Venetoclax combined with azacitidine for treatment of a blastic plasmacytoid dendritic cell neoplasm: A case report and literature review

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Abstract. Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare and highly aggressive hematological malignancy. The neoplasm is associated with a poor prognosis and frequent diagnostic challenges, mainly due to its cutaneous manifestations that can delay an accurate diagnosis or lead to its misclassification as other hematological disorders. In the absence of standardized protocols, the management of BPDCN relies on crucial judgment based on individual cases. The present study reports the case of a 51-year-old male with BPDCN who presented with a fever and bilateral limb pain, and achieved complete remission after only one cycle of azacitidine-venetoclax (VA) therapy. As the patient required concurrent administration of posaconazole (a potent CYP3A4 inhibitor), the dose of venetoclax was adjusted to a low dose of 200 mg per day, and the patient still achieved successful disease remission. The findings from this case study highlight the potential of VA therapy in the management of BPDCN. The study also emphasizes the diversity of the clinical manifestations of BPDCN and the crucial role of combination therapy in its management.

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Abbreviations: BPDCN, blastic plasmacytoid dendritic cell neoplasm; VA therapy, azacitidine-venetoclax therapy; pDC, plasmacytoid dendritic cell; AML, acute myeloid leukemia; ALL, acute lymphoblastic leukemia; OS, overall survival; CLS, capillary leak syndrome; BCL-2, B-cell lymphoma-2; HMA, hypomethylating agent; CR, complete remission; IL, interleukin; FCM, flow cytometry; CT, computed tomography; MRD, minimal residual disease; PBS, phosphate-buffered saline; CH, clonal hematopoiesis; HSCT, hematopoietic stem cell transplantation; allo-HSCT, allogeneic HSCT; CR1, first complete remission; DFS, disease-free survival; FDA, Food and Drug Administration; hyper-CVAD, hyperfractionated cyclophosphamide, vincristine, doxorubicin, dexamethasone; BM, bone marrow; PB, peripheral blood

Key words: BPCDN, venetoclax, azacitidine, combination therapy

Introduction

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare and highly aggressive hematological malignancy that originates from precursor plasmacytoid dendritic cells (pDCs) (1). BPDCN is characterized by unique clinical and pathological features, most commonly presenting with cutaneous involvement, including nodules, plaques or tumors. Varying degrees of bone marrow (BM) involvement, lymphadenopathy, splenomegaly and/or cytopenias have been reported (2). Being a rare condition, BPDCN is often misdiagnosed as other hematological or dermatological conditions (3), leading to delayed treatment initiation and a poor prognosis (4). Before December 2018, the management of BPDCN primarily relied on intensive chemotherapy with regimens used to treat acute myeloid leukemia (AML) or acute lymphoblastic leukemia (ALL). However, treatment responses were transient, and the overall survival (OS) rate was poor (5). Novel targeted therapies, such as tagraxofusp and SL-401, have been recently introduced to improve treatment outcomes in BPDCN. Tagraxofusp, a CD123-targeting monoclonal antibody, shows promise as frontline therapy; however, its clinical applications are limited due to severe adverse effects, such as capillary leak syndrome (CLS) and hepatotoxicity (6). Given that BPDCN cells frequently overexpress the antiapoptotic protein B-cell lymphoma-2 (BCL-2), BCL-2 inhibitors, including venetoclax, in combination with hypomethylating agents (HMAs) such as azacitidine have emerged as a potential therapeutic strategy, especially for elderly and relapsed/refractory patients (7).

The current study reports a case of BPDCN that presented with distinct clinical manifestations and showed a good treatment response. The aim of the present case study is to highlight the clinical variability of BPDCN, the role of targeted therapy in its management and the need for further research to optimize treatment strategies.

Case report

Case. A 51-year-old man was admitted to Beijing Luhe Hospital Affiliated to Capital Medical University (Beijing, China) in July 2024 due to bilateral shoulder and lower limb pain that had persisted for 1 month, and a fever that had been present for 1 week. The patient initially developed spontaneous pain in both shoulders in June 2024, which resolved with cephalosporin therapy administered at an external hospital for 6 days in the same month. The patient had sudden lower left limb pain progressing to intermittent lumbodorsal and bilateral lower limb pain. Symptomatic improvement was achieved with mecobalamin and vitamin B1 at an external hospital. Right lower limb pain worsened in July 2024, and 2 days later, a high fever (39˚C) with chills necessitated hospital admission after self-administered antipyretics. The patient's past medical history included a chronic hepatitis B carrier status for 30 years and a left upper lobectomy with appendectomy for bronchiectasis in 2001.

