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Plasma Cell Leukemia: An Overview

Jamal I*

Department of Hematology, Indira Gandhi Institute of Medical Sciences, India

***Corresponding author:** Iffat Jamal, Assistant Professor, Department of Hematology, Indira Gandhi Institute of Medical Sciences, Patna, India, Email: iffatjamal111@gmail.com

Editorial

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Abbreviations: PCL: Plasma Cell Leukaemia; IMWG: International Myeloma Working Group; MM: Multiple Myeloma.

Introduction

Plasma cell leukaemia (PCL) is a rare haematological malignancy which is classified into primary (pPCL) and secondary PCL (sPCL). The rising incidence of sPCL is attributed to improved survival in multiple myeloma Kyle's criteria define pPCL as 20% or more plasma cells and at least 2×109 /L plasma cells in the peripheral blood but the International Myeloma Working Group (IMWG) suggests that either one is sufficient for a diagnosis of PCL [1].

pPCL demonstrates an aggressive course and progresses rapidly without therapy. The prognosis is often poor with mortality within the first month as high as 15% [1,2]. Elevated lactate dehydrogenase, anaemia, increased serum beta-2 microglobulin, hypercalcaemia, hypoalbuminaemia and renal impairment are commonly seen in pPCL. Osteolytic lesions are less commonly seen in pPCL as compared with multiple myeloma. Untreated multiple myeloma may lead to sPCL within 20–22 months [2].

Differential Diagnosis

The diagnosis of pPCL is based on bone marrow morphology, flow cytometry, cytogenetic studies, serum and urine protein electrophoresis. The differential diagnoses need to be considered are amyloidosis, multiple myeloma, B-cell chronic lymphocytic leukaemia, hairy cell leukaemia, marginal zone lymphoma and reactive polyclonal plasmacytosis. Amyloidosis is often diagnosed on a positive Congo-red stain biopsy sample taken from the subcutaneous abdominal fat or involved organ system. In this case, the Congo-red stain of the bone marrow trephine biopsy was negative for amyloid deposition. Multiple myeloma without transformation to sPCL would not fulfil the diagnostic criteria of 20% or more clonal plasma cells on the peripheral blood film. Reactive polyclonal plasmacytosis is usually associated with infection or autoimmune disorders. Absence of kappa or lambda light-chain restriction excludes reactive polyclonal plasmacytosis [3].

Treatment

In the transplantation-eligible patient, high-dose therapy with autologous stem cell rescue is currently the most effective therapeutic modality to achieve long-term remission. A 3-drug bortezomib-based induction regimen, such as RVD, PAD, CVD (cyclophosphamide, bortezomib, and dexamethasone) is most commonly practiced. Post-transplantation consolidation and/or maintenance strategies with novel agents have not been extensively studied in pPCL, with only case reports and small case series describing prolonged remissions after maintenance treatment with thalidomide, lenalidomide, and bortezomib. Consolidation regimens that can be used after transplantation include 2-4 cycles of RVD. This treatment can be followed by maintenance therapy with lenalidomide and/or bortezomib until progression [4].

Discussion

The median age of diagnosis for pPCL is 55 years which is a decade younger in comparison with multiple myeloma (MM). Extramedullary involvement is common in pPCL with the IMWG suggesting a baseline 18-fluorodeoxyglucose positron emission tomography computed tomography to be performed in all newly diagnosed PCL. Flow cytometry is crucial in assessing plasma cell clonality. Polyclonal plasmacytosis can be attributed to infection which is commonly seen in pPCL as they have moderate to severe immunoparesis. Plasma cells in pPCL express CD20, CD38 and CD138 with CD56 positivity more frequently seen in MM. The genetic biology in pPCL differs in comparison with MM. Increased incidence of hypodiploidy, 17p deletion, TP53 and DIS3 mutations, t(11;14), t(4;14) and t(14;16) is seen in pPCL.The diagnosis of pPCL is often based on bone

marrow morphology, flow cytometry, cytogenetic studies, serum and urine protein electrophoresis. The differential diagnoses need to be considered are amyloidosis, multiple myeloma, B-cell chronic lymphocytic leukaemia, hairy cell leukaemia, marginal zone lymphoma and reactive polyclonal plasmacytosis. [4,5].

Conclusion

Plasma cell leukemia is a rare entity with grave prognosis. Early detection and proper management can increase the overall survival of the patients.

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