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A Bone to Pick with a Lung Disease

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Case Vignette

A 48-year-old smoker, paraplegic since the age of 20 years from a motor vehicle accident, was admitted with symptomatic nephrolithiasis for planned lithotripsy and percutaneous nephrostomy. He did not report respiratory symptoms. He had no known history of lung disease. He was born in the United States and had not traveled internationally. There were no occupational exposures, as the patient had never been employed. At the time of his accident, he required prolonged mechanical ventilation via tracheostomy but was subsequently decannulated. His oxygen saturation on admission was 93%. Lung auscultation revealed bibasilar dry crackles. Routine laboratory evaluation, including serum levels of creatinine, calcium, and phosphate, was unremarkable. There were no pulmonary function tests available for review. Echocardiography demonstrated normal left ventricular function and structurally normal heart valves. His preoperative chest radiograph is shown in Figure 1. Subsequent computed tomographic (CT) imaging of the chest is shown in Figure 2.

Questions

1. What are the abnormal findings on the chest radiograph (Figure 1)?
2. What abnormalities are apparent on the provided images from the chest CT examination (Figure 2)?
3. What is the most likely diagnosis?

[Continue onto next page for answers]
**Discussion**

The incidentally discovered chest radiograph abnormalities in this case consisted of bilateral loss of lung volume and diffuse reticulonodular opacities with areas of confluence—most notable in the upper lung zones—as well as areas of high attenuation suggestive of parenchymal calcification (Figure 1). Initial diagnostic considerations on the basis of that chest radiograph appearance centered on diffuse fibrotic lung diseases associated with upper lung zone involvement and calcification, among them sarcoidosis and pneumoconiosis, most notably silicosis. Subsequently, a chest CT scan revealed predominantly upper lobe emphysematous changes (Figure 2A, red arrows). Additional findings, primarily in the lower lung zones, were interlobular and intralobular septal thickening (Figure 2B, red arrows) as well as traction bronchiectasis (Figure 2C, red arrows), suggesting the presence of a fibrotic interstitial lung disease other than usual interstitial pneumonia. Strikingly, the areas of high attenuation on chest radiograph corresponded to an extensive bilateral network of mixed coarse and fine branching calcifications in the lung interstitium (e.g., Figure 2A, green arrows). Attenuation measurements of the calcified lung parenchyma were similar to that of the patient’s skeleton, which is best appreciated on bone window (Figure 2D). The radiological picture of chronic lung disease interlaced with widespread calcification led to the clinical diagnosis of a rare phenomenon known as diffuse pulmonary ossification.

**Calcium Deposition in the Lung**

The lung is a common site of calcium deposition, whether in the form of simple calcium salts, known as calcification, or as mature bone, termed ossification. The distinction between these two entities in the lung is one of histopathology rather than radiology, and calcification occurs far more frequently. Pulmonary calcification can be broadly categorized as metastatic or dystrophic. As the name suggests, metastatic processes are those originating outside the lung and involving it secondarily. The lung in these cases is usually otherwise structurally normal. Benign entities within this category reflect a systemic disturbance of calcium homeostasis that accompanies, for example, chronic renal failure or primary hyperparathyroidism. A number of cancers, most notably multiple myeloma and sarcoma, are capable of producing malignant metastatic pulmonary calcifications. The more common category of dystrophic calcification includes primary lung damage in which calcification is part of aberrant repair. Among the typical conditions in this category are infectious and noninfectious granulomatous processes (e.g., tuberculosis, histoplasmosis, sarcoidosis), amyloidosis, and healed varicella pneumonia.

