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Title: Clinical features common to five cases with secondary pulmonary alveolar proteinosis complicated with Behcet disease

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Body: Pulmonary alveolar proteinosis (PAP) is a rare lung disorder characterized by abnormal accumulation of surfactant materials in the lower respiratory tracts. It is classified into three distinct types according to etiology; autoimmune, secondary, and congenital PAP. Secondary PAP (SPAP) comprises ten percent of acquired PAP. Previously, we reported 40 cases of SPAP, in whom more than 70% occurred secondary to hematological disorders, with the majority being myelodysplastic syndrome (MDS). The present study focused clinical features of five patients (four female and one male) who developed PAP

during 6 months to 18 years after the onset of Behçet's disease (BD), with underlying trisomy 8-positive MDS in four of them. Oral and cutaneous BD lesions were involved in all cases, but ocular lesions were observed in only one case. Intestinal BD was recognized in three patients who had undergone potent immunosuppressive therapy that resulted in overwhelming sepsis. In the two surviving patients, PAP and BD were managed successfully, although both individuals had a high risk of MDS at diagnosis. Thus, we figure out the common and different points among SPAP patients complicated with BD. The differential diagnoses of SPAP should be ruled out when lung complications are encountered during the course of BD.