Unusual presentation of an ALK-positive systemic anaplastic large cell lymphoma (ALCL): acute respiratory distress syndrome (ARDS) and fatal evolution.

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INTRODUCTION

ALK-positive ALCL frequently presents at an advanced stage with systemic symptoms and extranodal involvement. However, outcome usually shows a significantly better prognosis than other T-cell lymphomas, with a 5-year OS of 70 to 90%. To our knowledge, acute respiratory distress syndrome (ARDS) due to lymphomatous lung infiltration has never been described in this entity.

CASE REPORT

A 65-year-old female patient presents a progressive dyspnea, associated with fever, a dry cough and a mild diffuse papulonodular rash.

The blood tests show a reactive neutrophilic hyperleukocytosis with CRP at 329 mg/l, LDH just above the normal range and slight signs of cytolysis.

PET scanner reveals some small-sized supra- and infradiaphragmatic hypermetabolic lymphadenopathies (<2 cm) with a low SUV of maximum 9.9 and a massively evolving bilateral pleuropneumonia compared to a scanner performed 7 days earlier.

The clinical course is rapidly unfavorable with development of an ARDS which is treated empirically by intravenous corticosteroids, broad spectrum antibiotics and an antifungal treatment at the intensive care unit.

The first endobronchial biopsy shows subacute inflammation without neoplastic infiltration. Then, a biopsy by EBUS of mediastinal lymphadenopathies and a transbronchial lung biopsy show a tumoral infiltration of these lymphadenopathies and the pulmonary parenchyma by a lymphoid CD30/ALK+ population. Rare neoplastic cells on standard HE coloration are highlighted by immunochemistry after clinico-pathological confrontation. The neoplastic lymphoid population is the same as found in the skin biopsy and the bone marrow biopsy.

Blood smear shows a major neutrophilic reaction with a white blood cell count of >132.000/mm³ overlapping the very low percentage of circulating lymphomatous cells (i.e. less than 1% of the lymphocyte count of 15.360/mm³). Those lymphomatous cells are finally revealed by buffy coat.

A multidrug therapy with brentuximab-cyclophosphamide-adriamycin and prednisolone cannot stabilize the acute situation and the patient rapidly dies.

DISCUSSION

The clinical presentation was unusual, fulminant and fatal, regarding the classical good prognosis of this entity.

ARDS in lymphomas has most frequently an infectious origin. Pulmonary lymphomatous involvement, that is moreover corticoresistant and that evolves in ARDS is extremely rare. So far, it only has been reported for more frequent lymphomas such as diffuse B-cell lymphomas but has never been described in ALK-positive T-cell lymphomas.

Leukemic peripheral blood involvement in ALCL is uncommon and requires a more aggressive therapy, including hematopoietic stem cell transplantation. Only a dozen cases have been reported, mainly children with a very pessimistic prognosis, regardless of ALK-positivity which is known as a factor of good prognosis.

The scant neoplastic cells visible on standard HE coloration contrast with the massive population revealed by immunochemistry. The major intravascular component is an unusual aspect. In front of this unusual histological presentation, CD30 and ALK stains were performed

on first intention on a lung biopsy only due to the clinical information and the lymphoid population seen on the skin biopsy.

CONCLUSION

This case highlights the exceptional and fatal presentation as a non-infectious ARDS of an ALK-positive ALCL with lymphomatous pulmonary involvement which has never been described before.

As diagnosis could only be made by immunochemical complementing, we foreground the importance of the clinico-pathological confrontation, even more when imaging findings and the clinical course are atypical or discordant.