The unexpected clinical behavior of a-PAP

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A 42-year-old woman, smoker (20 pack/years) painter and sculptor exposed to dusts, diagnosed with pulmonary alveolar proteinosis (PAP) was referred to us on April-2019 for further evaluation and treatment with inhaled-granulocyte-macrophage-colony-stimulating-factor (i-GM-CSF) (Figure 1A, Figure 1B). Oxygen saturation on room air was 88% and DLCO 35% predicted. Meanwhile of the results of anti-GM-CSF titer and in relation of her moderately good clinical condition the patient was advised to quit

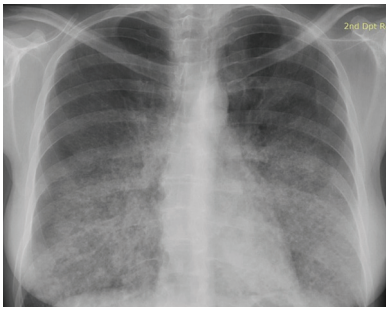

FIGURE 1a. In this posteroanterior (PA) chest radiography (CXR) confluent ground glass and consolidation specifically distributed in the pericardial and lower lung fields are detected. Note that the lateral costophrenic angles should be filled with pleuritic fluid if this was a common cardiac failure.

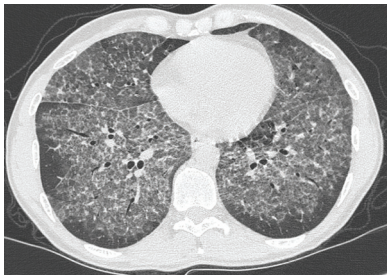

FIGURE 1b. In the synchronous high-resolution computerized tomography (HRCT) diffuse ground glass opacities not reaching the pleura are identified. There is also a diffuse interstitial involvement around the interlobular lines involved.

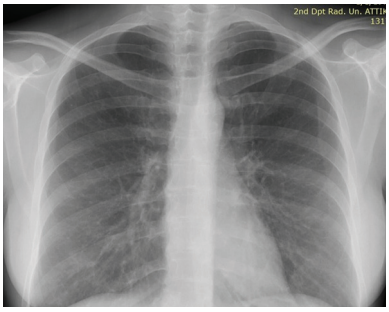

FIGURE 1c. In this PA CXR four months later the lung parenchyma looks normal.

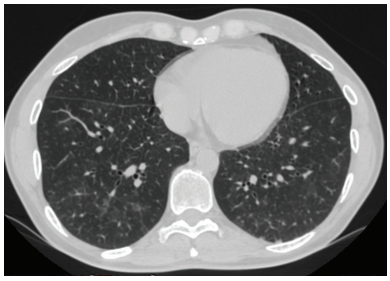

FIGURE 1d. The synchronous HRCT depicts a normal lung without significant scars from the previous insult except a minimal ground glass opacity in the superior segment of the right lower lobe.

smoking, avoid any harmful professional exposure and to initiate oral bromhexine syrup daily. Three months later anti-GM-CSF titer documented autoimmune-PAP (101.16 μg/ml). Upon reevaluation on July-2019 we made the serendipitous observation that all clinical, physiological and radiological parameters (Figure 1C, Figure 1D) of the disease were totally normalized and the disease was unexpectedly in complete "spontaneous" remission. Anecdotal reports as well as our personal experience underline the possible beneficial effect of bromhexine in PAP that may act as facilitator of washing-out stagnant surfactant[1-6].

Statement

This is to certify that the images have not been previously published and that the authors have no conflict of interest.

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