A physical examination revealed generalized pallor of the skin and mucosa, a 0.5x0.5-cm purple rash (which had existed for several years without change) on the right medial malleolus, a 10-cm surgical scar on the right thorax, absence of palpable superficial lymphadenopathy, negative sternal tenderness, and no lymphadenopathy or hepatosplenomegaly. A BM aspirate smear demonstrated disrupted trilineage hematopoiesis accompanied by focal stromal fibrosis and scattered hematopoietic cells. BM aspiration showed hypercellularity, with 62.5% of abnormal cells characterized by large size and basophilic vacuolated cytoplasm (Fig. 1A). Some cells also exhibited a tailing phenomenon (Fig. 1B) and were negative for peroxidase staining.

Immunophenotyping using flow cytometry (FCM) demonstrated that the abnormal cells were positive for terminal deoxynucleotidyl transferase, human leukocyte antigen-DR, CD56, CD304, CD123 (dim), CD4 (dim), CD99, CD2, CD71 (dim), and CD38 and negative for CD34, CD303, myeloperoxidase, CD117, CD13, CD33, CD64, CD10, CD11b, CD11c, CD15, CD16, CD14, CD9, CD7, c/mCD3, CD5, CD8, CD19, CD20, CD22, cCD79a, CD94, CD161, CD81, CD30, cytokeratin, CD61, CD41, CD41a, CD42a, CD42b and Ki67 (Fig. 2). Myeloid hematological neoplasm-related gene variation testing identified ASXL2 p.Q612X (38.28%) and ETV6 c.1253+1G>T (47.32%) mutations, with negative leukemia fusion genes and a normal male karyotype [46,XY(20)]. Laboratory examinations demonstrated pancytopenia: Neutrophils, $1.20x10^9$/l (normal range, 2.00-7.50x10^9/l); hemoglobin, 82.00 g/l (normal range, 120.00-160.00 g/l for adult males); and platelets, $44.00x10^9$/l (normal range, 100.00-300.00x10^9/l). Peripheral blood (PB) contained 8% blasts. Biochemical analyses revealed increases in lactate dehydrogenase (518 U/l; normal range, 109-245 U/l), C-reactive protein (64.52 mg/l; normal range, 0-10 mg/l), β2-microglobulin (3.12 mg/l; normal range, 1.00-3.00 mg/l) and alkaline phosphatase (191 U/l; normal range, 40-150 U/l). Coagulation studies indicated elevated D-dimer levels [8.51 mg/l (fibrinogen equivalent units); normal range, 0-0.55 mg/l (fibrinogen equivalent units)], whereas cytokine profiling revealed increased interleukin (IL)-6, IL-8 and IL-10 levels. Findings from positron emission tomography-computed tomography (CT) demonstrated diffuse hypermetabolic BM (SUVmax 4.9) and multiple metabolically active lymphadenopathies. CT of the chest revealed post-left lobectomy changes with bilateral supraclavicular and axillary lymphadenopathy. Based on the aforementioned symptoms and outcomes, the patient was diagnosed with BPDCN.Cycle one of VA therapy (100 mg/m^2 azacitidine on days 1-7 + 200 mg venetoclax daily on days 1-21) was initiated for the patient in July 2024 (day 3 post-admission). Meanwhile, 300 mg posaconazole was administered orally on a daily basis for 2 weeks per cycle. A BM aspirate was performed in August 2024 (day 14 after starting treatment). No BPDCN cells were identified on the BM aspirate smear (Fig. 1C and D). Flow cytometry demonstrated 0% aberrant cells in the BM, and the previously positive BPDCN-associated markers were undetectable (Fig. 2). These findings confirmed CR with minimal residual disease (MRD) negativity [there is currently no international consensus on the criteria for defining MRD negativity in BPDCN. In Beijing Luhe Hospital Affiliated to Capital Medical University, MRD negativity (detected by flow cytometry) is defined as BPDCN-associated aberrant immunophenotypic cells in BM <0.01%].

