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Case Report

De Novo Plasma-Cell Leukemia: About A Case

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Abstract: Primary plasma cell leukemia: a rare type of leukemia and plasma cell proliferation Plasma cell leukemia is defined by the presence of more than 20% plasma cells in the peripheral blood or a number of circulating plasma cells greater than 2G/L**.** The primary form: observed may be de novo, and may reveal a multiple myeloma until then unknown. This has similarities with multiple myeloma and has some special particularities that authors will expose in the light of a case diagnosed in the laboratory hematology Hassan II university Hospital of Fez**.**
Keywords: Plasma-cell leukemia, laboratory of medical biological analysis, CHU Hassan II Fez.

INTRODUCTION

Plasma cell leukemia is malignant haemopathy characterized by malignant proliferation of plasma cells in the blood greater than 2 G/L (2 x 109/L). It represents 1 to 2% of plasma-cell neoplasia. The primary form occur de novo in a patient not attending multiple myeloma [1-3]. We report in this work a case of de novo plasma-cell leukemia collected in the hematology laboratory of CHU HASSAN II through which we will discuss the diagnostic and therapeutic peculiarities of this disease.

OBSERVATION

It is a 45-year-old patient with no known pathological history, seen in the emergency department for a serious alteration of the general state, physical asthenia, intermittent bone pain and epistaxis evolving for seven (7) months. The clinical examination found a cachectic patient with a body mass index of 22 kg/m^2, a blood pressure (BP) measured at 90/60 mmHg, dehydrated, no palpable peripheral adenopathy, no hepato-splenomegaly, the conjunctiva and the mucous membranes were pale. A biological assessment was therefore requested: The corrected serum calcium was 5.66 mmol/L, serum creatinine: 480 mmol/L, protein: 73 g/L, proteinuria 24 hours at 1.78 g/24h, LDH at 427 IU/l. The hemogram showed hyperleukocytosis (GB: 24 G/L), normoglycemic normogranic anemia (Hb: 8.1 g/dl) and thrombocytopenia (PLQ: 7000 /IU). The blood smear showed 75% of circulating plasma cells, 18 G / L, made of large, centrally located, and binucleated plasmocytes. (Figure 1 and 2). A sternal puncture was performed, which showed medullary infiltration by more than 85% of plasma cells, most of which are dystrophic (Figure 3). Radiography of the skeleton showed no bone deficiency. Evolution was marked by the patient's death in a severe sepsis table one month later.

