Case Report

Cicatricial Organizing Pneumonia with Dendriform Pulmonary Ossification: An Unusual Cause for a Recurrent Pneumothorax

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Cicatricial organizing pneumonia is an uncommon form of organizing pneumonia, which may manifest as persisting linear opacities on computerized tomography (CT) scan mimicking a fibrosing interstitial pneumonia. It may also manifest with pulmonary ossification, which is a metaplastic bone formation within the lung tissue. The latter presentation could be either nodular or dendriform, both secondary to underlying lung disease and rarely idiopathic. Dendriform pulmonary ossification (DPO) has rarely been described as a cause of spontaneous pneumothorax. We present a case of a 55-year-old male with history of recurrent pneumothoraces and worsening dyspnea on exertion. A CT of the chest revealed progressive bilateral sub-pleural and peribronchovascular reticular opacities associated with densely ossified branching and nodular opacities. Video-assisted thoracoscopic biopsy of the lung demonstrated cicatricial organizing pneumonia with areas of marked diffuse DPO. The case highlights that dendriform pulmonary ossification arising from cicatricial organizing pneumonia should be considered in the differential diagnosis of recurrent pneumonias among patients with lower lobe sub-pleural reticular opacities. The case highlights that dendriform pulmonary ossification rarely can cause spontaneous pneumothorax and can be associated with cicatricial organizing pneumonia and reticular opacities on imaging.

1. Introduction

Organizing pneumonia (OP) is characterized by the presence of organizing fibromyxoid proliferations within the lumens of respiratory bronchioles and alveolar ducts. Peripheral and/or peribronchiolar consolidations are the most frequent findings of OP on a computerized tomography (CT) scan mimicking a fibrosing interstitial pneumonia. OP is often steroid-responsive and reversible with total resolution of radiological opacities, but occasionally may recur [1, 2].

Cicatricial OP (OPc) is a newly described entity in the pathology literature [3, 4] and distinguished from conventional OP by formation of irreversible dense fibrous bands and small nodules in the background of conventional OP [4]. The reported CT findings of OPc are variable and range from typical imaging findings of OP to lower lobes predominant peribronchovascular and sub-pleural reticular opacities with or without pulmonary ossification [4]. The latter is characterized by metaplastic ossification in the lung and is classified into nodular (NPO) and dendriform (DPO) subtypes [5]. Dendriform pulmonary ossification is usually described in association with chronic lung disease including usual interstitial pneumonia (UIP) [5, 6] although it can be seen in isolation [7, 8]. Dendriform pulmonary ossification presenting with spontaneous pneumothorax has been reported in a few occasions [9–14] but rarely with OPc [4]. We report a case of cicatricial organizing pneumonia with DPO who presented with recurrent pneumothorax. The clinical, radiological and pathological findings as well as the pulmonary functions have been described.

2. Case Report

A 55-year-old, nonsmoker male presented with a recurrent large left pneumothorax requiring a chest tube placement. He had a same side pneumothorax three and six years earlier, also requiring a chest tube placement. The patient also reported
Table 1: Pulmonary function test over several years.

| Year | FEV1 Value | Predicted % | FVC Value | Predicted % | FEV1/FVC % | DLco Value | Predicted % |
|------|------------|-------------|-----------|-------------|------------|------------|-------------|
| 2019 | 2.22       | 55%         | 3.01      | 57%         | 74         | N/A        | N/A         |
| 2017 | 2.62       | 69%         | 3.77      | 76%         | 70         | 20.74      | 68%         |
| 2015 | 2.96       | 76%         | 4.11      | 82%         | 72         | 24.76      | 80%         |
| 2014 | 3.15       | 81%         | 4.16      | 82%         | 76         | 24.76      | 80%         |
| 2013 | 3.41       | 87%         | 4.47      | 88%         | 76         | 23.61      | 76%         |
| 2012 | 3.39       | 85%         | 4.50      | 88%         | 75         | 32.39      | 103%        |

FVC: forced vital capacity, FEV1: forced expiratory volume in 1 second, DLCO: diffusing capacity of the lung for carbon monoxide (ml/min/mmHg), N/A: not available.