Three cycles were completed without the occurrence of severe complications. After completing initial treatment at Luhe Hospital Affiliated to Capital Medical University, the patient was subsequently managed in an external hospital, so further follow-up details could not be obtained.

Materials and methods

BM aspirate smears. BM aspirate smears were prepared by placing drops of aspirated marrow onto glass slides and air-drying. Smears were stained with a Wright-Giemsa Composite Stain Kit (cat. no. BA-4017; Changde Bickman Biotechnology Co., Ltd.) according to the manufacturer's instructions: The stain was applied to fully cover each smear and staining was performed at 25˚C for 1-2 min. After staining, excess stain was rinsed off with distilled water, and slides were air-dried again. Cellular morphology was examined by light microscopy (Olympus CX43; Olympus Corporation).

FCM. Sample preparation and staining. Ice-cold acetic acid (190 μl) was mixed with 10 μl BM aspirate, and cells were then counted using a hemocytometer. This step was performed to estimate the total cell number, determine the appropriate antibody panels and total staining volume, and set up control tubes. For surface immunostaining, the required antibodies were added to the specimen, gently mixed and incubated in the dark for 15 min. Subsequently, 3 ml lysing solution was added and gently mixed to lyse erythrocytes, and incubated for another 10 min. An appropriate volume of phosphate-buffered saline (PBS) was added, and the mixture was centrifuged at 362 x g for 5 min. The supernatant was discarded, and the pellet was washed once with PBS. After gentle mixing, the mixture was centrifuged again at 362 x g for 5 min, and the supernatant was discarded. Finally, the pellet was resuspended in 300 μl PBS, gently mixed and subjected to flow cytometric acquisition. For intracellular antibody staining, the specimen was first incubated with surface antibodies in the dark for 15 min. Next, 100 μl of fixative was added, gently mixed and incubated in the dark for 5 min. Next, 3 ml of lysing solution was added, gently mixed and incubated in the dark for 10 min