**Diffuse Pulmonary Ossification**

Pulmonary ossification is an uncommon and indolent process defined as heterotopic bone formation in lung tissue with or without marrow elements. Since the original description by Luschka in 1856, this disease process has been further divided into two categories—nodular and dendriform—with the recognition that overlap cases likely exist. The nodular form is characterized by lamellar bone deposition in the alveolar spaces. It is usually a sequela of chronic passive pulmonary vascular congestion, as might occur in heart failure or mitral stenosis. The dendriform type refers to branching spicules of bone involving the interstitium and alveolar septa, with only sporadic erosion into alveoli. This entity, to which our case is most analogous, is associated with chronic lung diseases such as emphysema and pulmonary fibrosis, although idiopathic occurrence has also been described. The pathogenesis of dendriform pulmonary ossification (DPO) is poorly understood but is believed to evolve from dystrophic calcification in a milieu of inflammation and fibrogenesis that is conducive to an imbalance between osteoblastic and osteoclastic activity. Given the rarity of DPO, incidence and prevalence figures are not readily available. As an example, review of 1,393 random autopsies by Lara and colleagues identified eight cases (0.6%).

The most common patient group affected by DPO appears to be men older than 60 years of age, which could simply reflect the demographic profile of the typical accompanying lung diseases. The respiratory symptoms, if any, of those with DPO are usually attributable to the underlying lung disease. There are no specific laboratory abnormalities associated with this condition, and pulmonary function testing is again reflective of the primary lung pathology. On plain chest radiography, isolated DPO may manifest as a very subtle increase in interstitial markings, and the presence of calcification may not be apparent. If DPO is associated with an underlying parenchymal disease, which is the norm, then the manifestations of that disease may dominate the radiographic picture. Chest CT imaging is much more informative in DPO, revealing a lacework of subcentimeter densities within the interstitium that correspond to the Hounsfield unit range of bone. In most cases, a coexistent chronic lung disease will be demonstrated by CT imaging. In fact, the finding of DPO in conjunction with diffuse fibrotic lung disease has been suggested as a distinguishing feature of usual interstitial pneumonia from nonspecific interstitial pneumonia. Although technetium 99 m-methyl diphosphonate bone scintigraphy would be expected to identify areas of bone formation in DPO, it is unlikely to add substantively to the impression based on thin-section CT scanning.

In the setting of the characteristic imaging abnormalities described above, the diagnosis of DPO can be established without histological confirmation. Most reports in which tissue sampling was performed describe the findings obtained on surgical lung biopsy, although bronchoscopic lung biopsy has also been diagnostic in selected cases. Histopathology reveals bone formation, with or without marrow elements, with surrounding interstitial inflammation. The indolent nature of this process obviates the need for further surveillance, and management is based on the underlying lung disease. No interventions are currently known to interrupt the pathogenesis of DPO.

**Follow-Up**

Our patient underwent the planned urological procedure without any complications and was discharged home after extensive counseling on the importance of smoking cessation. Pulmonary function tests were not obtained before discharge. Interestingly, the chemical analysis of his kidney stones yielded calcium phosphate (carbonate apatite), which is the crystal component of bone.
Answers

1. What are the abnormal findings on the chest radiograph (Figure 1)?

The chest radiograph demonstrates symmetrical loss of lung volume with bilateral coarse interstitial opacities, including areas of high attenuation suggestive of calcium deposition.

2. What abnormalities are apparent on the provided images from the chest CT examination (Figure 2)?

Chest CT coronal reconstruction (Figure 2A) set to lung window demonstrates upper lobe–predominant emphysema (red arrows) with extensive interstitial calcifications exhibiting both coarse and fine branching (green arrows). In addition, areas of interlobular and intralobular septal thickening (Figure 2B, red arrows) as well as traction bronchiectasis (Figure 2C, red arrows) are present in the lower lung zones and are also associated with the same calcification pattern. An axial image set to bone window (Figure 2D) highlights the extreme density of the calcifications, which is comparable to that of the patient’s osseous structures (representative density value range of 600–1,000 Hounsfield Units).

3. What is the most likely diagnosis?

Diffuse pulmonary ossification complicating chronic lung disease.

Recommended Reading

Chan ED, Morales DV, Welsh CH, McDermott MT, Schwarz MI. Calcium deposition with or without bone formation in the lung. Am J Respir Crit Care Med 2002;165:1654–1669.


