Diffuse pulmonary ossification in early-stage idiopathic pulmonary fibrosis

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DESCRIPTION
Diffuse pulmonary ossification (DPO) is an uncommon condition that is characterised by metaplastic bone formation in the lung.1 Secondary DPO usually occurs in a background of fibrosing interstitial lung diseases (ILDs).2 Here, we report a case of early-stage idiopathic pulmonary fibrosis (IPF) with DPO, which was confirmed by surgical lung biopsy.

A previously well 76-year-old man was admitted to our hospital with diffuse abnormal shadows on a chest X-ray as part of a routine health check-up. Chest high-resolution computed tomography (HRCT) scan on admission showed subpleural fine reticulation with minimal bronchioloectasis without honeycombing (indeterminate for usual interstitial pneumonia (UIP) pattern) and multiple punctate lesions of high density suggestive of calcifications predominantly in the bilateral lower lobes (figure 1A). The same-level image with a bone window setting (width, 1500 HU; level, 200 HU) showed multiple small nodular and dendriform ossifications in the subpleural regions (figure 1B).

Coronal section of chest HRCT showed fine reticulation with subpleural distribution in the bilateral lower lobes predominance and numerous punctate linear, branching, dendritic calcifications, appearing mainly in areas of reticulation (figure 1C). Arterial blood gas analysis and pulmonary function test showed normal range. Examination of bronchoalveolar lavage fluid (BALF) revealed alveolar macrophages, 97.4%; lymphocytes, 2.2%; neutrophils, 0.4% and eosinophils, 0%, with normal total cells counts and a CD4/CD8 ratio of 1.0. Cultures of sputum and BALF were negative for fungal, bacterial or mycobacterial pathogens. We performed video-assisted thoracoscopic surgery (VATS) due to diagnosis of underlying a fibrosing ILD. At macroscopic findings, there were seen extensive parenchymal and subpleural grey-whitish ossifications (1 scale=1 mm). (B) The microscopic findings revealed multiple ossified nodules ranging from 0.1 to 1.5 mm in diameter within both the interstitium and the airspaces on a background of predominantly usual interstitial pneumonia pattern in the lower lobe (H&E stain) (scale bar=2 mm). (C) There were scattered irregular, branching and nodular ossification in the non-fibrotic areas (H&E stain) (scale bar=2 mm). (D) At higher magnification, marrow elements and osteoblasts were present (H&E stain) (scale bar=200 μm).

Figure 1 Chest high-resolution computed tomography (HRCT) images on initial visit. (A) Chest HRCT showed subpleural fine reticulation with minimal bronchioloectasis without honeycombing and multiple punctate lesions of high density suggestive of calcifications predominantly in the bilateral lower lobes. (B) The same-level image with a bone window setting (width, 1500 HU; level, 200 HU) showed multiple small nodular and dendriform ossifications in the subpleural regions. (C) Coronal section of chest HRCT showed fine reticulation with subpleural distribution in the bilateral lower lobes predominance and numerous punctate linear, branching, dendritic calcifications, appearing mainly in areas of reticulation.

Figure 2 (A) At macroscopic findings, there were seen extensive parenchymal and subpleural grey-whitish ossifications (1 scale=1 mm). (B) The microscopic findings revealed multiple ossified nodules ranging from 0.1 to 1.5 mm in diameter within both the interstitium and the airspaces on a background of dominantly usual interstitial pneumonia pattern in the lower lobe (H&E stain) (scale bar=2 mm). (C) There were scattered irregular, branching and nodular ossification in the non-fibrotic areas (H&E stain) (scale bar=2 mm). (D) At higher magnification, marrow elements and osteoblasts were present (H&E stain) (scale bar=200 μm).

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seems likely that triggers such as severe and extensive lung injury, or genetic predisposition may be involved. Hans et al described that predominant DPO with fibrosis of UIP pattern may differ from IPF in several clinical features, including extremely slow progressive disease behaviour and the absence of radiological honeycombing despite long-lasting disease. However, Egashira et al recently reported that DPO is common in patients with fibrosing ILDs and significantly more prevalent in patients with IPF than in those with other fibrosing ILDs. Therefore, we decided to make a diagnosis of idiopathic interstitial pneumonia on the basis of histopathological findings obtained by VATS. Eventually, DPO may be a useful CT sign for diagnosis of early-stage IPF and distinguishing IPF from other fibrosing ILDs.

Learning points

► Diffuse pulmonary ossification (DPO) may be a useful CT sign for diagnosis of idiopathic pulmonary fibrosis (IPF) and distinguishing IPF from other fibrosing interstitial lung diseases.
► DPO can be associated with a variety of disease severity in IPF.
► It is possible that predominant DPO with fibrosis of usual interstitial pneumonia pattern may differ from IPF but long-term observational studies of larger case series are required.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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