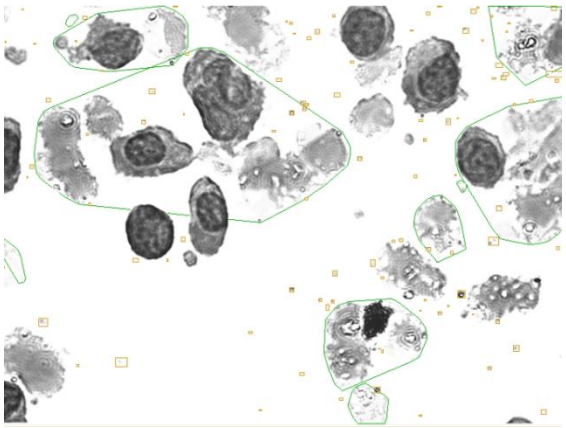

Fig-1: Blood smear of plasma circulating cells

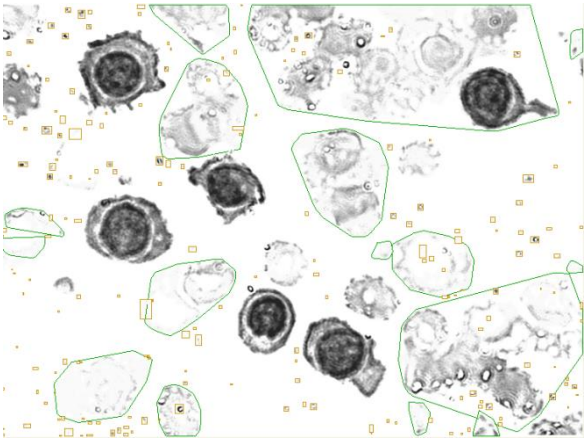

Fig-2: Blood smear of plasma circulating cells

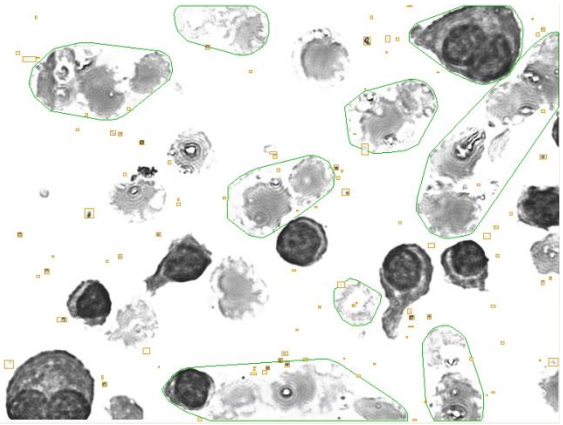

Fig-3: medullary smear of dystrophic plasma cells

DISCUSSION

Plasma cell leukemia is considered a leukemic variant of multiple myeloma. It is primitive in 60% of cases and secondary to multiple myeloma in 40% of cases. Due to the rarity of this disease, only a few cases have been reported in the literature [1]. It is characterized by its aggressiveness and poor prognosis. The longest survival reported being 28 months that of our patient was one month. The diagnosis of de novo plasma cell leukemia is biological, based initially on the blood-smear data and the May Grunwald Giemsa (MGG) blood smear which shows blood plasma levels greater than 2 G/L or Circulating plasma cell counts greater than 20% of the leukocyte formula [4].

Plasma cells are sometimes difficult to identify on blood smears and the use of immunophenotyping in ambiguous forms is essential for diagnosis. Leukocytosis varies from 30 G/L to 87.6 G/L. Normal white blood cell count or leukopenia may be recovered [5, 6]. Our patient had moderate leucocytosis. The two other strains are often associated with normoglycemic normogranic anemia with hemoglobin of less than 10 g/dl in 45 to 87.5% of patients and thrombocytopenia [7]. Our patient had anemia with hemoglobin at 8.1 g/dL and thrombocytopenia at 7000 /ul. The myelogram or osteomedullary biopsy shows diffuse plasma plasmacytic infiltration ranging from 50 to 100%. This plasmocyte population consists of plasma cells with an eccentric nucleus, a strongly basophilic cytoplasm with an archoplasm, and large dystrophic plasma cells with a double nucleus, triple or even multiple nuclei, with a cytoplasm of the vacuolized urea [8].

In our patient, invasion was significant (95%). These plasmocytes secrete a complete immunoglobulin or light chains found in 39% of cases with 50% of cases the lambda isotope. Primary non-secret cell plasmocyte leukemias have been reported [8]. Renal insufficiency is found in 80 to 100% [7]. Hypercalcemia is common. Calcium levels greater than 2.86 mmol/l (115 mg/l) are found in 44% of patients [9]. The serum LDH level is greater than 300 IU / l in 63% of cases and more than 460 IU/L in 48% of cases [10]. The immunophenotyping of peripheral or medullary plasma cells allows identifying some phenotype differences without a characteristic profile being identified. Hyperexpression of surface antigens CD38 and CD138 [7, 10] is frequent CD28 is present in only 38% of cases [7, 10] and CD56 in less than 20% of cases [7, 11]. A few studies have analyzed the expression of the adhesion molecules considered responsible for the anchoring of plasma cells in the stroma of the bone marrow. In our patient, immunophenotyping was not

performed. The genetic abnormalities found [7, 10] are non-specific but some findings are worth mentioning, such as the loss of chromosome material from the long arm of chromosome 13 (80% of cases) and chromosome 16 (80% of cases) the high frequency of translocation t (11; 14) (q13; q32) (33% of cases) and the rarity of hyperdiploidy. The most common molecular abnormalities are the amplification and hyperexpression of the c-myc oncogene (in 2/3 of the cases), the mutations of the p53 gene [12], the hyperexpression of MDM2 (murine double minute 2) [13], hypermethylation of the P16 gene [13] and mutations of the K-Ras and N-Ras [14] genes found in 30-50% of cases.

In addition to molecular anomalies, several molecules are secreted by the medullary stroma and the microenvironment and are involved in plasma proliferation such as lectin, interleukin 3 and 6, vascular endothelial growth factor (VEGF) and insulin growth Factor 1). The prognosis is very poor [7, 15], the median overall survival would be 7 months [1]. Factors of poor prognosis (b2 microglobulin, S-phase plasma cells, renal function, serum calcium levels and serum LDH levels) are often found [10]. Translocation (11,14) is correlated with a better prognosis [1].

CONCLUSION

Primitive plasma cell leukemia is a rare malignant haemopathy. Several points distinguish it from secondary plasma cell leukemia complicating multiple myeloma. It's very dark prognosis justifies the testing of innovative and very promising treatments such as thalidomide analogs and proteasome inhibitors.

ETHICAL APPROVAL

All procedures performed in studies involving human participants were in accordance with the ethical standards of the Institutional and / or National Research Committee and the 1964 Helsinki Declaration and its subsequent amendments or comparable ethical standards.

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