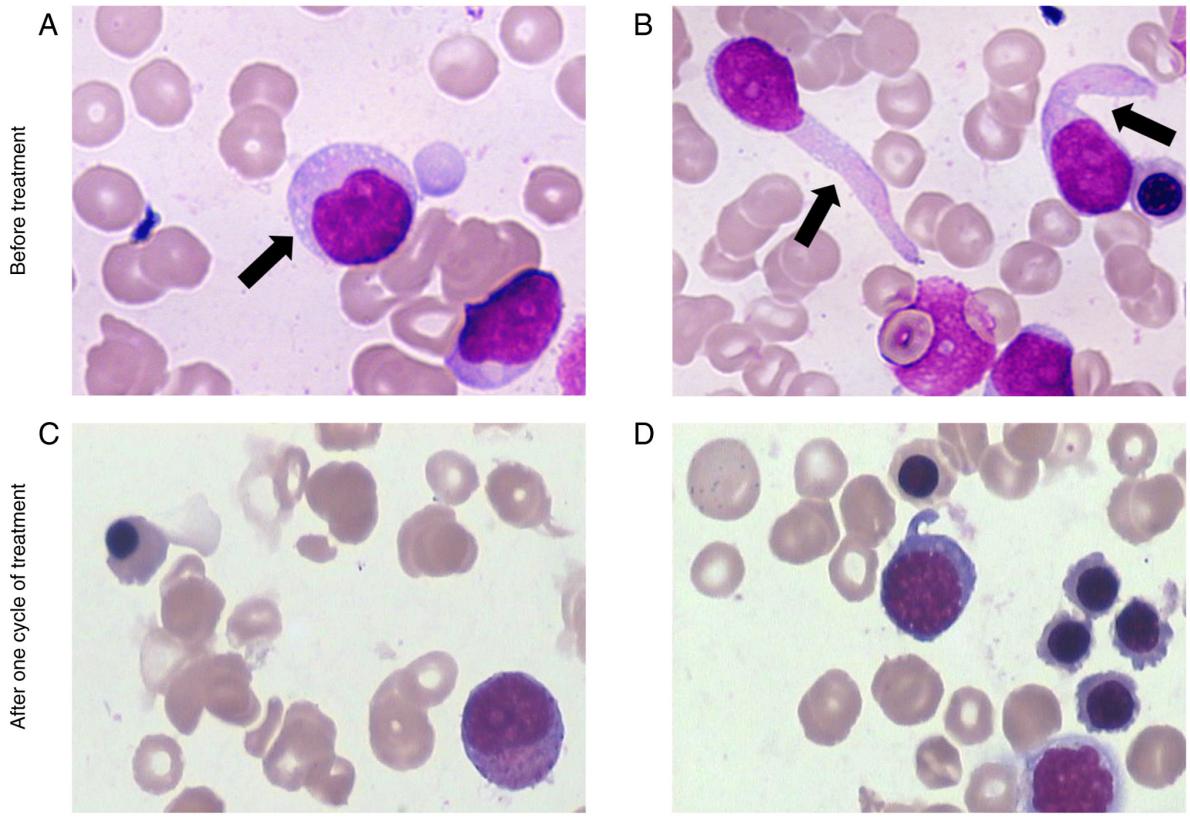

Figure 1. Wright-Giemsa-stained BM aspirate smear images (A and B) before treatment and (C and D) after one cycle of treatment, with a magnification of x1,000). In the pre-treatment BM aspirate smear, the cytoplasm is basophilic, with occasional (A) vacuoles (arrow) and (B) tail-like protrusions (arrow). (C) After one cycle of treatment, mature erythrocytes, promyelocytes, myelocytes and other hematopoietic cells were observed, with no BPDCN cells identified. (D) The remaining fields showed hematopoietic cells consistent with those in (C), and no BPDCN cells were identified. BM, bone marrow.

to lyse residual erythrocytes. The mixture was centrifuged at 362 x g for 5 min, and the supernatant was discarded. A total of 50 μl of permeabilization reagent was added to the pellet, gently mixed and incubated in the dark for 3 min. Intracellular antibodies were then added, gently mixed and incubated in the dark for 15 min. An appropriate volume of PBS was added for washing, and the mixture was centrifuged at 362 x g for 5 min; the supernatant was discarded. Finally, the cells were resuspended in 300 μl PBS, mixed thoroughly and analyzed by flow cytometry. All experimental steps were performed at room temperature (25˚C).

Data analysis. FCM files acquired on the instrument were imported into the analysis software. Cell populations were first gated using a two-parameter dot plot reflecting forward scatter and side scatter to exclude cell debris. A second two-parameter dot plot was then used to discriminate singlets from cell aggregates, thus excluding aggregated cells. Subsequently, a dot plot of CD45 fluorescence intensity vs. side scatter was generated to gate CD45-positive nucleated cells. Within the gated nucleated cell compartment, a series of dot plots were used to evaluate the expression of myeloid and lymphoid markers. These marker expressions were further compared against normal reference profiles to screen for aberrant cell populations. The proportions of each cell population and the intensity of marker expression were calculated and recorded. Dot plots and statistical data were exported, and these results were integrated with morphological review findings to generate the final bone marrow immunophenotyping report.

Reagents. The reagents used included lysing solution for FCM (cat. no. 349202), BD IntraSure Kit RUO (containing fixative and permeabilization reagents; cat. no. 641776) and BD FACSFlow Sheath Fluid (cat. no. 342003) (all BD Biosciences). The fluorescently labeled antibodies used were as follows: CD1a (cat. no. 560945), CD2 (cat. no. A07744), CD3 (cat. no. 555335), CD4 (cat. no. 340133), CD5 (cat. no. 665001), CD8 (cat. no. A07757), CD13 (cat. no. 557454), CD14 (cat. no. 665753), CD19 (cat. no. 652804), CD25 (cat. no. 560503), CD26 (cat. no. 340426), CD33 (cat. no. 664937), CD34 (cat. no. 652837), CD38 (cat. no. A07778), CD45RA (cat. no. 663496), CD45RO (cat. no. 340438), CD56 (cat. no. 347747), CD64 (cat. no. 652830), CD71 (cat. no. 665339), CD94 (cat. no. 559876), CD117 (cat. no. 664936), CD123 (cat. no. 560087), human leukocyte antigen-DR (cat. no. 665745), Myeloperoxidase (cat. no. 665337) (all BD Biosciences), CD7 (cat. no. 007-103-3), CD99 (cat. no. 099-101-3), CD304 (cat. no. 304-102-3), terminal deoxynucleotidyl transferase (cat. no. 815-101-3) (Suzhou Sizhengbai Biotechnology Co., Ltd.) and CD303 (cat. no. 354206; BioLegend, Inc.).

