Remembering Other Causes of Alveolar Siderophages

Macrophage Activation Syndrome

To the Editor:

We read the interesting article of Dr. Ksienski and colleagues1 that talks about cocaine, pulmonary edema, and siderophages. These cells are hemosiderin-laden alveolar macrophages characteristically found in BAL fluid or lung biopsy from patients with diffuse alveolar hemorrhage. Classically, diffuse alveolar hemorrhages are classified based on histologic appearance as capillaritis (Wegener granulomatosis, systemic lupus erythematosus, and propylthiouracil related), bland hemorrhage (cogulopathies, congestive heart failure, and rapamycin related), diffuse alveolar damage (ARDS and Crack cocaine inhalation), and miscellaneous conditions (lymphangioleiomyomatosis and pulmonary capillary hemangiomatosis).

However, we consider that macrophage activation syndrome should be included in the differential diagnosis of alveolar siderophages.2–5 Macrophage activation syndrome is a rare disease characterized by an immune dysregulation with excessive hemophagocytosis in lung/bone marrow/spleen (producing hemosiderin-laden macrophages), splenomegaly, bicytopenia, hypertyglyceridemia, hyperferritinemia > 500 µg/L, low natural killer-cell activity, increased plasma serum CD25, and fever.2 In addition, macrophage activation syndrome usually occurs in response to a triggering agent, such as tumors or infections (especially Epstein-Barr virus). Furthermore, macrophage activation syndrome may present with the triad of diffuse alveolar hemorrhage (lung infiltrates, a falling hemoglobin level, and worsening dyspnea).3–5 Therefore, chest physicians and pathologists should remember macrophage activation syndrome in patients with pulmonary infiltrates and falling hemoglobin levels and/or alveolar siderophages.

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REFERENCES