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PRIMARY PLASMA CELL LEUKEMIA: REPORT OF TWO CASES

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ABSTRACT

Plasma cell leukemia is a rare malignant hematologic disorder characterized by the presence of more than 20% plasma cells in the leukocyte differential count or a circulating plasma cell count exceeding 2×10^9/L (2G/L). It can be either primary, accounting for approximately 60% of cases, where it presents directly in a leukemic form, or secondary in 40% of cases, arising as a complication of preexisting multiple myeloma. Due to its rarity, only a few cases have been reported in the literature. This condition is marked by its aggressive nature and poor prognosis. Through two cases diagnosed at the hematology laboratory of the Cheikh Khalifa Ibn Zaid International University Hospital in Casablanca, the authors highlight the clinical, biological, and prognostic features of this disease.

KEYWORDS: Plasma cell leukemia, plasmacytosis, multiple myeloma.

INTRODUCTION

Plasma cell leukemia (PCL) is a rare and aggressive malignancy characterized by the abnormal proliferation of plasma cells in the bloodstream. It is defined by a circulating plasma cell count exceeding 2 G/L or constituting more than 20% of the leukocyte differential. Representing approximately 1 to 3% of acute leukemias, PCL is classified into two forms: primary PCL (pPCL), which arises de novo in patients without a prior diagnosis of multiple myeloma (MM), and secondary PCL (sPCL), which develops from the leukemic transformation of preexisting MM. Although PCL shares several features with MM, it also exhibits distinct clinical, biological, and prognostic characteristics.

PATIENT AND OBSERVATION

Case Report 1

A 69-year-old patient with no prior medical history presented with symptoms that began five months earlier, characterized by inflammatory back pain without fever, along with general health deterioration.

Upon admission, the clinical examination revealed pale conjunctiva. Morphological imaging identified a vertebral compression fracture at D8. Preoperative assessment showed renal insufficiency with a blood urea level of 0.64 g/L and a creatinine level of 22 mg/L.

The complete blood count (CBC) revealed:

- White blood cells: 9.3 G/L
- Hemoglobin: 8.6 g/dL
- Mean corpuscular volume (MCV): 102 fL
- Mean corpuscular hemoglobin (MCH): 34 pg
- Platelets: 150 G/L

A peripheral blood smear examination showed 26% plasma cells. Bone marrow aspiration revealed an 88% infiltration by dystrophic plasma cells.

Biochemical analysis showed a total protein level of 64 g/L and hypergammaglobulinemia at 18 g/L, with multiple monoclonal bands detected on serum protein electrophoresis (SPE). Immunofixation (IF) confirmed the presence of a monoclonal immunoglobulin of the IgG Kappa type. The calcium-phosphate balance was within normal limits. Standard skull X-rays revealed multiple punched-out osteolytic lesions.

Case Report 2

A 60-year-old patient with a history of chronic smoking was admitted for inflammatory-type back pain accompanied by a persistent cough, fever, and general health deterioration.

On clinical examination, the patient was febrile, with no signs of purpura or hepatosplenomegaly, and lymph nodes were not palpable. Standard spinal X-rays revealed a vertebral compression fracture at L1, while chest X-rays suggested an infectious pneumonia.

The complete blood count (CBC) showed

- Leukocytosis: 19.6 G/L
- Normochromic, normocytic anemia: 11.8 g/dL
- No thrombocytopenia

A peripheral blood smear examination revealed 30% plasma cells. Bone marrow aspiration showed a hypercellular marrow infiltrated by 45% dystrophic plasma cells.

Serum protein electrophoresis (SPE) indicated hypogammaglobulinemia. Renal and calcium-phosphate parameters were within normal limits.

DISCUSSION

PCL is a rare disease characterized by a malignant clonal proliferation of plasma cells. Compared to multiple myeloma (MM), PCL tends to occur at a younger age, with a median diagnosis age of 55 years, whereas MM is typically diagnosed at a median age of 65 years.[1] The most common presenting symptoms are related to bone marrow failure. The clinical presentation is more aggressive than that of multiple myeloma (MM), with a higher frequency of extramedullary involvement, reported in 23% to 100% of cases depending on the series.[2] The most significant sites of involvement include the liver and spleen, found in 52% and 40% of primary PCL cases, respectively. The diagnosis of PCL is based on biological findings, primarily relying on the complete blood count (CBC) and May-Grünwald Giemsa (MGG)-stained peripheral blood smear (Figure 1). It is characterized by a blood plasmacytosis exceeding 2 G/L or a circulating plasma cell count greater than 20% of the leukocyte differential. Plasma cells can sometimes be challenging to identify on blood smears, making immunophenotyping essential for confirming the diagnosis in ambiguous cases. The diagnostic workup is supplemented by a bone marrow aspiration (Figure 2) or a bone marrow biopsy, along with serum protein electrophoresis with immunofixation, 24-hour urine protein electrophoresis, and a biochemical panel (Figure 3 and 4). Compared to multiple myeloma (MM), PCL is more frequently associated with anemia, thrombocytopenia, hypercalcemia, renal failure, and elevated serum levels of lactate dehydrogenase (LDH) and β2-microglobulin, both reflecting tumor burden. Additionally, cytogenetic analysis of the clonal plasma cell population using fluorescence in situ hybridization (FISH) is essential for further characterization. The presence of a (4;14) translocation involving the gene encoding immunoglobulin heavy chains or a deletion of chromosome 13 is considered a poor prognostic factor.[3] The treatment approach for PCL is determined by factors such as the patient's age, clinical condition, disease extent, and biological parameters.