Instrumentation and software. Flow cytometric acquisition was performed using a BD FACSCalibur flow cytometer (cat. no. E97600149; BD Biosciences). Data analysis was performed on a computer equipped with macOS 10.1, using BD CellQuest Pro (for both data acquisition and analysis) and BD FACSComp (for instrument calibration).

Myeloid hematological neoplasm-related gene variation testing. Genomic DNA was extracted from all samples

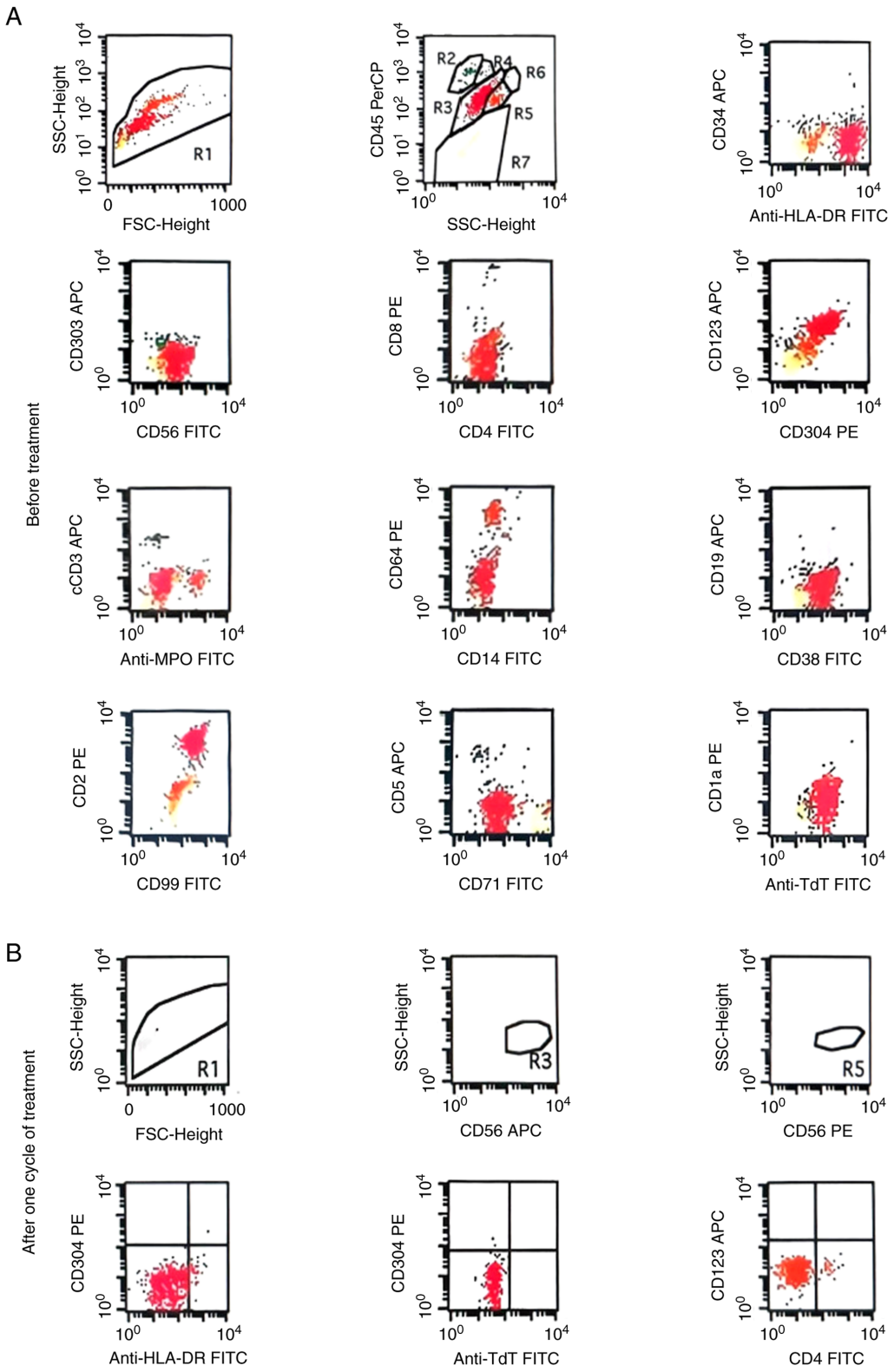

Figure 2. Flow cytometry prior to treatment indicating that the R3 population consists of abnormal cells, accounting for ~75.07% of nucleated cells. (A) This abnormal cell population showed 100% positive expression of CD4 (dim), CD56, CD123 (dim), CD304, TdT, HLA-DR, CD99, CD2, CD71 (dim) and CD38, while being negative for CD303, CD3, CD14, CD19, CD34 and MPO. (B) After one treatment cycle, the proportion of abnormal cells in the bone marrow decreased to 0.00%, and the positive expression rates of CD4, CD56, CD123 and CD304 were all 0%. (This figure only displays the major relevant immunophenotypes and does not present the complete detection results of all immunophenotypes). FSC, forward scatter; SSC, side scatter; TdT, terminal deoxynucleotidyl transferase; HLA-DR, human leukocyte antigen-DR; APC, allophycocyanin; FITC, fluorescein isothiocyanate; PE, phycoerythrin.

and fragmented to ~200 bp by restriction enzyme digestion. Pre-libraries were generated by end repair, adapter ligation and pre-amplification. Target enrichment was performed using Roche targeted capture (Roche Diagnostics GmbH) for a panel of 138 genes implicated in myeloid hematological neoplasms. Libraries were sequenced on an Illumina Inc., instrument

Table I. Clinical manifestation of 68 cases extracted from 57 studies related to blastic plasmacytoid dendritic cell neoplasm (covering case reports published between January 2020 and October 2024) (9-65).

Main lesion region	Percentage of cases (n/total n)
Skin	88.24 (60/68)
BM	83.82 (57/68)
PB	66.18 (45/68)
Lymph node	47.06 (32/68)
Spleen	33.82 (23/68)
Liver	13.24 (9/68)
CNS	10.29 (7/68)
Lung	7.35 (5/68)
Nose	2.94 (2/68)
Breast	2.94 (2/68)

BM, bone marrow; PB, peripheral blood; CNS, central nervous system.