3. Discussion

Organizing pneumonia is usually steroid-responsive and pathologically characterized by presence of loose fibromyxoid plugs within the lumens of the respiratory bronchioles and alveolar ducts [1, 2]. Cicatricial OP is distinguished from conventional OP by formation of irreversible dense fibrous bands and small nodules in a background of small or large foci of conventional OP [4]. In OPc, fibromyxoid plugs as seen in OP may be seen transitioning to more hyalinizing fibrous band/nodule of cicatricial OP and into foci of ossification on imaging as well as on pathology [4]. In our case, foci of DPO consisting of islands of ossifying fibrosis were present throughout both the left upper and lower lobes. In addition, there were scattered foci of hyalinizing/cicatrical type organizing pneumonia [4], representing a more chronic form of organizing pneumonia. Isolated focal plugs of hyalinized intra-alveolar tissue can be incidentally seen on lobar resections for unrelated primary indications and should not be confused with OPc. OPc tends to show bilateral and diffuse or patchy distribution on imaging [3, 4]. Fibrosing OP is a different from OPc, though it is not clearly determined what this pattern may represent. It may be form of disease where the fibrous tissues cause expansion of the alveolar septal interstitium such as in late organizing stages of diffuse alveolar damage or fibrotic forms of nonspecific interstitial pneumonia (NSIP) [3].

On CT, conventional OP presents as patchy bilateral peribronchial and subpleural consolidations [15–19], which may be migratory [20]. The classic “atoll” [20] or the “reverse halo” sign [18] is only seen in 20% cases manifesting as ground glass opacities with surrounding crescentic or ring-shaped consolidation [21]. Perilobular opacities bordering the periphery of the secondary pulmonary nodules are observed [16, 19, 22] in more than half of OP cases. Manifestations of OPc include ground glass opacities, often associated with consolidations [15] and 1–10 mm nodular opacities with consolidation [16, 17]. Less common imaging features are subpleural or peribronchial irregular reticular opacities with areas of consolidation [15, 16, 19] and large nodules or mass-like consolidation [23]. Only 40% of the reported OPc cases show typical imaging appearance and other cases show variable nonspecific imaging findings including peribronchial and peripheral reticular opacities with or without evidence of pulmonary ossification [4]. Reticular opacities on CT imaging are seen in cicatricial OP cases where fibrous bands and nodules are the predominant feature on pathology with minor conventional OP. Presence of branching high densities on CT is suggestive of dendriform pulmonary ossification.

Pulmonary ossification is a metaplastic process where mature bone is present in the alveolar interstitium and/or alveolar spaces. Pulmonary ossification is classified into DPO and NPO. NPO is usually a localized process of lamellar bone and can occur in the setting of chronic congestion as seen in mitral valve stenosis. Unlike DPO, NPO usually does not contain bone marrow (fat or hematopoietic cells) [5]. On CT, NPO manifests as lower lobe predominant small, often highly attenuating, centrilobular nodules that may coalesce [24]. In DPO,
Figure 1: Axial chest CT images using lung (a), (c) and bone (b), (d) windows show bilateral branching dense nodular opacities (arrows) with mild associated reticulation. Some of the nodule are high in attenuation and almost iso-dense to ribs on bone windows. Axial images using lung widow (e), (f) at the level of lung bases were obtained 5 years apart and show evidence of progression.

Figure 2: Large area of subpleural diffuse pulmonary ossification revealing lamellar bone with a more complicated pattern and areas of marrow elements including fat (arrowhead). Adjacent areas of cicatricial OP (arrows) are present (Hematoxylin and eosin; 12.5x).

Figure 3: Areas of OP (arrow) with adjacent foci of cicatricial OP transitioning to DPO (arrowhead) (Movat stain; 20x).
Spontaneous pneumothorax has been reported in several cases of DPO [9–14] including one case with cicatricial OP and DPO [4]. This case with cicatricial OP and DPO reported by Churg et al. [4] is likely the same case was reported by Tsai et al. [10]. In our case, foci of DPO were present in the subpleural areas, presumably causing recurrent pneumothoraces. It has been suggested that a subpleural sharp bony spicule may cause the pneumothorax by puncturing the visceral pleura [10, 13, 14]. None of the reported cases nor our case has other causes to explain the pneumothoraces, such as cystic or bullous lung disease, trauma, or bronchopleural fistula.

In summary, we present a case of cicatricial OP with DPO presenting with recurrent pneumothoraces and slow progressive pulmonary physiologic restrictive impairment. Cicatricial OP should be considered in the differential diagnosis of peribronchial or subpleural reticular opacities with DPO. Radiologists and clinicians alike should be aware of this newly described entity as distinct from other classical fibrosing processes, its potential association with DPO, and the presumed association between subpleural DPO and spontaneous pneumothorax.

**Conflicts of Interest**

The authors declare that they have no conflicts of interest.

**Acknowledgments**

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