European Journal of Internal Medicine 68 (2019) e5–e6

Contents lists available at ScienceDirect

European Journal of Internal Medicine

journal homepage: www.elsevier.com/locate/ejim

Internal Medicine Flashcard

An atypical cause of dyspnea

Charlotte Kaeuffer[a,*], Marie-Pierre Ledoux[a], Raoul Herbrecht[a,b]

[a] Department of Oncology and Hematology, Hôpitaux Universitaires de Strasbourg, Strasbourg, France
[b] Université de Strasbourg, Inserm, UMR-S1113 / IRFAC, Strasbourg, France

ARTICLE INFO

Keywords:
Dyspnea
Crazy paving pattern
Chest CT-scan
Bronchoalveolar lavage

1. Case description

A 53-year old woman was admitted to hospital with a six-month history of cough and dyspnea, weight loss and left abdominal and lumbar pain. Inspiratory crackles were diffusely heard during physical examination. The chest radiography revealed bilateral symmetric opacities. Chest CT-scan showed bilateral crazy paving patterns with diffuse ground-glass opacities, inter and intralobular septal thickening and alveolar infiltrates with peripheral sparing. Bronchoscopy with bronchoalveolar (BAL) returned abundant opaque and milky lavage fluid (Fig. 1). Blood cell count, liver and renal function tests were normal.

The diagnosis of pulmonary disease was suspected by clinical and radiographic findings, and finally confirmed by the appearance of lavage fluid. In parallel, ultrasonography revealed a large heterogeneous mass in the left kidney in the setting of a biological inflammatory syndrome. These were further explored and related to a renal *Aspergillus fumigatus* abscess based on repeated positive urine cultures and a positive culture from a renal biopsy. What is your diagnosis for the pulmonary disease?

2. Diagnosis

The crazy paving pattern on CT-scan and the opalescent milky BAL fluid suggest the diagnosis of pulmonary alveolar proteinosis. Pulmonary alveolar proteinosis is a rare disease with an estimated prevalence of 0.37 per 100,000 individuals [1]. This condition is characterized by accumulation of lipoproteinaceous material in the pulmonary alveoli as a consequence of deficient clearance by alveolar macrophages [1,2]. The increased protein levels reduce the surface area available for gas exchange in the alveoli, thereby impairing lung function. Patients with pulmonary alveolar proteinosis often present with exertional dyspnea, cough, weight loss, and fatigue. Pulmonary alveolar proteinosis occurs in three clinically distinct forms: congenital [due to mutations in the genes encoding surfactant protein or beta-chain of the receptor for granulocyte-macrophage colony-stimulating factor (GM-CSF)], secondary (to hematologic cancer, HIV infection, immunosuppressive therapy or toxic exposure) and acquired [1,2]. The acquired form represents 90% of cases and is related to the presence of neutralizing anti-GM-CSF antibodies. Patients with pulmonary alveolar proteinosis are at increased risk for various opportunistic bacterial and fungal infections because of impaired macrophage function. *Nocardia* sp. and *Aspergillus* sp. infections are the most common of these opportunistic infections [3]. The standard of care for pulmonary alveolar proteinosis is whole-lung lavage to remove excess protein and help reduce respiratory symptoms. Significant improvement of respiratory function has also been reported in patients treated with GM-CSF.

* Corresponding author at: Department of Oncology and Hematology, Hôpitaux Universitaires de Strasbourg, 67098, Strasbourg, France.
E-mail address: charlotte.kaeuffer@chru-strasbourg.fr (C. Kaeuffer).

https://doi.org/10.1016/j.ejim.2019.05.004
Received 25 January 2019; Received in revised form 28 March 2019; Accepted 8 May 2019
Available online 05 June 2019

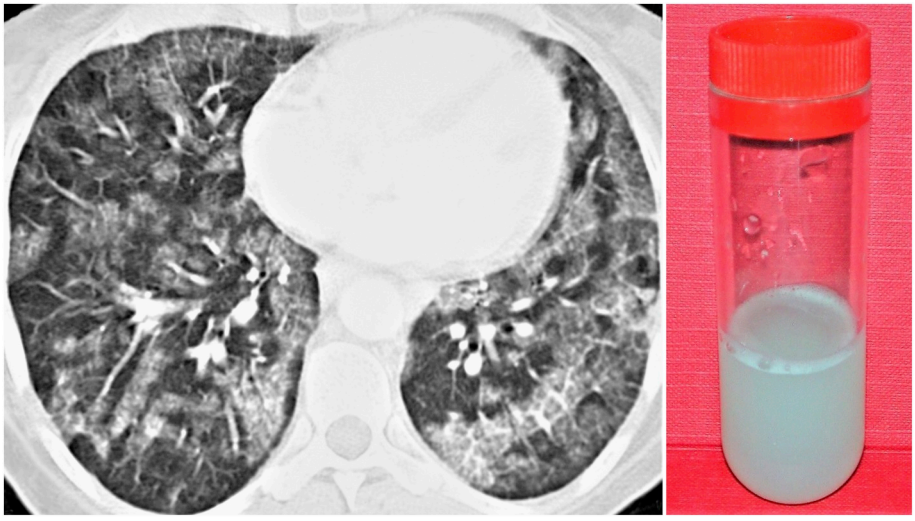

Fig. 1. Left panel: CT-scan showing bilateral diffuse crazy paving patterns; Right panel: Opalescent milky bronchoalveolar lavage fluid.

Declarations of interest

None.

References

[1] Trapnell BC, Whitsett JA, Nakata K. Pulmonary alveolar proteinosis. N Engl J Med 2003;349:2527–39. https://doi.org/10.1056/NEJMra023226.

[2] Ioachimescu OC, Kavuru MS. Pulmonary alveolar proteinosis. Chron Respir Dis 2006;3:149–59. https://doi.org/10.1191/1479972306cd101rs.

[3] Seymour JF, Presneill JJ. Pulmonary alveolar proteinosis: progress in the first 44 years. Am J Respir Crit Care Med 2002;166:215–35. https://doi.org/10.1164/rccm.2109105.