A 63 year-old previously healthy man presented with marked lymphocytosis. Physical examination showed splenomegaly but no adenopathy. The blood count was 38.5 x 10^9 leukocytes with 88% lymphoid cells, 170 g/L hemoglobin and 246 x 10^9/L platelets. The lymphoid cells were large with a single thick-rimmed nucleolus in a round regular nucleus. The chromatin was moderately dense and the cytoplasm was pale blue (Figure 1). Cytological diagnosis was prolymphocytic leukemia (PLL). Immunophenotyping of lymphoid cells by flow cytometry showed a B proliferation CD19+, CD5+, CD23+, CD24, Fm C7–, CD79+ with strong expression of surface IgM k. Cytogenetic study of blood cells was normal without 14:18 translocation and cyclin D1 was not expressed.

The patient was asymptomatic for several months before developing, in a few days, enlarged, tumoral, cervical lymph nodes. Cytological examination of these lymphadenopathies showed voluminous cells: their nuclei had prominent nucleoli and very thin chromatin. The cytoplasm was deep blue (Figure 2). A diagnosis of immunoblastic lymphoma was confirmed by histological examination.

This patient appears to have Richter’s syndrome, that is an aggressive lymphoma which occurred several months after the diagnosis of lymphocytic leukemia. We re-assessed the diagnosis of B-PLL with regard to the cytologic aspect of the lymphoid cells and the strong expression of IgM despite the negativity of FmC7. Richter’s syndrome occurs in about 5% of patients with chronic lymphoid leukemia, but to our knowledge, has not been reported in patients with PLL.