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CASE STUDY

Primary diffuse alveolar septal amyloidosis with multiple cysts and calcification

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Primary diffuse alveolar septal amyloidosis with multiple cysts and calcification. S. Ohdama, S. Akagawa, O. Matsubara, Y. Yoshizawa. ©ERS Journals Ltd 1996.
ABSTRACT: We report an extremely rare case of primary diffuse alveolar septal amyloidosis associated with multiple cysts and calcification. Development of multiple cysts may have resulted from fragile alveolar walls, as a consequence of amyloid deposition both on alveolar walls and around capillaries.
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Diffuse alveolar septal amyloidosis localized exclusively to the lung is extremely rare. We present a case of diffuse alveolar septal amyloidosis associated with multiple cysts and calcification.

Case report

A 54 year old woman presented with dizziness when she visited an otorhinolaryngologist in March 1989. She was diagnosed as having Ménière's syndrome and corticosteroids were initiated leading to rapid improvement. She was referred to our hospital in April 1989 for further evaluation of abnormal chest shadows when she was asymptomatic. The patient's past medical history revealed that she had been treated for pulmonary tuberculosis with para-aminosalicylic acid (PAS) and streptomycin for 1 yr at the age of 16 yrs. She had undergone a total hysterectomy for uterine myoma at the age of 50 yrs. She had never smoked. On physical examinations, no evidence of respiratory distress, hepatosplenomegaly, or macroglossia were disclosed.

Laboratory examination revealed a total leucocyte count of 6.2×10^9 cells·L^{-1} with a normal differential, and haemoglobin of 146 g·L^{-1}. Erythrocyte sedimentation rate (ESR) was 10 mm·h^{-1}. Urinalysis was normal, with negative Bence Jones protein. Blood biochemical examination were normal. A tuberculin skin test was positive (15 mm transverse diameter). Measurement of serum immunoglobulins revealed a slightly increased immunoglobulin G (IgG) of 15.9 g·L^{-1} and normal levels of immunoglobulin A and M (IgA and IgM) (3.28 and 1.46 g·L^{-1}, respectively). The following data were negative: C reactive protein, rheumatoid factor, antinuclear factor, anti-SS-A antibody, anti-SS-B antibody, parathyroid hormone. An electrocardiogram was normal. Sputum was negative for acid-fast bacilli.

Pulmonary function tests revealed normal forced vital capacity (FVC) of 2.49 L (105% of predicted), reduced forced expiratory volume in one second (FEV_1) of 1.61 L (65% pred), increased forced expiratory flow at 50% VC/25% VC (FEF_{50}/FEF_{25}) ratio of 4.0, a mildly increased residual volume/total lung capacity (RV/TLC) of 41%, and mild reduction in carbon monoxide transfer factor (78% pred). Blood gas analysis was within normal limits arterial carbon dioxide tension (P_{a,CO_2}) 5.2 kPa (39.1 mmHg), arterial oxygen tension (P_{a,O_2}) 10.7 kPa (80.0 mmHg), and mildly increased alveolar to arterial oxygen tension difference $P_{(A\text{-}a),O_2}$ of 3.2 kPa (24 mmHg).

Histological examinations of gastric mucosa obtained by gastroendoscopy were normal. Chest roentgenogram showed diffuse reticulonodular shadows and a fine stippled appearance in the middle and lower lungs (fig. 1).

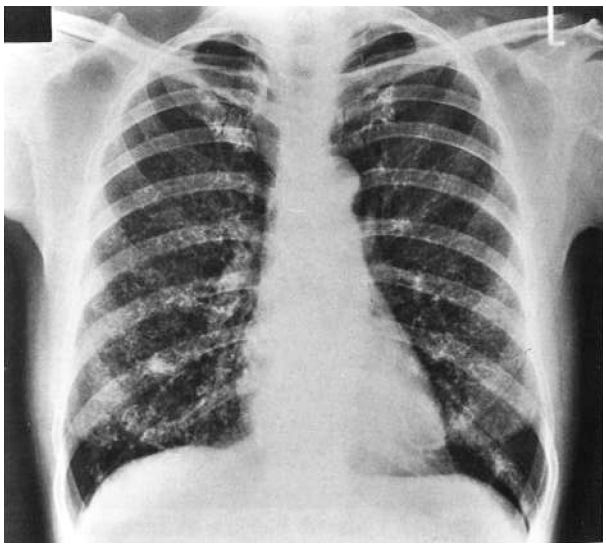

Fig. 1. – Chest radiograph demonstrating diffuse reticulonodular shadows and fine stippled appearance in the middle and lower lungs.