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Plasma Cell Leukemia: 2 Cases Reports and Review of Literature

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Abstract | **Cases Report**

Plasma cell leukemia (PCL) is a rare disorder which develops spontaneously (primary PCL) or evolves in patients with multiple myeloma (secondary PCL). It is defined by the presence of 2 × 109/L peripheral blood plasma cells or plasmacytosis accounting for more than 20 % of the differential white cell count. PCL presents more often extramedullary involvement, anemia, thrombocytopenia, hypercalcemia, as well as impaired renal function. Cytogenetic abnormalities and mutations observed in PCL lead to escape from immune surveillance and independence from the bone marrow microenvironment with changes in expression of adhesion molecules or chemokines receptors. The outcome of PCL has improved with combination approaches with novel agents (including bortezomib and immunomodulatory drugs, such as lenalidomide) and with autologous stem cell transplantation. Allogeneic hematopoietic stem cell transplantation is currently available for young patients. This article is an overview of this rare and severe disease and the different therapeutics options that are recommended through 2 observations.
Keywords: Plasma cell leukemia, diagnosis, cytology, prognosis.

INTRODUCTION

Plasma cell leukemia (PCL) is a rare form of leukemia and an unusual manifestation of multiple myeloma. There are two forms of PCL: the primitive PCL (60-70% of cases) occurring at novo in patients without preexisting multiple myeloma and diagnosed from the outset during the leukemic phase, and the secondary PCL, corresponding to a late evolutionary event in patients with multiple myeloma. Through this work, we report 2 observations.

Case Report 1

67 years old patient, with no notable pathological history, admitted for anemic syndrome, and spinal pain with deterioration of general condition.

Biological assessment revealed an inflammatory syndrome with elevated sedimentation rate and CRP, an aregenerative normocytic normochromic anemia, and 67% plasma cells in the peripheral blood. The myelogram revealed 50% medullary plasmocytosis with dystrophic plasma cells (figure 1). At the protein electrophoresis, we noted presence of a monoclonal peak at the level of gamma globulins and at the IEPP presence of IgG lambda monoclonal gammopathy with positive Bence Jones proteinuria: lambda type.

The ionogram revealed hypercalcemia with impaired renal function. The radiological assessment revealed the presence of lytic lesions in the skull with an old lumbar fracture and a compression of D12. Hypercalcemia was treated by hyperhydration, diuretics and corticosteroid with good evolution. For his leukemia, the patient started chemotherapy, received 2 cures and was lost sight of.

Case Report 2

55 years old patient, known to have multiple myeloma; admitted for bone pain and deterioration of the general condition. The blood count revealed hyperleucocytosis at 20G/L, a normochromic normocytic aregenerative anemia (Hb: 9g/dl), and thrombocytopenia at 110G/L.

The cytological study on the blood smear showed a level of plasma cells at 24%, with the presence of red blood cells in rolls (figure 2).

At myelogram; the bone marrow was 60% invaded by dystrophic plasma cells (flamed cytoplasm, vacuolated cytoplasm, binucleate nucleus, etc.). The sedimentation rate was 87mm/1hour. Other biological parameters revealed hyperproteinemia at 120g/l, LDH at 1894UI/L, acute renal failure with preserved diuresis (creatinemia at 143mg/l), hypercalcemia corrected at 120 mg/l and a slight increase in phosphoremia at

63mg/l. Protein electrophoresis and immunofixation revealed monoclonal gammopathy at 69.2g/l (Ig G lambda). The patient died within 70 hours of admission.

DISCUSSION

Plasma cell leukemia, the most aggressive variant of monoclonal gammopathy, has been described by both Gluzinski and Reichenstein more than a century ago [1]. It is a rare lymphoproliferative disorder characterized by malignant proliferation of plasma cells in the bone marrow and peripheral blood. It is a rare form of multiple myeloma, defined by the presence of more than 20% of plasma cells or circulating plasma cells greater than 2 G/L in circulating blood. There are two variants; the primitive form (60% of cases) is observed at novo in patients who have presented no prior signs of myeloma, while the secondary form (40% of cases) consists of leukemic transformation of multiple myeloma already known, and will be in this case the ultimate evolution of the disease which in general relapsed or refractory (only 1% of multiple myeloma will evolve to a secondary plasma leukemia).

Clinically, the PL has a more aggressive presentation than multiple myeloma, with a very large tumor mass [3]. The main reasons for consultation are anemic or haemorrhagic syndrome and symptoms related to hypercalcemia. Extra-medullary involvement by plasma cell infiltration (hepatomegaly, splenomegaly, lymphadenopathies and tissue plasmocytomas) is more common in primitive PL. In contrast, bone disease is more evolved in the multiple myeloma compared to the PCL.

The medullary microenvironment plays a key role in the pathogenesis of multiple myeloma by triggering the signaling cascades involved in the proliferation, migration and survival of the myeloma cell (MC) [7]. The disruption of these mechanisms could be responsible for the development of the PCL [8]. A number of adhesion molecules are involved in the passage of plasma cells [9]. Thus, the absence of the CD56 antigen, a neuronal cell adhesion molecule, impairs the anchorage of MC to the medullary stroma and thus promotes their passage through the blood. In addition, the deficient expression of this molecule is responsible for a weaker interaction of MC with each other and an increase in the secretion of metalloproteinase-9 (MMP-9) [9, 10]. The downregulation of CD106 and activated CD29 as well as a decrease in the expression of HLA-1 and CD40 surface molecules in the PCL compared with MGUS is also noted [11]. In any case, it is a rare entity with a poor prognosis, especially since there is no known effective therapy.

CONCLUSION

The plasma cell leukemia is a very serious pathology but fortunately extremely rare. The diagnosis is simple based on the discovery of circulating plasma cells on the blood smear by a trained biologist. Many advances have been made in the understanding of the pathophysiological mechanisms and the characterization of the disease, but still a way remains to be done to propose an effective therapy and improve its prognosis.

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