A case of pulmonary alveolar microlithiasis with Sjögren’s syndrome

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Pulmonary alveolar microlithiasis (PAM) is a rare and chronic disorder characterized by the accumulation of calcium phosphate precipitates in both lung alveolar spaces. Its etiology is unknown, usually seen in equal proportions in both sexes, and in almost every age (1,2). It is characterized by miliary pattern formed by 1-3 mm in size micronodules which tended to coalesce in middle and lower zones of both lungs. Although the final diagnosis of the disease is lung biopsy, bronchoalveolar lavage, sputum cytology and radiological diagnosis can also be made in some cases. Sjögren’s syndrome is an autoimmune disease characterized by keratoconjunctivitis sicca and xerostomia which affects the lacrimal glands and salivary glands. It may be associated with other autoimmune diseases, such as can be found isolated. We presented here a case that diagnosed with Sjögren’s syndrome with radiological diagnosis of PAM.

A 29-year-old male patient with 1-2 years of increasing shortness of breath, cough, and a small amount of sputum was admitted. He has smoked 30 pack-years. On physical examination, vital signs were stable, oxygen saturation is 95%. There were fine crackles bilaterally and rhonchi in some places. Bilateral diffuse micronodular opacities in the lower zones were observed in postero-anterior chest X-ray. In thorax computerized tomography, bilateral diffuse micronodular calcifications were detected intensively in the lower lobes of lungs. Mild restriction were present in pulmonary function test (FEV1: 63%, FVC: 73%, FEV1/FVC: 73). There was no abnormal finding was detected by echocardiography. Diffuse lung scan with technetium-99m methylene diphosphonate showed an increase in activity in both lungs. There was no calcification in his body other than the lungs. Bronchoscopy was planned with the diagnosis of PAM. Although a short-acting sedative had been given to him, the patient didn’t fit the operation, so the operation was failed. Sputum cytology had no evidence of PAM. The collagen tissue markers were performed for differential diagnosis of patient. Anti-SSA was positive and the patient has followed by rheumatology with the diagnosis of Sjögren’s syndrome. Analysis of the family (mother, father and two siblings) revealed no pathology. The patient with good performance status has been followed with symptomatic treatment.

PAM is a rare disease with unknown etiology. Hereditary, metabolic abnormalities, calcium-phosphorus metabolism disorders and abnormalities of the immune system are accused of the formation of PAM.
gene have been implicated (3). In the absence of the gene encoding the phosphate carriers of type II alveolar cells, phosphorus can’t be removed from intraalveolar cavity and the calcium-phosphate stones are formed. The most common symptoms are shortness of breath, cough, chest pain, and rarely hemoptysis. Our case has complained of dyspnea, cough, and a small amount of sputum for about two years. The disease progresses, causes pulmonary hypertension and respiratory failure. Pulmonary function tests are usually normal, advanced disease could provide restrictive dysfunction. Our case had a pulmonary function test with a slight restriction. Radiological imaging of the lung as in our case, there are micronodular calcifications tend to coalesce especially in the middle and lower zones of both lungs and paracardiac areas. The final diagnosis based on showing dilated alveoli in the lung tissue or cytological observations of calcium phosphate stones (4). However, in some cases due to the typical radiographic appearance diagnosis could be put with radiological findings. In our case, there were the typical features of the clinical and radiological findings of PAM. There was also accompanied by Sjogren’s syndrome. In order to notify, co-incidence of PAM and Sjogren syndrome firstly presented here.

CONFLICT of INTEREST
None declared.

REFERENCES