Three key elements are required for a clinical diagnosis including a compatible history of silica exposure & the appropriate latency from the time of first exposure, chest imaging that shows opacities consistent with silicosis and an absence of another diagnosis more likely to be responsible for the observed abnormalities.[1] Spirometry shows an obstructive pattern, as was in the index case, followed by a restrictive pattern in advanced disease, with reduced DLCO.[12] Only in patients with

atypical features of the disease like unilateral PMF, bronchoalveolar lavage (BAL) and/or transbronchial lung biopsy (TBLB) or open lung biopsy may be considered. BAL may show silica particles, as well as a preponderance of neutrophils; a lung biopsy reveals the characteristic silicotic nodules and may also reveal an alternative diagnosis or complication like TB or lung cancer.[13]

Silicosis is an irreversible condition with no cure. The disease will generally progress even without further exposure, but the rate of deterioration is probably reduced.[2] Management is aimed at stopping further exposure and change of profession, smoking cessation and prompt treatment of concomitant mycobaterial infection. Experimental treatments like whole lung-lavage, parenteral administration of a polymer, polyvinyl pyridine N-oxide (PVNO); inhalation of aluminum and administration of tetrandrine may be tried.[14,15]

In low-income countries like Pakistan, new cases of silicosis and associated lung cancer, COPD and TB are likely to be seen for decades because necessary reduction of silica use will take time to be achieved. The index case was a young person from Gujranwala who ended up in permanent disability due to chronic silicosis. According to him, he was completely unaware of the hazards associated with his dusty occupation and there are many similar work places in his area. In screening programs of construction workers, silicosis may be underestimated due to less sensitivity of simple radiographs in detecting early disease in comparison to chest CT scans. [16,17] There is a worldwide reduction in silicosis-related deaths over the past few decades.[1,16] Unfortunately, there is paucity of data regarding its prevalence and incidence in our setup. Economic trends have skewed from agriculture to industry in the recent years and newer industries with unknown occupational hazards are emerging. Hence effective measures must be taken to increase awareness of silica-related occupational exposures among the factory workers, health care providers and the community in general.

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