with 2x150 bp paired-end reads, yielding an average on-target coverage of 1,500 times. Sequence variants were detected via Sentieon software; public databases (ClinVar, http://www.ncbi.nlm.nih.gov/clinvar/; COSMIC, https://cancer.sanger.ac.uk) were used to filter germline variants, and the final report included somatic single-nucleotide variants and short insertions/deletions.

Discussion

The diagnosis of BPDCN poses significant challenges and is frequently misdiagnosed as other hematological or dermatological malignancies (8). A systematic review of case reports related to BPDCN published between January 2020 and October 2024, identified 68 patients from 57 reports retrieved through PubMed (https://pubmed.ncbi.nlm.nih.gov/) (9-65). Eligible cases were defined as newly diagnosed patients with BPDCN whose diagnosis was confirmed by flow cytometry in accordance with the 2022 World Health Organization classification of hematopoietic and lymphoid tumors (66). Exclusion criteria included recurrent or refractory BPDCN, duplicate reports and insufficient diagnostic information. Analysis of these cases revealed that BPDCN most commonly involved the skin, BM, PB and lymph nodes (Table I). The highly variable clinical manifestations, ranging from localized cutaneous lesions to systemic involvement, pose substantial difficulties in the early and accurate diagnosis of the disease.

The patient in the current case report initially presented with pain and fever as clinical symptoms, with the lesion site originating in the BM. Laboratory tests revealed 75.07% abnormal cells in the BM and only 8% blast cells in the PB. To the best of our knowledge, no study has directly clarified the mechanism of BPDCN BM-PB tumor-burden difference. We hypothesize two plausible explanations. First, BPDCN originates from BM clonal hematopoiesis (CH), and CH clones can be preserved via autologous stem cell transplantation and undergo cross-lineage evolution (67). This may help explain why BPDCN cells tend to proliferate and persist in the BM. Second, multiple studies have confirmed the involvement of the CXCR4/CXCL12 axis as the core mechanism for the retention of various tumor cells by the BM (68-70). Given that normal pDC precursors (from which BPDCN cells originate) highly express the chemokine receptor CXCR4 (71), we speculate that this may also lead to the retention of BPDCN cells in the BM and their reduced release into the PB.

