ISSN: 3065-5145
Case Report
World Journal of Medicine and Health Care

Diagnosis of Primary Plasma Cell Leukemia: About One Case

Abddaoui M*, Lemhouer K, Tlemçani L & Amrani Hassani M

Central Laboratory of Medical Analysis, University Hospital Center Hassan II- Fez

***Corresponding author:** Meryeme Abddaoui, Central Laboratory of Medical Analysis, Flow Cytometry Unit. University Hospital Center Hassan II of Fez.

Submitted: 16 June 2025 **Accepted:** 24 June 2025 **Published:** 03 July 2025

https://doi.org/10.63620/MKWJMHC.2025.

Citation: *Abddaoui, M., Lemhouer, K., Tlemçani, L., & Amrani Hassani, M. (2025). Diagnosis of Primary Plasma Cell Leukemia: About One Case. Wor Jour of Medic and Heal Care, 3(4), 01-03.*

Abstract

Plasma cell leukemia is a rare lymphoproliferative disorder characterized by the monoclonal proliferation of plasma cells in the marrow and blood peripheral. It is defined by a blood plasmacytosis greater than 2 G/l or a plasma cell level greater than 20% of leukocytes. It can be primitive or secondary to multiple myeloma. In this context, we report 1 case of primary plasma cell leukemia, through this observation we describe clinical, biological, prognostic and diagnostic characteristics of this pathology.

Keywords: Diagnosis, Multiple Myeloma, Plasma-Cell Leukemia.

Introduction

Plasma cell leukemia (PCL) is a malignant proliferation of plasma cells represents about 1 to 3% of acute leukemias. It may be primary in 60% of cases and it manifests itself immediately as a leukemia or as a secondary event, in 40% of cases, complicating a previously diagnosed multiple myeloma [1]. Given the rarity of this condition, only a few cases have been reported in the literature. It is characterized by its aggressiveness and poor prognosis.

Observation

A 53 -year-old patient with no reported pathological history was hospitalized in the internal medicine department for serious alteration of the general state, physical asthenia, intermittent bone pain and epistaxis evolving for five (5) months. The clinical examination found a cachectic patient with a body mass index of 19 kg/m2, a blood pressure measured at 90/60 mmHg, dehydrated, no palpable peripheral adenopathy, no hepato-splenomegaly, the conjunctiva and the mucous membranes were pale. Biologically, we noted: anemia at 8,2 g/dl a regenerative normochromic normocytic, hyperleukocytosis 9100/mm3, thrombocytopenia at 64 G/l, CRP at 123 mg/l, hypercalcemia at 120 mg/l, renal failure with serum creatinine at 35 mg/l, urea at 0.85 g/l and 24-hour proteinuria at 0.98 g/24 h. B2 microglobulin at 10.34 mg/l. The blood smear shows circulating plasma cells at 76%, The medullogram shows the presence of 79% of dysmorphic plasma cells (Figure 1). The immunophenotyping by flow cytomery objectifies a monoclonal population kappa, CD38+, CD138-, CD19- cells, low expression of CD45 and aberrant expression of CD56 (figure 2), Evolution was marked by the patient's death in a severe sepsis table two month later.

