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Immunophenotypic Diagnosis of Low Clone Hairy Cell Leukemia (A Case Study)

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Abstract: Tricholeucocyte leukemia is a rare type B chronic lymphoproliferative disorder that affects only the adult, mainly male, evoked before the association: splenomegaly, pancytopenia and characteristic decrease of monocytes. The diagnosis is based on the identification of tumor cells in the blood, marrow and / or spleen. Complete blood cytology by immunophenotyping lymphocytes by flow cytometry makes it possible to evoke the diagnosis. We reported a case of hairy cell leukemia with a low B-cell clone, describing the various diagnostic aspects including flow cytometry immunophenotyping.

Key words: Flow Cytometry · Hairy Cell · Immunophenotyping · Low Clone

INTRODUCTION

Hairy cell leukemia (HCL) is a rare hematological malignancy, whose recognition is not always easy and can be confused with other blood diseases; it is therefore essential to obtain an accurate diagnosis to determine the best treatment options. Flow cytometry allows the diagnosis of HCL by the detection of markers characteristic of hairy cell leukemia cells: CD11c, CD25, CD103, CD123 [1].

Case Study: A 41-year-old patient who had diffuse abdominal pain with asthenia, on clinical examination, there is splenomegaly reaching the umbilicus without hepatomegaly or associated peripheral lymphadenopathy.

The complete blood count (CBC) showed white blood cells (WBCs) 2100 / mm^3 with 400 / mm^3 neutrophils and 50 / mm^3 monocytes after correction, the hemoglobin level was 7.6 g / dl; Average RBC volume at 88 μ^3, platelets 50, 000 / mm^3. The tumor lysis balance is normal with serology Ag Hbs, Ac Hvc, HIV, TPHA, VDRL negative.

It is noted that the cytology automaton can provide a result including a monocytosis, by confusing the monocytes with the hairy cells, which must be known to characterize and to highlight the study of the blood smear. The blood smear showed the presence of large cells with an extensive cytoplasm, weakly basophilic with fine cytoplasmic projections. The nucleo-cytoplasmic ratio is low and the rounded nucleus often eccentric. Nuclear chromatin has a finely dispersed appearance and the nucleolus with little or no visible appearance is small (Fig. 1).

Immunophenotyping on peripheral blood by flow cytometry showed a lymphocyte population with a strong expression of CD45.

The immunophenotypic study of the CD45 + lymphocyte subpopulations shows the presence of a CD3 + T population estimated at 87.9%, an estimated CD38 + CD56 + NK population of 4.3% and a 7.8% CD19 + B lymphocyte (Fig. 2).

The study of isotype restriction of light chains shows the absence of kappa / lambda imbalance (Fig. 3).

An expanded panel of monoclonal antibodies was performed targeting CD19 + lymphocytes, demonstrating an immunophenotypic profile made of high intensity CD5 +, CD5 +, CD23-, FMC7 +, CD79b, high intensity IgM, CD10-, CD38-, CD103 +, CD11c +, CD25 +, CD123 +.

The matutes score in our case is equal to 1 and we noted the expression of the markers specific to hairy cells.

The treatment received by our patient consisted of a splenectomy to suppress hypersplenism and the administration of interferon alpha. Five weeks after the start of treatment, the patient died.

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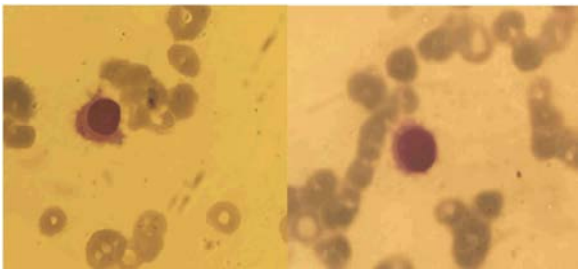

Fig. 1: Cytological appearance of hairy cells (x100)

