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Guest Editorial

Plasma cell leukemia: An overview

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1. 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.[1,2]

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%.[3]

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.[4,5]

The diagnosis of pPCL is based on peripheral blood smear, bone marrow morphology, flow cytometry, cytogenetic studies, serum and urine protein electrophoresis.[6]

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.[7]

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.

1.1. Treatment

Newer treatment options include cycles of subcutaneous bortezomib oral thalidomide and dexamethasone.

2. Discussion

The median age of diagnosis for pPCL is 55 years which is a decade younger in comparison with multiple myeloma (MM).

The most common subtype is IgG followed by light-chain-only PCL. 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.[5]

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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.[6] 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.[7–9]

3. Conclusion

Plasma cell leukaemia (PCL) is the most aggressive form of all plasma cell neoplasms. Prognosis is often dismal in primary PCL (pPCL) with the median overall survival below 1 year if treatment is delayed. Early diagnosis and effective therapy are vital to improve survival in pPCL.

4. Conflict of Interest

None.

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