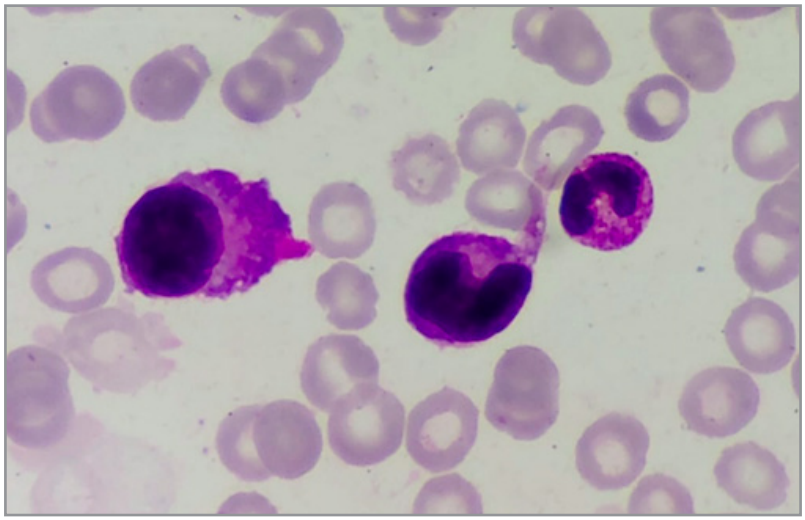

Figure 1: Plasma Cell Leukemia: Marrow Smear

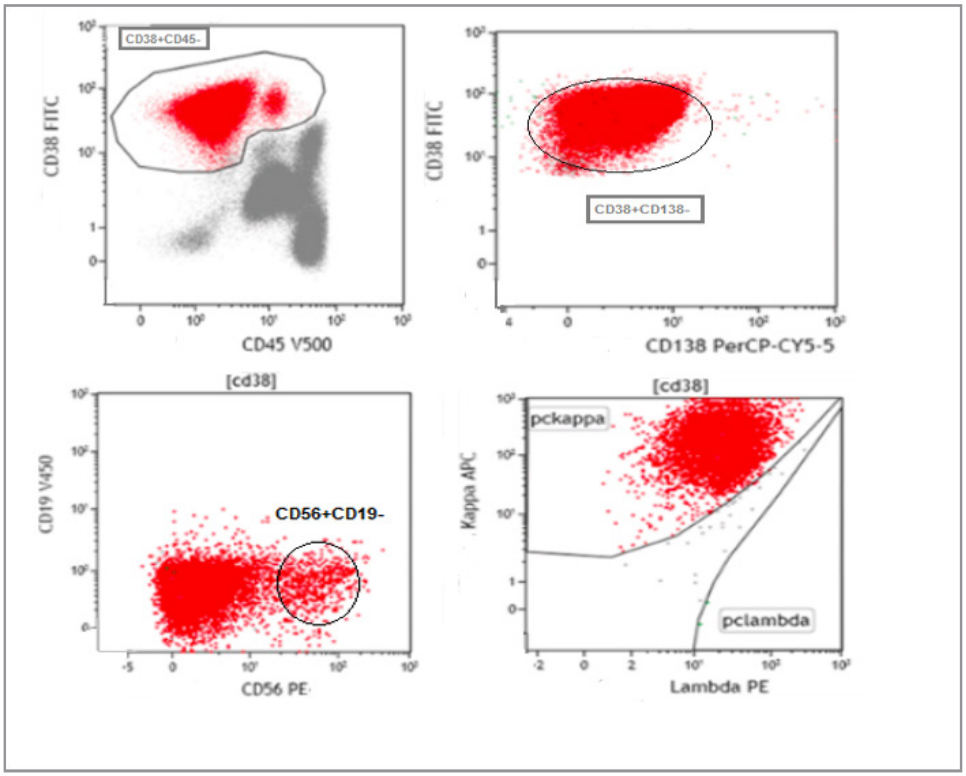

Figure 2: Immunophenotypic Profile of Primary Plasma Cell Leukemia (CD38+CD56+CD19-CD138-CD45- Kappa+)

Discussion

Primitive plasma cell leukemia (pPL) is a rare form of leukemia which occurs immediately at diagnosis unlike the secondary Plasma cell leukemia (sPL) which corresponds to the unfavorable course (in 2% to 4% of cases) of advanced multiple myeloma (MM) [1, 2]. It is characterized by its aggressiveness and poor prognosis.

The majority of pPL affects men with an M/F ratio of around 3/2. The median age at diagnosis of pPL ranges from 49.5 to 65 years depending on the series, in our case it concerned a man aged 53 years old. The longest survival reported being 28 months that of our patient was two months [3].

Leukocytosis ranges from 30 G/L to 87.6 G/L. Normal white blood cell counts or leukopenia may be present [4, 5]. Normochromic normocytic anemia with a hemoglobin level of less than 10 g/dl and thrombocytopenia are often present in 45 to 87.5% of cases. Our patient had a moderate leukocytosis, anemia with hemoglobin at 8.2g/dL and thrombocytopenia at 6400 /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 [6].

In our patient, invasion was significant (95%). Renal insuffi-

ciency 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 [8].

The diagnosis of de novo plasma cell leukemia is biological, based initially on the blood-smear data which shows Circulating plasma cell counts greater than 20% of the leukocyte formula [9]. Plasma cells are sometimes difficult to identify on blood smears: Plasmacytosis in the blood of PL differs from a plasmacytosis reactive to an infectious event or immunological by its monoclonal character. in this context the use of immunophenotyping in ambiguous forms is essential for diagnosis. Flow cytometry has its place to demonstrate the clonality of plasma cells and exclude other syndromes lymphoproliferative, especially lymphoplasmacytic lymphoma. The phenotypic profile of our case study shows the presence of a monoclonal population with low CD45 expression and CD38+ CD56+CD138- CD19-. The CD138 is a membrane marker of normal and dysmorphic plasma cells and dysmorphic plasma cells and absent on the surface of immature plasma cells. CD56 is a marker of TNK lymphocyte population, its expression is inconstant in plasma cell dyscrasia, the CD56 antigen [neural cell adhesion molecule] is important for adhesion of plasma cells to the medullary stroma. We observe the loss expression of this antigen on primitive PL cells and secondary, which may explain their migration into the circulation peripheral. The cytogenetic abnormalities described in the pPL are quite heterogeneous, based on small retrospective studies. Over 80% have hypoploidism or diploidism, which is a factor in poor prognosis. For our patients, the genetic study was not realized.

Age, acute evolution of symptoms, clinico-biological aspect, immunophenotypic profile and absence of myeloma history exclude the possibility of secondary myeloma leukemia and suggest a primary leukemia.

In terms of therapy, no chemotherapy has been instituted because of the aggressive form of leukemia which had led to the death of our patient. The treatment of plasma cell leukemia is poorly codified.

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.

Declaration of Interest

The authors declare that they have no conflict of interest in relation with this article.

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