Melphalan-prednisone chemotherapy appears to be less effective, with a response rate of 20–30% and a median overall survival of approximately 4 to 8 months. In contrast, multi-agent chemotherapy regimens, such as the VAD protocol (vincristine, doxorubicin, dexamethasone), show improved response rates of 40–60% and a median survival of 10 to 20 months. For younger patients, autologous stem cell transplantation following induction therapy has further enhanced survival outcomes.

Recent preliminary data suggest that novel agents, particularly bortezomib, whether used alone or in combination with other chemotherapy regimens, may significantly improve clinical outcomes in primary plasma cell leukemia (pPCL).[4,5]

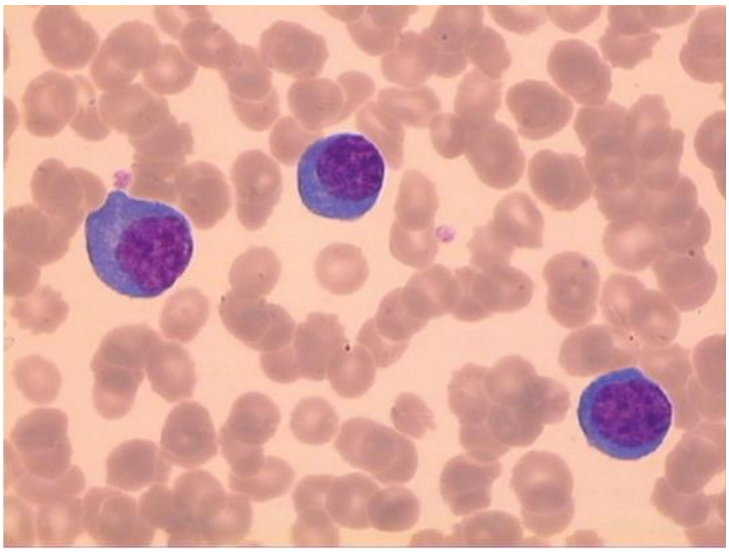

Fig. 1: High-magnification blood smear (100× objective) showing two plasma cells on the left and a lymphoplasmacytoid cell on the right.

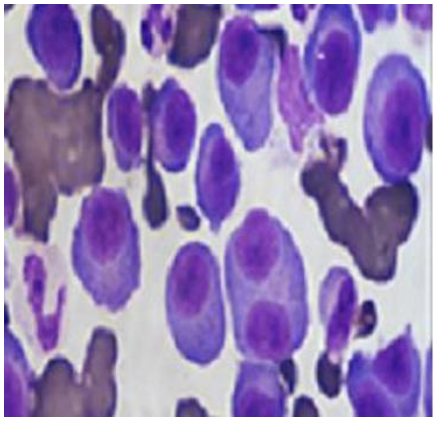

Figure 2: Bone marrow smear stained with MGG (100× objective), showing infiltration by predominantly immature and dystrophic plasma cells.

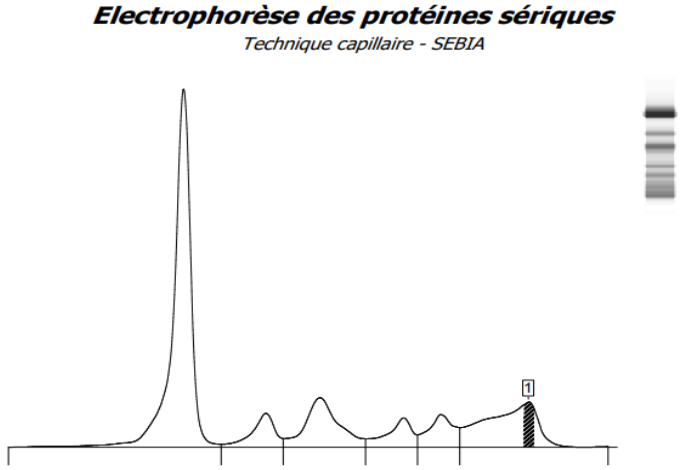

Figure 3: Presence of a narrow monoclonal peak migrating in the gamma globulin region.

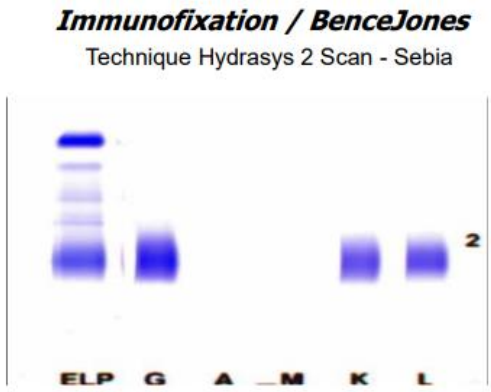

Figure 4: Presence of a monoclonal immunoglobulin of the IgG Kappa type.

CONCLUSION

Plasma cell leukemia is a rare disease with a poor prognosis. While it shares certain characteristics with multiple myeloma (MM), it also exhibits distinct clinical, biological, and prognostic features. PCL requires intensive treatment, now incorporating novel therapies such as proteasome inhibitors and thalidomide analogs. In younger patients, intensified treatment strategies, including autologous or allogeneic stem cell transplantation, have contributed to improved outcomes.

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