Progressive Massive Fibrosis in Workers Outside the Coal Industry: A Case Series from New Mexico

Landon Casaus, MD1
Sapna Bhatia, MD1
Akshay Sood, MD, MPH1, 2

1Department of Internal Medicine
University of New Mexico School of Medicine
Albuquerque, NM, USA
2Miners’ Colfax Medical Center
Raton, NM USA

Abstract

Four clinical patterns of diffuse lung disease may be seen with silicosis: acute silicosis or silicoproteinosis (the latter resembling pulmonary alveolar proteinosis), simple nodular sclerosis, accelerated silicosis, and progressive massive fibrosis (PMF). The intensity and duration of exposure as well as host susceptibility dictates the presentation and progression of PMF. Although most cases of PMF in the literature are reported among coal miners in whom this disease has shown a recent increase in prevalence, this disease can also be seen in exposed workers outside the coal industry. In this article, we will review the clinical, physiological, and pathological manifestations of the disease, illustrated by three case examples of PMF among non-coal miners from New Mexico. Diagnosis and management of patients with PMF can be difficult, and carries medicolegal implications for the patient. Physicians and policymakers need to be aware of PMF in workers exposed to silica within and outside the coal industry.

Introduction

The worldwide prevalence of silicosis peaked by the beginning of the 20th century during the development of mechanized industry (1). Outbreaks of silicosis are still noted in the developed world, particularly where workers are consistently exposed to silica particles that are small enough to be inhaled (≤10 µm in diameter) and at levels above a “safe” concentration (action level of 25 µm/m³ as a time-weighted average over an 8-hour work day, as recommended by the U.S. Occupational Safety and Health Administration or OSHA) (2,3). The four Appalachian coal mining states of Pennsylvania, West Virginia, Virginia, and Kentucky accounted for more than 75 percent of all silicosis-related deaths in the United States (U.S.) in 2007 (4). A recent study however indicates that the age-standardized mortality rate from silicosis in the U.S. in 2014 was amongst the highest in the mining intense regions of the Southwest, particularly in the Four Corners area where the borders of New Mexico, Arizona, Utah, and Colorado meet (5). The number of diagnosed silicosis cases has increased in New Mexico between 2000 and 2011, and residents of New Mexico are twice as likely to die from or with silicosis when compared to the rest of the country for reasons that are unexplained (6).
Four clinical patterns of diffuse lung disease may be seen with silicosis: acute silicosis or silicoproteinosis (the latter resembling pulmonary alveolar proteinosis), simple nodular sclerosis, accelerated silicosis, and progressive massive fibrosis (PMF). PMF represents the coalescence of multiple small pneumoconiotic opacities to form larger opacities or conglomerate masses measuring over 10 millimeters in size on a chest radiograph, with smaller rounded opacities usually seen in simple silicosis. Silicotic opacities are classified on their shape, size, and profusion using the International Labour Organization’s (ILO) International Classification of Radiographs for Pneumoconiosis system (commonly referred to as B reads) (7-9). The 1970-2017 radiographic data from the National Institute for Occupational Safety and Health (NIOSH) surveillance program concluded that the national prevalence of coal workers’ pneumoconiosis in coal miners with 25 years or more of tenure now exceeds 10% (10). This is an increase from the previous estimate of 7% in 2012 (11,12). A resurgence of progressive massive fibrosis in coal miners has also been described, particularly those working in smaller mines (13). The rate of PMF in silica exposed workers outside of the coal mine industry, similar to those illustrated in this paper, is unknown. We herein describe three New Mexico non-coal miners with PMF that were followed at the University of New Mexico Occupational Pulmonary Medicine Clinic. Each of the three cases had already received compensation under the United States Energy Employees Occupational Illness Compensation Program, based upon prior abnormal B reads of chest radiographs. The epidemiology, pathogenesis, and management of PMF is also reviewed.

Case reports

Case 1

An 83-year-old man presented in 2017 with worsening dyspnea over the prior 10 years. He worked at a federal national laboratory in northern New Mexico, from 1962-1992 as a construction worker. His work included digging ditches, removing insulation, demolishing buildings, breaking up concrete with jackhammers, and working around sandblasters in enclosed areas, without any respiratory protection. He had a 5-pack year smoking history, and quit 50 years prior.

