Secondary pulmonary alveolar proteinosis in acute myeloid leukemia

A 68-year-old woman was diagnosed with acute myeloid leukemia (AML) derived from a myelodysplastic syndrome and was treated with 17 cycles of azacitidine. Symptoms of high fever, general fatigue and hypoxemia appeared; therefore, she underwent chest computed tomography (CT) and a diffuse pulmonary shadow with a crazy-paving appearance was noted in her both lungs (Figure 1). Crazy-paving appearance is defined as a network of smooth line shadows in ground glass opacity by CT examination, and this appearance is frequently observed in patients with pulmonary alveolar proteinosis (PAP).\(^1\) Levels of interstitial pneumonia markers, such as serum krebs von lungen-6 (KL)-6, surfactant protein (SP)-D and SP-A, were elevated (1539, 161.3 and 145 U/ml, respectively). We performed a bronchoscopic examination, and her bronchoalveolar lavage fluid (BALF) had a remarkably cloudy appearance (Figure 2). PAP was suspected, and tests for serum anti-granulocyte macrophage colony-stimulating factor (GM-CSF) antibody were negative; therefore, she was diagnosed with secondary PAP (sPAP) and AML.

PAP is a rare respiratory disease in which reduced GM-CSF-dependent surfactant substances stored in the peripheral air cavity of the lung cause respiratory failure.\(^2\) BALF has a white turbid appearance, deposition of acidophilic/non-structural substances that have a granular appearance during optical microscopic examination and foamy macrophages. PAP is an auto-immune disorder, and anti-GM-CSF antibodies are detected in serum samples.

sPAP is a rarer disorder, is not dependent on GM-CSF and mainly occurs owing to a hematological disease.\(^3\) The mechanism underlying sPAP remains unclear, and there are no specific treatments for patients with sPAP; therefore, sPAP has a poor
prognosis. If patients with a hematological disease have a diffuse pulmonary shadow on CT, such as a crazy-paving appearance, sPAP should be considered.

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References

Figure 2. Her bronchoalveolar lavage fluid (BALF) had a remarkably cloudy appearance.