Case report: Richter syndrome with plasmablastic lymphoma

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Richter syndrome is a rare transformation of chronic lymphocytic leukemia into aggressive lymphoma. It occurs in 2% to 10% of cases. Most commonly, Richter syndromes harbor features of diffuse large B-cell lymphoma but cases of Hodgkin lymphomas or prolymphocytic lymphomas have been described. Here, we report the unusual transformation of a 5-point Matutes score CLL into plasmablastic lymphoma.

History of the disease

The patient is a 65 year old man followed for chronic lymphocytic leukemia for 5 years. At the time of diagnosis, he had trisomy 12 and Matutes score of 5. The patient was treated with a Mabthera-Fludarabine-Cyclophosphamide therapy. One year ago, the patient developed a severe thrombopenia treated under Ibrutinib. Later, the patient received successfully allogenic peripherical blood stem cell transplantation with 100% of bone marrow donor cells after 180 days.

Eleven months after transplantation, multiple myeloma was suspected with 34 g/L IgG Kappa. Bone marrow aspiration smear revealed an infiltration by atypical plasmacytoïd cells expressing weak CD45, HLADR-, Kappa+, CD19-, CD20-, CD38+ and CD138+. Results of the biopsy confirmed presence of plasmablastic cells and more recently, atypical plasmacytoïd cells were also detected in pleural fluid. To establish the clonal link between CLL and the plasmablastic lymphoma, restrict pattern analysis comparing a 2 year old bone marrow and the most recent biopsy was made. Clonal CDR1 and clonal CDR kappa were identical on the 2 specimens. The diagnosis of high grade plasmablastic lymphoma was established given the clonally related origin of the disease.