A 2017 chest radiograph showed small, upper lobe predominant, nodular opacities. A high-resolution computed tomography (CT) scan in 2009 showed innumerable micronodules in the upper lobes of the lung with a centrilobular distribution. A repeat CT scan obtained in 2017 (Figure 1) showed new-onset coalescence of several upper lobe nodules, as large as 1.5 cm x 2 cm.
His pulmonary function tests (PFT) showed mild obstruction with evidence of air trapping. A diagnostic bronchoscopy showed no evidence of infection or neoplasm.

Case 2

A 78-year-old man presented in 2014 with several-years history of progressive New York Heart Association Class III dyspnea. The patient worked as an underground uranium miner from 1960 to 1989 where he was exposed to hauling, “mucking” (a term referring to the loading of fragmented ore), and blasting. He wore a respirator intermittently. He had a five-pack year smoking history, quitting in 1981.

Chest x-ray showed innumerable micronodules, predominately in the upper lobes. A CT scan of the chest with 3 mm cuts in 2012 showed innumerable upper lobe predominant micronodules in a perilymphatic and centrilobular distribution, with coalescence in the upper lobes. Repeat CT scans in 2014 and 2015 demonstrated no disease progression (Figure 2).

Figure 2. Computed tomography scan of the chest demonstrating progressive massive fibrosis in the right upper lung and several silicotic opacities in bilateral upper lungs.
PFTs showed a mild restrictive defect. An infectious etiology was ruled out by negative sputum acid fast bacilli (AFB), and bacterial smears and cultures.

**Case 3**

A 79-year-old man presented with dyspnea at rest and upon exertion, and chronic bronchitis symptoms, with occasional hemoptysis. The patient worked as an underground uranium miner from 1959-1980 performing drilling, blasting and “mucking”, with significant self-reported exposure to dust and without use of respiratory protection. The patient reported a 15-pack year smoking history, but quit in 1976.

A chest x-ray showed hilar and mediastinal nodal calcifications with small scattered lung nodules. A HRCT scan of the chest in 2016 (Figure 3) showed multiple calcified nodules as well as calcified hilar and mediastinal lymph nodes.

![Computed tomography scan of the chest demonstrating progressive massive fibrosis with evidence of traction in both lobes.](image)

**Discussion**

PMF is seen in workers employed in industries that cut, grind, or drill silica-containing materials such as concrete, masonry, tile and/or rock (3). Most cases of PMF in the literature have been reported among coal miners, likely a reflection of the fact that coal miners undergo active surveillance due to governmental regulations (12). Although more commonly believed to occur in underground coal miners, PMF can be seen in surface coal miners as well (14). PMF outside the coal industry has been described in limited studies of barium miners, sandblasters, blacksmiths, welders, metal polishers, and quartz surface fabricators (15-17). More recently, PMF has been reported in
‘distressed’ denim jean industry workers (18). In this case series, we report PMF in New Mexico construction and uranium workers.

The latency for PMF is usually 10-30 years. Latency is greatly impacted by the exposure concentration and duration, as well as type of silica exposure. Additionally, it is influenced by underlying diseases, genetics, and smoking. Although PMF typically occurs in a setting of high cumulative dust exposures (14), some studies indicate that the host patterns of deposition and clearance of dust may be more relevant (19).

The pathogenesis of PMF is not completely understood; however, it is known that alveolar macrophages initiate a complex cascade results in inflammation and fibrosis (20). Histopathological findings include nodules, usually located near the respiratory bronchioles, composed of silica particles surrounded by whorled collagen in concentric layers. Larger masses of collagen define the lesion of PMF, which may be associated with avascular necrosis in the center and endarteritis in the periphery (21). The extensive fibrotic reaction in PMF is associated with high serum levels of interleukin (IL)-8 and intercellular adhesion molecule (ICAM)-1, which are important as neutrophil attractants and adhesion molecules (22).

