ポスター1-9 ／ AML : APL, BPDCN

2022/10/14 17:40~18:25 ポスター会場 マリンメッセB館 1F　アリーナ

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Background: Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare and aggressive subtype of myeloid malignancy. The etiology of BPDCN was closely associated with clonal hematopoiesis (CH). Therefore, elderly BPDCN patients frequently have gene mutations associated with CH, such as TET2, ASXL1, ZRSR2, and NRAS which are also noted in myelodysplastic syndromes (MDS). Although various regimens for these patients have been implemented, the standard strategy remains to be elucidated. Bcl-2, an anti-apoptotic protein, has been reported to be highly expressed in BPDCN cells. Venetoclax (VEN) targeting Bcl-2 was approved for the treatment of acute myeloid leukemia (AML) recently. We experienced a case of BPDCN associated with CH highly sensitive to VEN-based therapy.Case report: A 70-year-old man was attending outpatient as MDS. He came to our hospital because blasts appeared in peripheral blood. In addition, skin rash was observed in abdomen and enlarged lymph nodes were palpable in cervical and inguinal legions. The blasts in both bone marrow and skin lesion expressed CD4, CD56, CD123, and Bcl-2, and had TET2 and NRAS mutations. Because clinical features of this case were like AML whose blasts expressed Bcl-2, we selected the combination of VEN and azacytidine as induction therapy and achieved hematological complete remission at the end of first cycle. Conclusion: We experienced a case of elderly BPDCN sensitive to VEN-based therapy. Our result suggested that this combination is a therapeutic option for aged BPDCN patients who are not eligible for intensive chemotherapy.