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Title: Secondary pulmonary alveolar proteinosis associated with IgG4-related disease

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Body: Pulmonary alveolar proteinosis (PAP) is a rare respiratory disease characterized by pulmonary surfactant-derived lipoproteins accumulation within alveolar spaces. PAP has been classified in 3 forms: autoimmune, secondary and unclassified. Until now, there has been no report supporting the association between secondary PAP(SPAP) and IgG4-related disease. Here, we report a first case of SPAP associated with IgG4-related disease. A 61 year old man of Indian ethnicity complained of shortness of breath. CT scan demonstrated diffuse ground-grass opacities in the lung, and cervical, mediastinum, axilla and inguinal lymphnode enlargement. Serum IgG4 concentration was elevated. IgG4-related disease was diagnosed by immunohistochemical findings of inguinal lymphnode that revealed the increase of IgG4-positive cells. SPAP was diagnosed by pathological findings of the transbronchial lung biopsy revealing the accumulation of eosinophilic amorphous material in the alveoli. Autoantibodies against granulocyte-macrophage colony-stimulating factor in sera were below the sensitivity. These results confirmed the diagnosis of SPAP. Bronchoalveolar lavage of the right upper lobe was performed, which resulted in the improvement of the lung lesion. Seven months after the diagnosis, he had acute interstitial nephritis which was suspected IgG4-related kidney disease. After steroid treatment, the renal function and the lung lesion were improved. We reached a diagnosis of SPAP associated with IgG4-related disease.