The clinical diagnosis of PMF has three requirements: the patient must have a history of inhalational silica exposure significant enough to cause disease; chest imaging must be consistent with PMF; and other illnesses that mimic PMF must be reasonably ruled out (1). The disease presentation of PMF is highly variable. Patients may have debilitating symptoms of dyspnea on exertion and exercise intolerance, obstructive and/or restrictive patterns on PFTs, as well as experience complications such as cor pulmonale, spontaneous pneumothorax, and hypoxic respiratory failure (23). On the other hand, a normal spirogram is described in up to 11% of subjects with PMF, as also noted in Case 3 above (23). The level of pulmonary impairment in patients with PMF generally increases with increasing radiologic size of large opacities (23). Spirometry is repeated upon follow-up visits to assess for functional deterioration (24). Invasive tests such as arterial blood gas or cardiopulmonary exercise test are usually not indicated. Surveillance chest radiographs are classified for small and large opacities using the International Labour Organization’s (ILO) International Classification of Radiographs for Pneumoconiosis system (7-9). CT scan of the chest is more sensitive in diagnosing PMF than chest radiographs, and may be considered if the radiograph fails to show large opacities but demonstrates small opacities of relatively larger diameter or a tendency for opacities to coalesce (25,26). Lung tissue for histology or mineral analysis is rarely needed. The presence of atypical features in a patient with simple silicosis such as fever, hemoptysis, worsening dyspnea, weight loss, disproportionate fatigue, and the presence of a new infiltrate or cavitation of a pre-existing lesion on chest imaging should prompt the clinician to look for PMF, tuberculosis, or lung cancer. Patients with PMF are at elevated risk for concomitant tuberculosis. This risk is directly proportional to the level of profusion of silicotic small opacities (27), and the risk in patients with the highest level of profusion is comparable to that in patients with HIV infection (3). Autopsy studies from Welsh coal workers during the period 1952–1954 demonstrated tubercle bacilli in as many as 35% of cases with PMF (21). A recently published study from Brazil reported
coexisting microbiologically confirmed tuberculosis in about half of patients with PMF, raising concerns about tuberculosis infection as a risk factor for the development of PMF (15). Patients with silicosis are also at high risk for lung cancer (28), with a greater risk for lung cancer described in patients with PMF as compared to patients with simple coal workers pneumoconiosis in one study (29). Positron emission tomography with F-18 fluorodeoxyglucose is of limited utility in differentiating malignancy from PMF lesions (30).

The prevention of PMF remains a focus at the exposed workplace. This includes primary prevention such as worker education; control of airborne dust exposure via engineering and work practice interventions such as improving ventilation, providing a means of exhaust, adding water to the cutting surface, and using enclosed cabs or booths; and use of respiratory protective devices (3). In June 2018, OSHA mandated personal breathing zone air sampling to monitor exposure and medical surveillance of workers with exposure above the permissible exposure limits (31). Medical surveillance constitutes secondary prevention, facilitating early diagnosis and treatment. Surveillance should be done periodically and should include a medical examination and occupational questionnaire, chest radiograph with B read interpretation, tuberculosis screening, and spirometry, with referral of affected workers to a pulmonologist or occupational medicine physician for further evaluation (32).

Once PMF has been diagnosed, it is important to immunize the patient against influenza and pneumococcal infection, assess the need for oxygen supplementation, and encourage pulmonary rehabilitation. Exclusion of active tuberculosis is recommended and screening for latent tuberculosis infection by either skin testing or interferon gamma release assay should be considered (33). Systemic corticosteroids, inhaled aluminum citrate, poly(vinlypyridine-N-oxide) and whole lung lavage are unlikely to benefit patients with PMF and lung transplantation may be considered (4).

Patients with PMF are considered ‘totally disabled’ from coal mine employment under the Black Lung Benefits Act in the United States. Outside the coal industry, they may be eligible for benefits under the Social Security Impairment system or the state workers’ compensation systems.

**Conclusion**

PMF represents the coalescence of smaller radiographic pneumoconiotic opacities to those over 10 millimeters in size. The rate of PMF in American coal miners has recently increased. Although most cases of PMF are reported among coal miners, this is likely a reflection of the fact that coal miners undergo active surveillance due to governmental regulations. In this case series, we report PMF in workers outside the coal industry. Physicians and policymakers need to be aware of this condition in workers exposed to silica within and outside the coal industry.
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Abbreviations List

AFB: Acid fast bacilli
CT: computed tomography
HIV: Human immunodeficiency virus
HRCT: High resolution computed tomography
IL: interleukin
ICAM: Intercellular adhesion molecule
NIOSH: National Institute for Occupational Safety and Health
OSHA: Occupational Safety and Health Administration
PMF: Progressive massive fibrosis
PFT: Pulmonary function test
US: United States

References
