

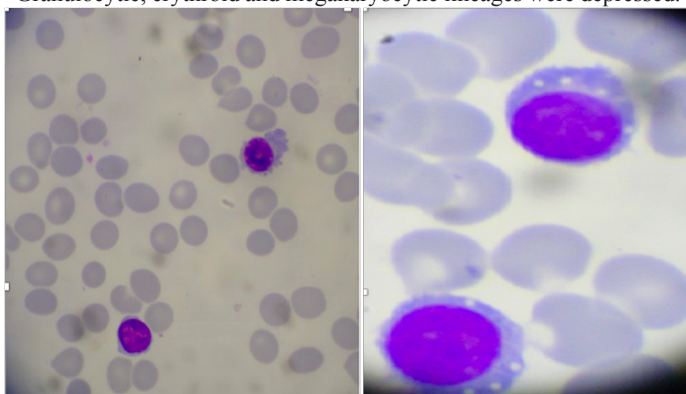
# Plasma cell leukemia with different expression of CD56 marker in the peripheral blood and bone marrow: A case report

## Introduction

Plasma cell leukemia (PCL) is a rare and aggressive entity of malignant plasma cell proliferation that corresponds to 2–4% of plasma cell myeloma (PCM) cases.<sup>1,2</sup> According to the World Health Organization (WHO), PCL is defined as a disorder in which clonal plasma cells constitute more than 20% of total leukocytes in the blood or their absolute count is more than  $2.0 \times 10^9/L$ .<sup>1,3</sup> The bone marrow is usually extensively and diffusely infiltrated. Neoplastic plasma cells are frequently present in extra-medullary sites such as the liver, spleen, body cavity effusions, and spinal fluid. PCL can be classified into: primary (de novo, 60–70% of all PCLs) or secondary which is a leukemic transformation occurring in approximately 1% of previously diagnosed PCMs.<sup>4</sup> Patients with PCL have aggressive disease, poor response to therapy, and a relatively short survival.<sup>1</sup>

## Case report

A 62-years-old female patient presented to the Hematology department at Alexandria main university hospital suffering from bone pain, fever, and dysuria. Past history and family history were irrelevant. On examination, chest and heart were free and there was no organomegaly (lymphadenopathy, hepatomegaly or splenomegaly). On laboratory evaluation, the complete blood picture revealed a hemoglobin level of 8.3 g/dL, hematocrit of 25.2%, white blood cell count of  $19.51 \times 10^9/L$  and platelet count of  $114 \times 10^9/L$ . Morphological examination of peripheral blood smear showed small to medium-sized atypical lymphoplasmacytoid cells (accounting for 85% of the differential WBCs count) with eccentric nuclei and basophilic cytoplasm (Figure 1A). Bone marrow aspirate examination revealed that 47% of total nucleated marrow cells were atypical lymphoplasmacytoid cells with a morphology similar to those in the peripheral blood (Figure 1B), and 13% were lymphocytes. Granulocytic, erythroid and megakaryocytic lineages were depressed.



**Figure 1** (A) PB smear showing small to medium-sized lymphoplasmacytoid cells with eccentric nucleus and basophilic cytoplasm. (B) BM aspirate showing atypical lymphoplasmacytoid cells with basophilic cytoplasm.

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Other laboratory tests showed total serum proteins of 6.2 g/dL, albumin of 3.4 g/dL and a monoclonal band of IgG, (lambda) by immunofixation (1.7g/dL). Total serum Calcium was normal (7.9 mg/dL), and renal functions were abnormal with increased blood urea (86 mg/dL) and serum creatinine (2 mg/dL).

