Case reports

Splenic lymphoma with villous lymphocytes : dissociated response with Fludarabin

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The SLVL is a B lymphocytic proliferation, classically treated by splenectomy. We present one patient treated by 3 cycles of Fludarabine with a partial respons.

Observations

We present a 73-year-old patient, hospitalized for asthenia for four weeks, with dyspnea, paleness, splenomegaly extends out from the costal border from 15 cm.

The hemogram shows a leukocyte count at 33.10^9/l of which 85% of lymphocyte, hemoglobin at 5.1 g/dl, (MCV at 85 fl), and platelet count at 177.10^9/l. A bone marrow biopsy (BMB) shows a very rich marrow, invaded by lymphocytes (primarily lymphoplasmacytic), malignant cells have abundant cytoplasm with small surface villous projections.

The cytological aspect and the BMB allow for the diagnosis of SLVL.

The immunophenotype shows B-cell-associated antigens (CD19, CD20, CD22 and IgMs), with positivity for CD5 and CD23. The karyotype again finds a deletion 13 (q13.q33). The biologic assessment shows no element in favor of a hemolysis.

The abdominal echography confirms the splenomegaly and shows multiple retro-peritoneals adenopathies.

Because of the initial refusal of the splenectomy, chemotherapy with fludarabine $25mg/m^2$/d for 5 days every 28 days is initiated, when the transfusional needs are 2 unit of red blood cells every 2 weeks.

After 3 cycles, the hyperleucocytosis regresses to 6.10^9/l of which $0,6.10^9$/l lymphocytes with few circulating villous lymphocytes (CVL), the splenomegaly no longer extends out more than 3 cm. However, the transfusional needs remain unchanged, without any sign of hemolysis (normal blood concentration of lacatate deshydogenase and bilirubin). A splenectomy, therefore, permits the need for transfusion to end. Now one year after the splenectomy, this result continues to hold true.

Discussion

SLVL has a chronic course which, and some patients without any symptoms does not require any therapeutic [3]. There is little data concerning chemotherapy as the primary treatment for SLVL.

Chlorambucil or Cyclophosphamide having a 40 % response rate [1], are inferior to the splenectomy which remains usual treatment. A review of the literature only notes 8 cases treated by fludarabine, 5 of which have received Chlorambucil as first line treatment. The biology normalized and the splenomegaly disappeared in 4 of the cases [2], in 1 case we observe a partial response of 1 year then a chemo-resistant relapse [3], and in 3

cases a major response is indicated without precision [3].

In this case, we noticed a partial response with the regression of the splenomegaly and of the hyperleucocytosis but the transfusional needs were unchanged. Thus, the fludarabine seems to have a role in SLVL but should be reserved for cases where the surgeon is contra-indicated. In the absence of a sign of hemolysis this observation poses the question of the cause of the severe anemia observed, does not seem to be attributable to a splenic sequestration (non-regression of the anemia in spite of the major regression of the splenomegaly), however completely corrected by the splenectomy.

References

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