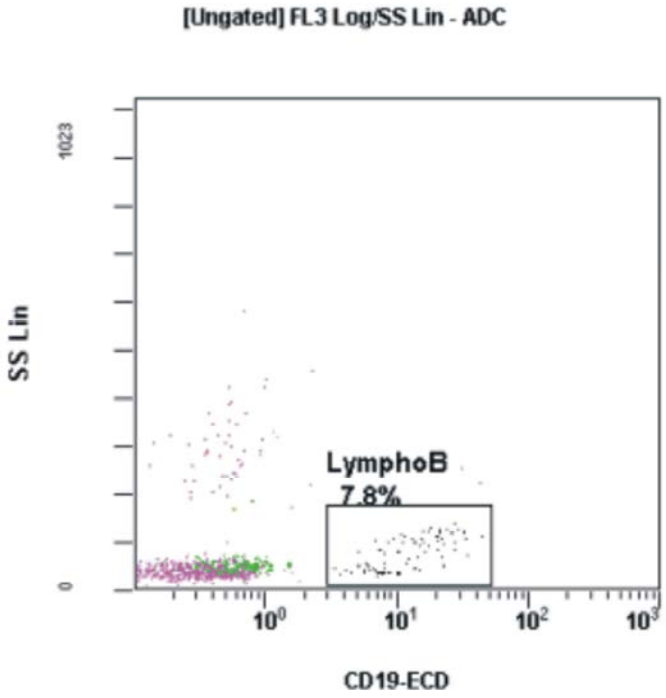

Fig. 2: Bi-parametric representation CD19 / SS

DISCUSSION

Hairy cell leukemia (HCL) is a rare adult disease that accounts for approximately 2% of all leukemias [2]. It is a type of B chronic lymphoproliferative syndrome characterized by an infiltration of the liver, spleen and bone marrow by a particular cell, the hairy beetle [3]. HCL is observed more frequently in humans from the fifth decade [1]. The clinical picture of HCL associates splenomegaly with pancytopenia, sometimes only neutropenia, thrombocytopenia or anemia, which is often discreetly macrocytic [4].

The diagnosis of HCL is based on the detection of hairy cells in the blood, marrow or spleen. In this case, the hemogram showed a cytopenia including a monocytopenia which contrasts with the analysis of the automata and which identify the hairy cells as monocytes. Hairy cells, few in number, are large cells with an extensive, weakly and irregularly basophilic cytoplasm with fine cytoplasmic projections. "Granulo-lamellar" cytoplasmic inclusions with the appearance of discretely basophilic rods with a clear central zone are sometimes detected. The nucleo-cytoplasmic ratio is low and the nucleus often eccentric. Oval or rounded, it can sometimes be kidney-shaped. Nuclear chromatin has a finely dispersed appearance and the nucleolus, little or no visible, is small and often unique (Fig. 1).

Lymphocyte immunophenotyping by flow cytometry on peripheral blood showed the presence of CD19 + CD20 + lymphoid B cells, which have the same FS / SS (Forward scatter / side scatter) characteristics as monocytes, expressing the aberrant marker CD5 usually negative in the cells. HCL described in the literature [5]. CD103, CD11c, CD25, CD123 are also positive with strong

