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Case Report

Blastic Plasmacytoid Dendritic Cell Neoplasia: A Rare Case Report

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Abstract

Blastic plasmacytoid dendritic cell neoplasia is a rare hematological malignancy whose pathogenesis has not been clarified yet. Skin, bone marrow, and lymph node involvement can be seen and is usually seen in men and older adults. Immunohistochemistry features of skin and bone marrow biopsy are important in diagnosis. Here, we presented a case of blastic plasmacytoid dendritic cell neoplasia.

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Introduction

BPDNC is a rare hematological malignancy that is considered to be caused by plasmacytoid dendritic cells and was included in the acute myeloid leukemia-associated precursor neoplasm subgroup by the World Health Organization (WHO) in 2008.[1] The patients frequently presented with common cutaneous lesions. Blastic cell infiltration can be seen in the peripheral smear. Lymphadenomegaly and pancytopenia can be detected in patients. Typically, CD4, CD56, CD123 expression is observed in the bone marrow and skin biopsy by flow cytometry/immunohistochemistry (IHC). The patients are generally men and older adults.[2] The median survival is 12-14 months from diagnosis. Advanced age and stage are thought to be associated with poor prognosis. Patients often respond to initial chemotherapy, but relapses are frequent. Multiagent chemotherapies such as CHOP, hyper-CVAD are frequently used. Stem cell transplantation can be an option in young and well-performing patients.

Case Report

The patient, 81 years old, known to have no comorbidities, was examined five months ago with

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newly developed swelling in the right arm and widespread rash on the whole body. The patient didn't have B symptom. On physical examination, there were multiple lymphadenomegalies in bilateral cervical, axillary, and inguinal regions, the largest of which was 2.5x2 cm in size. He had hepatomegaly. Splenomegaly wasn't detected. In laboratory tests, leukocyte: 3060/mm^3, neutrophil: 1,270/mm^3, lymphocyte: 1,620/mm^3, hemoglobin: 11.1 g/dL, platelet: 16,800/mm^3, ESR: 25 mm/h, LDH: 170 U/L, beta-2 microglobulin: 3.23 mg/dL. Platelet count in his peripheral smear was consistent with the hemogram. Aspiration and imprints were infiltrated with heterogeneous cells, some with lymphocyte morphology, some with narrow cytoplasm and large-small lymphoblasts without granules at bone marrow biopsy. Blast rate was evaluated as 51%. The immunophenotype was in flow cytometry CD3 (+), CD4 (+), CD5 (+), CD7 (+), CD8 (+), HLA-DR (+), CD20 (-), CD34 (-), and CD103 (-). IHC staining as CD4 (+), CD8 (+), CD38 (+), CD138 (+), CD20 (-), TDT (-), CD5 (-), and LCA (+) bone marrow biopsy interpreted CD4 expression as supporting T-cell neoplasia. The skin biopsy has been reported as punch biopsy showing atypical lymphoid infiltration. It resulted as IHC as CD3 (-), CD4 (+), CD7 (+), CD8 (-), CD20 (-), LCA (+), and MPO (-). Ki-67 index was reported as 50%. BPDCN was considered primarily supported by skin findings, pancytopenia, and flow. On PET-CT, diffuse increased metabolic activity was observed in the neck, right lung, mediastinum, bilateral axillae, subcutaneous soft tissue in the right humerus, right nipple, spleen, and all bone structures entering the imaging field. Based on the available information, the patient was considered as BPDCN. After four cycles of chemotherapy inter evaluation with the CVP protocol, the CVP protocol was planned to continue. The follow-up and treatment of the patient who has been applied cure CVP continues.

Discussion

In summary, BPDCN is a rare disease with poor prognosis.[3] Response to initial chemotherapy is good, but relapse is common. More studies are necessary to have a better understanding of the disease for proper management.

Conflict of Interests

Authors declare that there are none.

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