According to the Fifth Edition of the World Health Organization Classification of Tumors of Hematopoietic and Lymphoid Tissues in 2022 (66), the diagnosis of BPDCN relies primarily on immunophenotypic criteria, including expected positivity for CD123*, TCF4*, TCL1*, CD303, CD304*, CD4 and CD56, and negativity for CD3, CD14, CD19, CD34, lysozyme and myeloperoxidase. The immunophenotypic diagnostic criteria are defined as follows: i) In addition to CD4 and/or CD56 positivity, there is the co-expression of CD123 and at least one additional pDC marker (indicated as * in the aforementioned list); ii) when there is an absence of CD4 and CD56 expression but yet there is positivity for any three pDC markers along with the concurrent negativity for all anticipated negative markers (66). Pathological biopsy remains a cornerstone for its definitive diagnosis (72). Therefore, clinicians should integrate clinical symptoms with the timely selection of appropriate diagnostic modalities such as immunophenotypic analysis, skin biopsy and BM aspiration to optimize the diagnostic workflow for BPDCN.

The role of hematopoietic stem cell transplantation (HSCT) in treating BPDCN has been firmly established. Chemotherapy alone cannot sustain long-term remission in BPDCN; allogeneic HSCT (allo-HSCT) is key for durable remission. Notably, patients with BPDCN undergoing allo-HSCT during first CR (CR1) have a 1-year disease-free survival (DFS) rate of 80%, which is significantly higher than the <50% 1-year DFS rate in those treated with chemotherapy alone (73). A single-center study has reported that among patients with BPDCN who received allo-HSCT in CR1, the 3-year OS rate was >60%; by contrast, the 3-year OS rate was >40% for patients who were not in CR1 (74).

However, a number of patients are ineligible for HSCT due to their poor physical condition. Moreover, they require induction therapy to achieve remission before transplantation. Some targeted drugs are currently under active research, development and clinical applications, boasting broad prospects. These drugs include CD123-targeted agents, such as tagraxofusp, IMGN632 and SL-401 (7). Among them, tagraxofusp is the only drug approved by the US Food and Drug Administration (FDA) for the treatment of BPDCN (75). A clinical study has demonstrated its CR rate of 72% in treatment-naive patients (7). However, its use is contingent on strict patient-selection criteria (for example, patients with reduced ejection fraction, hyperbilirubinemia or hypoalbuminemia are ineligible) and it carries a risk of severe CLS (76).

Chemotherapy regimens tailored to treat myeloid leukemia, lymphoid leukemia or lymphoma are also commonly adopted to treat BPDCN. These regimens are viable options for first-line therapy (77). Studies have shown that the outcomes of ALL-oriented and lymphoma-oriented regimens [for example, hyperfractionated cyclophosphamide + vincristine + doxorubicin + dexamethasone (hyper-CVAD); and cyclophosphamide + doxorubicin + vincristine + prednisone] are better

than those of AML-oriented regimens (for example, ifosfamide + carboplatin + etoposide) (78-80). A retrospective study that recruited 100 patients with BPDCN demonstrated that first-line hyper-CVAD-based therapy achieved a CR rate of 80%, which was significantly higher than that of the CD123-targeted agent SL-401 and other regimens. This intensive chemotherapy regimen is suitable for younger patients with good performance status and is particularly indicated for individuals at high risk of central nervous system involvement (81).

BPDCN cells exhibit the characteristic of high BCL2 expression, and their survival is directly dependent on the BCL2 pathway. This finding provides a clear biological basis for the use of the BCL2 inhibitor venetoclax in treating BPDCN (82). The venetoclax-HMA regimen, which is a combination of venetoclax with HMAs (such as azacitidine and decitabine), has emerged as a highly promising therapeutic strategy for BPDCN. The regimen is particularly suitable for patients who cannot tolerate intensive chemotherapy, those ineligible for HSCT and those with relapsed or refractory disease following prior treatment (83,84). A retrospective study of 10 elderly or frail patients with BPDCN treated with VA showed that 60% of patients achieved CR, and 2 patients were successfully bridged to allo-HSCT (84). Another study reported that although two elderly patients with BPDCN with multiple relapsed and refractory disease (following prior lines of treatment) did not meet the 'formal response criteria' (namely, CR) after VA therapy, 1 patient experienced a 50% reduction in BM blasts, and both patients showed improvement in skin lesions (83).

