Case report

A 36-year-old male patient was admitted in intensive care unit (ICU) for acute respiratory failure. He was diagnosed five months before referral with T-cell prolymphocytic leukemia, with massive splenomegaly and hepatomegaly, characterized by bloodstream and bone marrow involvement by mature T-cells harboring CD3+/dim, CD8+, CD4-, CD5+ and CD7+ phenotype. He was initially treated with the purine analog pentostatin. He relapsed after two courses, four weeks before respiratory symptoms onset. On admission in ICU, respiratory rate was 36 cycles mn⁻¹, body temperature was 38.1°C, arterial pressure was 116/79 mmHg, heart rate was 128 mn⁻¹. Clinical examination found mild lung crackles, cardiovascular examination remained normal. Transthoracic echocardiography was normal. Chest CT scan showed worsening of bilateral alveolar condensations (Figure 1). PaO2/FiO2 was 155 mmHg under noninvasive mechanical ventilation showed worsening of bilateral alveolar condensations (Figure 1). PaO2/FiO2 was 155 mmHg under noninvasive mechanical ventilation. Flexible bronchoscopy with bronchoalveolar lavage was performed while the patient was under noninvasive ventilation. Cytological examination and immunophenotyping of BAL lymphoid cells confirmed the diagnosis of lung infiltration with prolymphocytic T-cell leukemia. An associated organized pneumonia was suspected. The rapid clinical and radiological response to corticosteroids following by immunotherapy with alemtuzumab strengthened our hypothesis. To our knowledge, this is the first reported case of prolymphocytic T-cell leukemia with a specific pulmonary lung involvement associated with an organized pneumonia.

Keywords: T-cell prolymphocytic leukemia; Respiratory failure; Leukemic lung

Acute Respiratory Failure in a Patient Presenting T-cell Prolymphocytic Leukemia: Specific Leukemic Lung Involvement?

Kim Blanc 1, Aurélie Lefebvre1, Nicolas Chapuis2, Jerome Tamburini2, Bouscary Didier3, Felipe Suarez4, Laurent Frenzel5 and Antoine Rabbat*1

1Service de pneumologie et Soins Intensifs Respiratoires, Hôpital Cochin, France
2Service d’hématologie, Hôpital Cochin, France
3Service d’hématologie, Hôpital Necker, GH Paris centre, AP HP, université René Descartes Paris 5, France

Abstract

A 36-year old patient with relapsing T-cell prolymphocytic leukemia was admitted in intensive care unit for acute respiratory failure and pulmonary infiltrates. A flexible bronchoscopy with bronchoalveolar lavage was performed while the patient was under noninvasive ventilation. Cytological examination and immunophenotyping of BAL lymphoid cells confirmed the diagnosis of lung infiltration with prolymphocytic T-cell leukemia. An associated organized pneumonia was suspected. The rapid clinical and radiological response to corticosteroids following by immunotherapy with alemtuzumab strengthened our hypothesis. To our knowledge, this is the first reported case of prolymphocytic T-cell leukemia with a specific pulmonary lung involvement associated with an organized pneumonia.

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Discussion

Prolymphocytic T-cell leukemia is a rare (less than 2% of lymphocytic leukemia in adults) hematological malignancy arising from mature T-cell lymphocytes but generally demonstrating a very aggressive outcome [1]. Besides blood and bone marrow involvement, spleen, liver and lymph node enlargement are commonly found in this disease (60%, 50% and 50% of cases, respectively) and skin infiltration is detected in up to 20% of patients [2] but pulmonary involvement has not been reported as yet. In our current report, after exclusion of infectious, cardiovascular, toxic and drug etiologies, a specific pulmonary involvement of T cell leukemia was suspected, and immunophenotyping of BAL lymphoid cells highly favored this hypothesis: we demonstrate leukemic lung involvement by the detection of T-cell prolymphocytic leukemia cells in bronchoalveolar fluids [3]. The absence of intraalveolar hemorrhage ruled out the hypothesis of a blood contamination of the bronchoalveolar fluids arguing in favor of a specific lung involvement by leukemic cells. Here, clinical and radiological picture associated with a complete BAL analysis including immunophenotyping on BAL lymphocytes is a useful diagnosis tool for detecting specific lung involvement, allowing early therapeutic intervention.

*Corresponding author: Rabbat A, Service de pneumologie et Soins Intensifs Respiratoires, Hôpitaux Universitaires Paris Centre, Hôpital Cochin, 27 rue du Faubourg Saint Jacques, 75679 PARIS cedex 14, France, Tel: +33158412087; Fax: +33158412088; Email: antoine.rabbat@chc.aphp.fr

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In our current report, we hypothesized that an OP was associated with specific leukemic lung involvement as severe and recurrent episodes of OP have been reported in indolent CD4+/CD8+ T cell leukemia [4]. While definitive proof of this hypothesis could only be given by analysis of lung tissue samples [5], the critical condition of our patient prevented lung biopsy procedures [6].

However, rapid clinical and radiological improvement upon corticosteroid therapy retrospectively strengthened the hypothesis of OP. As noticed previously in OP, alveolar inflammatory lesions are very sensitive to corticosteroid therapy [5]. Indeed, corticosteroid therapy initially given as a monotherapy (i.e. before initiation of alemtuzumab) had no impact on T-cell leukemia tumor burden, supporting the hypothesis that corticosteroids targeted OP inflammatory lesions rather than intra-pulmonary tumor localizations. The diagnosis of OP is difficult as its clinical and morphological features are shared with those resulting from respiratory infections [5]. Estimated incidence is 34/105 cases among patients suffering from haematological malignancies [7]. Among this population, OP is mostly diagnosed in a context of infections, radiation therapy, chemotherapy and stem-cell transplantation. However, OP is also suggested to arise directly from blood cancers prior treatment initiation and in the absence of documented infection [7]. Co-existence of specific lung involvement was not specified in those reported cases [7].

In our current report, we suggest that phenotypic detection of a clonal haematological disease on BAL fluid is a useful diagnosis tool for detecting specific lung involvement in the etiological investigation of acute respiratory failure, allowing rapid therapeutic intervention.

References