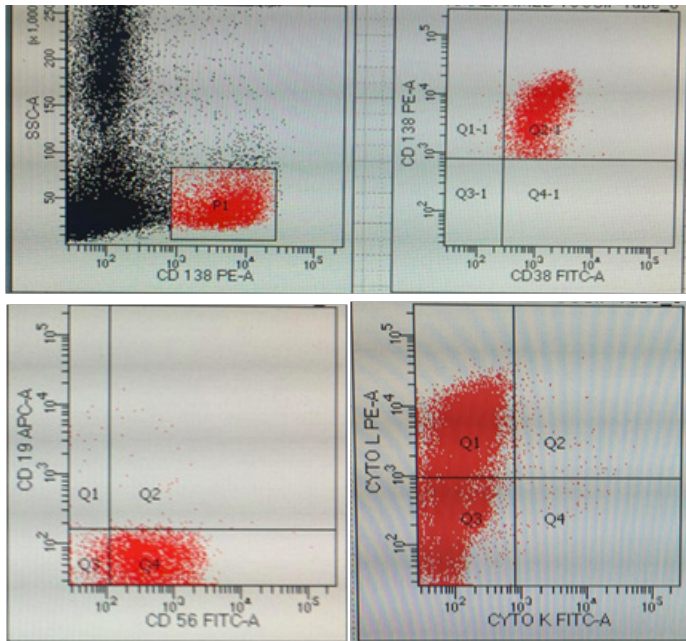
Flowcytometric immunophenotyping of the bone marrow revealed the presence of clonal plasma cells expressing CD138, CD38, CD56 and Cytoplasmic lambda light chain and negative for CD19 and Cytoplasmic kappa light chain, reflecting a plasma cell malignancy despite the morphologic features of lymphoplasmacytic lymphoma (Figure 2). Additionally, immunophenotyping of the peripheral blood was also performed and revealed the presence of clonal plasma cells with the same immunophenotype as the previously identified plasma cells from the bone marrow except that they were CD56 negative (Figure 3). Based on the previous findings, the final diagnosis of the patient was released as plasma cell leukemia.

## Discussion

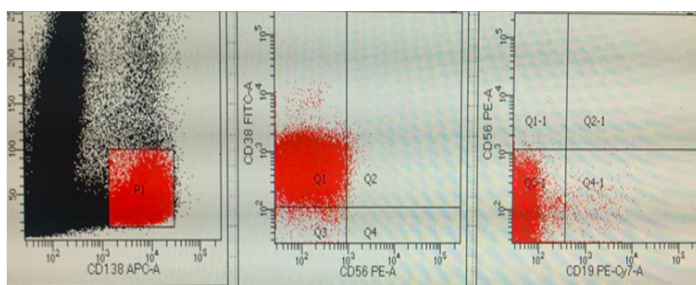
PCL is a rare disease with median patient age younger than myeloma at diagnosis. The bone marrow is usually extensively infiltrated which leads to bone marrow suppression and high incidence of anemia and thrombocytopenia.<sup>2,5,6</sup> Neoplastic plasma cells are frequently found in extramedullary sites such as the liver, spleen, body cavity effusions, and spinal fluid.<sup>1</sup> This appears to be the result of a lower expression of cell adhesion molecules, such as CD56, facilitating the release of leukemic plasma cells from the BM microenvironment.<sup>5,7</sup>

Our patient had no extramedullary involvement at diagnosis, but she presented with anemia, bone pains and renal affection. Her PB and BM aspirate showed extensive infiltration by lymphoplasmacytoid cells that had similar morphology, which made the differential diagnosis between lymphoplasmacytic lymphoma (LPL) and PCL difficult by morphological analysis alone. Normal plasma cells are CD19<sup>+</sup>/CD45<sup>+</sup>/CD38<sup>bright</sup>/CD56<sup>-</sup> while neoplastic plasma cells are usually CD19<sup>-</sup>/CD45<sup>-</sup>/CD38<sup>+</sup>/CD56<sup>+</sup>.<sup>7,8</sup> However, in this case plasma

cells in BM are CD56 positive, while in PB are CD56 negative. CD56 mediates cell-cell and cell-matrix interactions. So, loss of CD56 expression could accelerate the process of metastatic spread.<sup>9,10</sup> The regulation of CD56 is controlled by a very precise mechanism. CD56 can be downregulated during migratory events and re-expressed when the cell reaches a specific site or organ.<sup>7</sup> This finding is also in accordance with study by Pellat-Deceunynck et al,<sup>7</sup> who found migrated plasma cells in the peritoneal cavity of multiple myeloma patients. Cells localized in the peritoneal cavity expressed CD56 and have high proliferation activity.<sup>7</sup>



**Figure 2** Flow cytometric immunophenotyping of the BM showing lambda restricted clonal plasma cells that are CD56 positive.



**Figure 3** Flow cytometric immunophenotyping of the PB showing lambda restricted clonal plasma cells that are CD56 negative.

## Acknowledgments

None.

## Conflicts of interest

The author declares no conflict of interest.

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