In the present study, VA therapy was chosen for the patient, who had poor general condition at treatment onset (intermittent fever for 1 week, maximum temperature of 39˚C, lethargy and generalized anemic appearance) and was intolerant of intensive chemotherapy or CD123-targeted agents. Hematological toxicity is a major concern with venetoclax-based regimens (82). Common grade 3/4 adverse events include febrile neutropenia, leukopenia and anemia, whereas infectious complications are primarily pneumonia and sepsis (85). These toxic effects require close monitoring and supportive care to ensure patients' tolerance to treatment. Other therapeutic options include combination regimens involving pralatrexate, enasidenib and bortezomib (76,80).

Notably, the dosage of venetoclax used to treat BPDCN in one previous study ranged from 400-800 mg per day (83). However, in clinical practice, drug interactions are a crucial factor impacting the formulation of treatment regimens. In the present case study, venetoclax dosage adjustment was necessary as the patient's neutropenia required posaconazole intervention (a potent CYP3A4 inhibitor) as antifungal prophylaxis. The FDA recommendation of reducing venetoclax dosage to 70 mg when co-administered with posaconazole is included in the package insert (86). A previous study demonstrated that reducing the venetoclax dosage to 50 mg in patients with AML did not compromise treatment efficacy when used in combination with potent CYP3A4 inhibitors (86). Another study that enrolled 43 patients with relapsed and refractory AML and related myeloid malignancies used venetoclax in combination therapy and found that 3 patients who received higher than the recommended dosages of venetoclax still achieved a clinical response (83). Nevertheless, due to differences in disease pathogenesis, tumor classification and other aspects between BPDCN and AML, these dosage regimens cannot be directly applied to BPDCN treatment. Therefore, based on empirical treatment, 200 mg/day of venetoclax was administered to the current patient. Further large-scale randomized controlled trials are warranted to determine the optimal dosage regimen of VA when used in combination with CYP3A4 inhibitors to treat BPDCN.

Overall, VA therapy demonstrated favorable efficacy in treating BPDCN in the present patient, achieving rapid remission while maintaining good tolerability. However, the lack of long-term follow-up data limits a comprehensive assessment of treatment durability. Nonetheless, this case report provides a clinical reference for selecting therapeutic options for patients with BPDCN who are unable to tolerate intensive chemotherapy or CD123-targeted agents. Moreover, when the patient received the VA therapy in combination with posaconazole, the venetoclax dose was adjusted empirically and was not reduced to the FDA-recommended dose range. Nevertheless, the patient achieved a favorable clinical response and no adverse events were observed. This finding suggests that the dose-reduction thresholds for venetoclax when co-administered with CYP3A4 inhibitors in the treatment of BPDCN still need validation in prospective clinical studies with larger sample sizes. Given the rarity of BPDCN, the ongoing collection of real-world data and additional well-designed clinical trials is imperative to optimize treatment paradigms, refine treatment strategies and improve patient outcomes.

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Availability of data and materials

The data generated in the present study may be requested from the corresponding author.

Authors' contributions

XC and HZ designed the study, advised on patient treatment and analyzed patient data. YL was responsible for collecting clinical, imaging and pathological data of the patient and was responsible for the conception, design and content of the manuscript. XC and HZ confirm the authenticity of all the raw data. YL and XC wrote the original draft. HZ reviewed and edited the original draft. YL revised the manuscript. All authors have read and approved the final version of the manuscript.

Ethics approval and consent to participate

The present study, which includes this case as part of a project evaluating the efficacy of venetoclax combined with azacitidine in treating hematological diseases, was approved by the Ethics

Committee of the Affiliated Beijing Luhe Hospital of Capital Medical University (approval no. 2024-LHKY-087-02) and was conducted in accordance with the guidelines of the Declaration of Helsinki.

Patient consent for publication

Patient written consent was obtained for publication of images and data.

Competing interests

The authors declare that they have no competing interests.

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