Pathologic Quiz Case

Recurrent Spontaneous Pneumothorax in an Industrial Worker

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A 38-year-old man experienced progressive shortness of breath with spontaneous pneumothoraces 2 times, which required chest tube placements and hospitalization for medical management. A computed tomographic (CT) scan of the chest performed at the time of the second chest tube placement demonstrated a small, dependent, right pleural effusion with adjacent consolidation, mild pleural thickening near the left base, and prominent interstitial markings at both lung bases (Figure 1). Past medical history was significant for a 20-pack-year history of cigarette smoking and an occupation as an industrial arc welder for 15 years. An open lung biopsy was performed at the time of the second chest tube placement to evaluate the cause of the interstitial lung disease.

Histologic sections of the open lung biopsy demonstrated patchy areas of consolidation secondary to a marked increase in the number of alveolar macrophages with a bronchocentric-angiocentric distribution. These macrophages contained abundant cytoplasmic, coarse, brown-black foreign particles (Figure 2) and abundant birefringent pointed spicules, 5 μm or less in length (Figure 3). Prussian blue stain revealed abundant hemosiderin particles in these same macrophages (Figure 4), with absence of staining in the uninvolved portions of the lung parenchyma.

What is your diagnosis?
Pathologic Diagnosis: Mixed Dust Fibrosis in an Arc Welder (Pulmonary Siderosilicosis)

Pulmonary siderosis may result from the exposure to inert metallic iron or iron oxides by arc welders (arc welder’s lung), iron workers, and hematite miners.1 Irregular, coarse, brown-black particles of iron oxides and variable numbers of hemosiderin particles accumulate in alveolar macrophages in the lung parenchyma with a perivascular distribution. In extreme cases, the lung parenchyma may even assume a rust-to-brown coloration on gross examination. Bronchopulmonary lavage has been suggested as a potential method to reduce the level of siderosis in selected cases.2

When exposure occurs in conjunction with silica dust, a form of mixed dust fibrosis called siderosilicosis can result. In contrast to isolated siderosis, siderosilicosis may be associated with the development of significant interstitial fibrosis. Coexistent cigarette smoking contributes to and accelerates the lung injury and pulmonary function compromise.3

The clinical, radiographic,4 and progressive pulmonary functional defects of mixed dust fibrosis or siderosilicosis may be similar to those observed with silicosis. Histologically, however, the progressive development of stellate interstitial fibrous lesions is observed with mixed dust fibrosis,1 in contrast to the classic silicotic nodules that occur with silicosis. The stellate lesions are small, discrete, and centered around respiratory bronchioles and adjacent small arteries, with intervening uninvolved lung parenchyma present. The severity of pulmonary compromise generally corresponds to the extent of these lesions.

When compared with the general population, welders are at an increased risk for developing lung cancer.5-7 Tobacco use and exposure to asbestos are relatively common among these workers and contribute to this risk.

The isolated carcinogenic potential attributed to various nonasbestos dust and metallic particles in the lung remains controversial and is the subject of ongoing scientific investigation.

Arc welders may also experience acute respiratory distress secondary to exposure to metal fumes, called metal fume fever.8 Onset of symptoms usually occurs several hours following exposure, consisting of low-grade fever, nonspecific muscle pains, and intense shaking chills. Radiographic studies typically demonstrate bilateral, patchy, interstitial infiltrates, compatible with acute respiratory distress syndrome. Open lung biopsy reveals focal, mild interstitial pneumonia, without evidence of an infectious or immune process. With mild exposure, symptoms are self-limited and resolve spontaneously. Intense medical support, including mechanical ventilation, may be required for severe cases.

Pneumothorax is a common complication of pulmonary disease or injury. Spontaneous pneumothorax, however, often occurs in the absence of significant pulmonary disease.9 Spontaneous pneumothorax is generally due to the rupture of subpleural blebs located at the apex of the upper lobe or in the superior segment of the lower lobe. The origin of these blebs is unclear. Chest tube placement to ensure adequate re-expansion and sealing of the air leak is generally required. For patients with recurrent pneumothorax, surgery to remove, oversew, or staple any residual blebs may be indicated.

In this patient, the spontaneous pneumothoraces were believed to be due to the rupture of an emphysematous apical bleb on the pleural surface, unrelated to the coexistent pulmonary disease. The siderosilicosis was an incidental finding, which was initially suggested by a chest x-ray film to evaluate the second pneumothorax and then confirmed by the open lung biopsy.

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References