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A Successful Case of Primary Plasma Cell Leukemia Treated with Daratumumab-Based Therapy Followed by Autologous Bone Marrow Transplantation

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INTRODUCTION

Context: Primary plasma cell leukemia is a rare and aggressive variant of plasma cell neoplasm, and its diagnosis is based on the percentage (≥ 20%) of circulating plasma cells in the peripheral blood. It accounts for about 0.5-2% of all plasma cell dyscrasias, and the median age of presentation is 55 years. It is even rarer in young adults; only a few isolated case reports have been reported. In this case, we are representing an aggressive form of plasma cell leukemia that was successfully treated with daratumumab therapy and autologous bone marrow transplantation.

CASE REPORT

In October 2021, a 38-year-old man presented to the emergency department with complaints of extreme sweating, fatigue, urinary difficulties, and lower abdominal pain. Initial investigations revealed anemia (Hb = 8.0 g/dL), significantly elevated urea (26.55 mmol/L), and creatinine (1142 µmol/L). The patient was admitted to the nephrology department and initiated on immediate dialysis. Abdominal ultrasound showed splenomegaly (180 mm).

Due to unexplained anemia and splenomegaly, a hematology consultation was requested. Immunofixation revealed hypogammaglobulinemia with kappa light chains at 4.9 mg/L and normal lambda chains at 26.3 mg/L (lambda/kappa ratio: 5.36). The sedimentation rate was 150 mm/h. No osteolytic lesions were found on X-ray imaging, and serum calcium levels were within the normal range.

Peripheral blood smear demonstrated 22% circulating plasma cells. Bone marrow aspiration and biopsy confirmed extensive infiltration with plasma cells expressing lambda light chains. The cells were CD38+ and CD138+, but CD56–. A diagnosis of primary plasma cell leukemia was established, and the patient was transferred to the hematology unit.

Initial treatment with the VTD-PACE protocol was initiated. However, after two cycles, bone marrow aspiration still showed >90% plasma cells, and the patient remained dialysis-dependent, with a lambda/kappa ratio of 100 (1200/11.9 mg/L), indicating refractory disease.

The patient was referred to an international center for further treatment. He received a combination therapy with Daratumumab, Thalidomide, and Bortezomib. After two cycles, he achieved remission and successfully underwent an autologous bone marrow transplantation. As of July 2022, the patient remains in remission, continues subcutaneous Bortezomib every two weeks, and is no longer on dialysis.

DISCUSSION/CONCLUSION

This patient presented with a clinically aggressive form of primary plasma cell leukemia, exhibiting hallmark features such as severe anemia, acute renal failure, absence of bone lesions, >20% circulating plasma cells in peripheral blood, splenomegaly, and bone marrow infiltration by CD56-negative plasma cells.

The combination of daratumumab-based therapy followed by autologous bone marrow transplantation led to clinical remission and restoration of renal function. This case highlights the importance of early diagnosis and the potential efficacy of novel monoclonal antibody-based regimens in treating refractory primary plasma cell leukemia, especially in younger patients.

REFERENCES

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