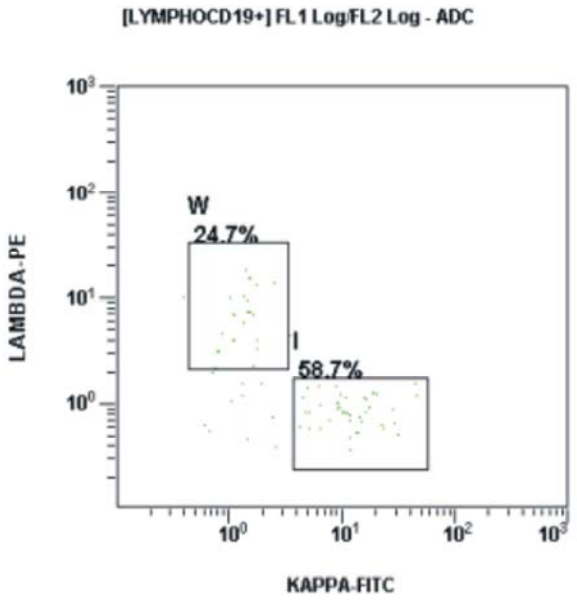

Fig. 3: Kappa / lambda bi-parametric representation

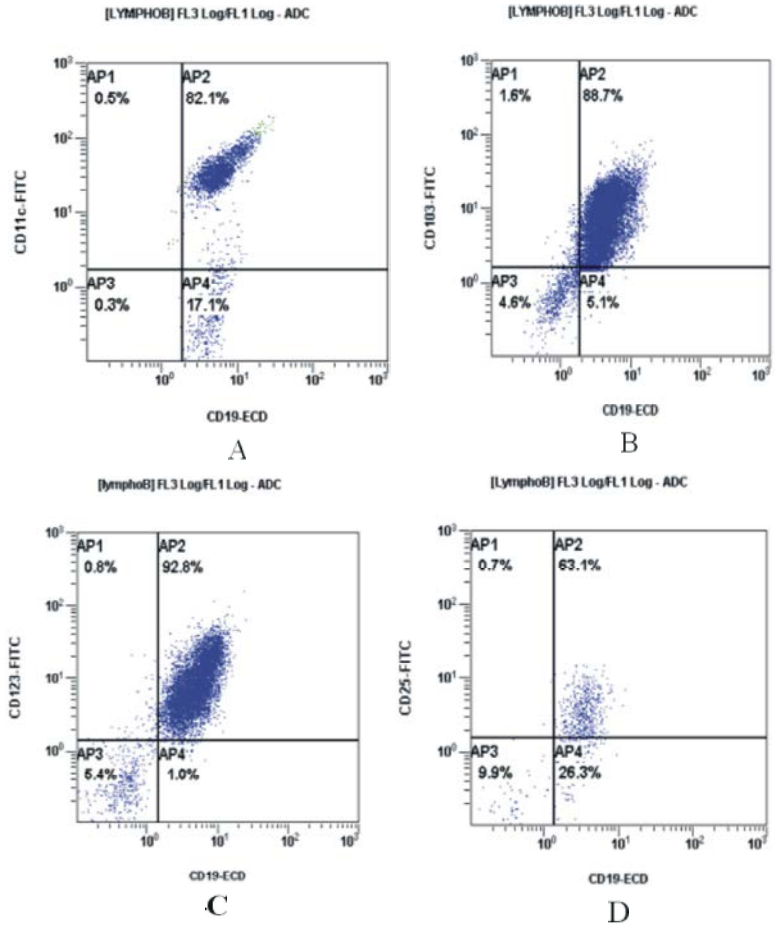

Fig. 4: A) bi-parametric representation CD19 / CD11c; B) bi-parametric representation CD19 / CD103; C) bi-parametric representation CD19 / CD123; D) bi-parametric representation CD19 / CD25

expression of IgM surface immunoglobulins and CD79b and non-expression of CD23, CD10, CD38; Immunophenotypic profile characteristic of most HCLs [6]. The search for an κ / λ isotype restriction is mandatory by labeling with CD19 in search of chronic lymphoproliferative syndrome type B, with the exception of our case because the malignant cells are very rare and do not modify the ratio κ / λ of B lymphocytes. It is then preferable to search for them by their specific phenotype CD19 + CD103 + CD11c + CD25 + CD123 + (Fig. 4).

An immunological score has been developed for the diagnosis of HCL. Based on the expression of four markers (CD103, CD11c, CD25 and CD123), one point is awarded for a positive expression and 0 points for a negative expression [7]. Ninety-eight per cent of HCL cases have a score of 3 or 4, in contrast to variant hairy cell leukemia or splenic marginal zone lymphoma, which is a differential diagnosis problem where the score is usually 0 or 1. [8].

CONCLUSION

The diagnosis of HCL is usually based on clinical-biological characteristics; splenomegaly, cytopenia including monocytopenia, flow cytometry immunophenotyping that allows the diagnosis of classical HCL and other B-type lymphoproliferative disorders that cause differential diagnosis and also the study of residual disease (MRD) which is extremely important: the absence of detection makes it possible to hope for a complete cure of the disease.

Declaration of Interests: The authors declare that they have no conflicts of interest in relation to this article.

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