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ELEVENTH ANNUAL MEETING

BLASTIC PLASMACYTOID DENDRITIC CELL NEOPLASM (BPDCN): INITIAL RESULTS OF A NOVEL, ONGOING BPDCN INTERNATIONAL REGISTRY PROGRAM

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INTRODUCTION

BPDCN is a rare, clinically aggressive hematologic malignancy originating from the precursors of plasmacytoid dendritic cells. In 2008 it was recognized by WHO as a distinct entity.

AIMS

Due to rarity and heterogeneous presentation of disease, at present there is no worldwide consensus on the optimal treatment modalities. Launched on July 1, 2022, the aim of our global registry is to establish a novel, large database of patients with BPDCN in order to generate data-driven diagnostic and treatment recommendations.

METHODS

The registry collects retrospective and prospective data on clinical presentation, diagnostics, treatment regimens, and outcomes of patients diagnosed with BPDCN after January 1, 2010. The data is collected electronically through eCRF and Excel form. The registry is included in ClinicalTrials.gov with following number: NCT05430971.

RESULTS

Through May 15, 2023, 14 centers from 10 countries have joined the registry and another 13 centers are onboarding. Twenty-six retrospective and 2 prospective patients are currently included in the registry; 75% are male. By the time of recruitment, 10 patients were alive from which 5 underwent allogenic stem cell transplant (alloSCT). Median age at diagnosis was 61.5 years (4–97). In 93% of patients, the tumor cells were triple positive with CD4+, CD56+, CD123+ immunophenotype; 50% of patients presented with cutaneous manifestation. Twenty-one percent of patients received the initial treatment with AML-based, 64% with ALL-based, and 7% with lymphoma-based regimens. Twenty-one percent of patients underwent alloSCT, of which one died. While 36% of patients were diagnosed after 2018, only 1 patient received tagraxofusp, a CD123-directed therapy that received FDA approval in 2018. Sixty-nine percent of patients achieved complete response, of which 89% were treated with ALL-based regimens. Fifty-three percent experienced relapse. In all patients the cause of death was disease progression except one.

CONCLUSION

Current analysis of a 28-patient cohort of BPDCN demonstrates a high degree of heterogeneity in presentation and treatments. The outcome of ALL-based treatment trended towards superior as compared to AML-based, but relapse rate remained high. Further global collaboration is needed to collect additional data and to identify the best diagnostic and therapeutic approaches for this unique and challenging disease.

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Blastic Plasmacytoid Dendritic Cell Neoplasm International Regsitry

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