

Abstract

Primary plasma cell leukemia (PPCL) is a rare hematological malignancy with an incomprehensible pathogenesis and a very aggressive course. Risk factors that can cause the development of PPCL and affect its course are unknown.

Objective

Evaluating four PPCL patients' clinical and laboratory results was aimed.

Materials & Methods

Four PPCL patients diagnosed in Gülhane Training and Research Hospital between March 2019 and June 2020 were evaluated in terms of laboratory and clinical course retrospectively.

Results

The mean age of the patients at the time of diagnosis was 66 (44-76), and M/F:1/1. None of the patients had a history of Multiple myeloma (MM). Leukocytosis was detected in two patients at the time of diagnosis, two patients presented with low white blood cell count. The mean white blood cell count was $59.625 \times 10^3/\mu\text{L}$ (3800-197.000), and the average plasma cell rate detected in peripheral smear was 54.5% (20-100), and the mean plasma cell number was $55.500 \times 10^3/\mu\text{L}$ (600-188.000). Leukapheresis procedure was applied to one patient twice due to the findings of leukostasis. Anemia was detected in all patients at the time of diagnosis, the mean Hb value was 9.9 g/dL, thrombocytopenia was detected in 3 patients, and the mean platelet count was $101 \times 10^3/\mu\text{L}$. Immunfixation results were; Ig G lambda in two patients, free lambda in one patient, and free kappa monoclonality in 1 patient. The genetic examination results of the

patients could not be reached. One patient was out of follow-up and 3 patients were given Bortezomib-cyclophosphamide-dexamethasone (VCD) in first-line treatment. Two patients died during follow-up. Average survival time was calculated as 77 days for these patients. One patient who was given 1 cycle VCD and 1 cycle lenalidomide-dexametasone (RD) left follow-up and treatment. But 1 year after diagnosis and without treatment, patient presented with leukocytosis and treatment programme started again with VRD. Autologous stem cell transplantation could not be planned due to the short life span of the patients.

Conclusion & Discussion

While the incidence of PPCL is 0.4-1.2/million, progression from MM is seen in 0.5% of the cases. PPCL is defined as the presence of 20% of the leukocyte count from clonal plasma cells or the presence of $2 \times 10^9/\text{L}$ clonal plasma cells in number. There is no difference between MM and

PPHL in terms of CD38 and CD138 positivity, but it has been reported that CD56 is more frequently positive in MM and CD20 in PHL. CD27 positivity in PHL indicates increased antiapoptotic activity. Although approximately 150 multiple myeloma patients were followed up in our clinic between the same dates, secondary PCL cases were not detected, and the fact that all of the diagnosed cases were PPCL created a picture incompatible with the incidence rates reported in the literature. In addition, lambda monoclonality was shown in three of our four patients, and this situation made us think that the question of whether it constitutes a feature for PPCL, unlike myeloma, should be examined. However, more patients are needed for further evaluation and determination of risk factors in this aggressive disease. It is necessary to identify poor prognostic indicators and urgently undergo autologous/allogeneic transplantation in eligible patients using multiple bortezomib-based treatment regimens.