

COULD THE HEMATOLOGIC CANCER YOU JUST DIAGNOSED...



ACTUALLY BE BPDCN?

What the CD123 diagnostic
marker can tell you*

HAVE YOU SEEN A CASE OF BPDCN?

Blastic plasmacytoid dendritic cell neoplasm (BPDCN):

- Is an **aggressive** and **deadly** hematologic cancer^{1,2}
- May be **misidentified** because of its **similarity** to other **hematologic malignancies**^{1,3-5}
- Primarily involves **skin**, **bone marrow**, and **peripheral blood**, with variable dermatologic and hematologic presentations^{1,4,6,7}
- Is diagnosed through immunophenotype analysis, with confirming markers including **CD123**, **CD4**, and **CD56**^{1,8*}

BPDCN MAY BE MISTAKEN FOR^{1,3-5}

- AML
- ALL
- Leukemia cutis
- MDS
- Myeloid sarcoma
- CMML
- NK/T-cell lymphoma
- CTCL

BPDCN CAN AFFECT A RANGE OF PATIENT DEMOGRAPHICS¹



~75% are men⁹

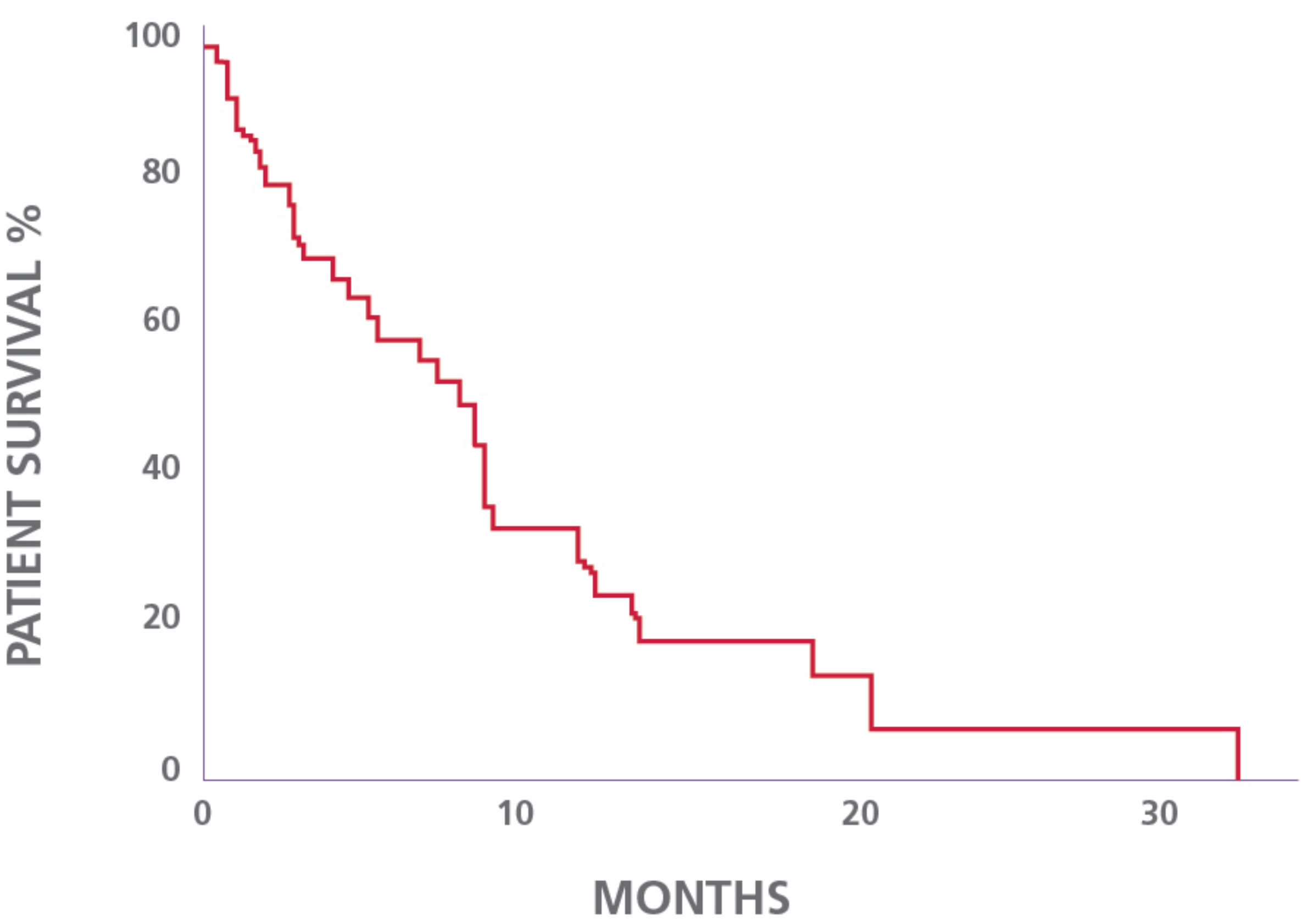


Typically **60 to 70 years of age**, but all ages can be affected¹



Affects **all races** and **geographical locations**¹

HISTORICAL OVERALL SURVIVAL⁹



- In a retrospective analysis, the mean time between the onset of lesions and the final diagnosis of BPDCN was 6.2 months¹⁰
- Historically, median overall survival for BPDCN is approximately 8 to 14 months after diagnosis^{1,2}
- BPDCN may rapidly progress to an aggressive leukemia^{4,9}

Early and accurate diagnosis may be critical in improving patient outcomes¹

AML = acute myeloid leukemia; NK = natural killer; ALL = acute lymphoblastic leukemia; MDS = myelodysplastic syndrome; CMML = chronic myelomonocytic leukemia; CTCL = cutaneous T-cell lymphoma.

*BPDCN diagnosis can include other markers, such as CD4, CD56, TCL1, TCF4, and CD303 (BDCA2).^{1,11}

YOUR AML PATIENT... MAY NOT HAVE AML



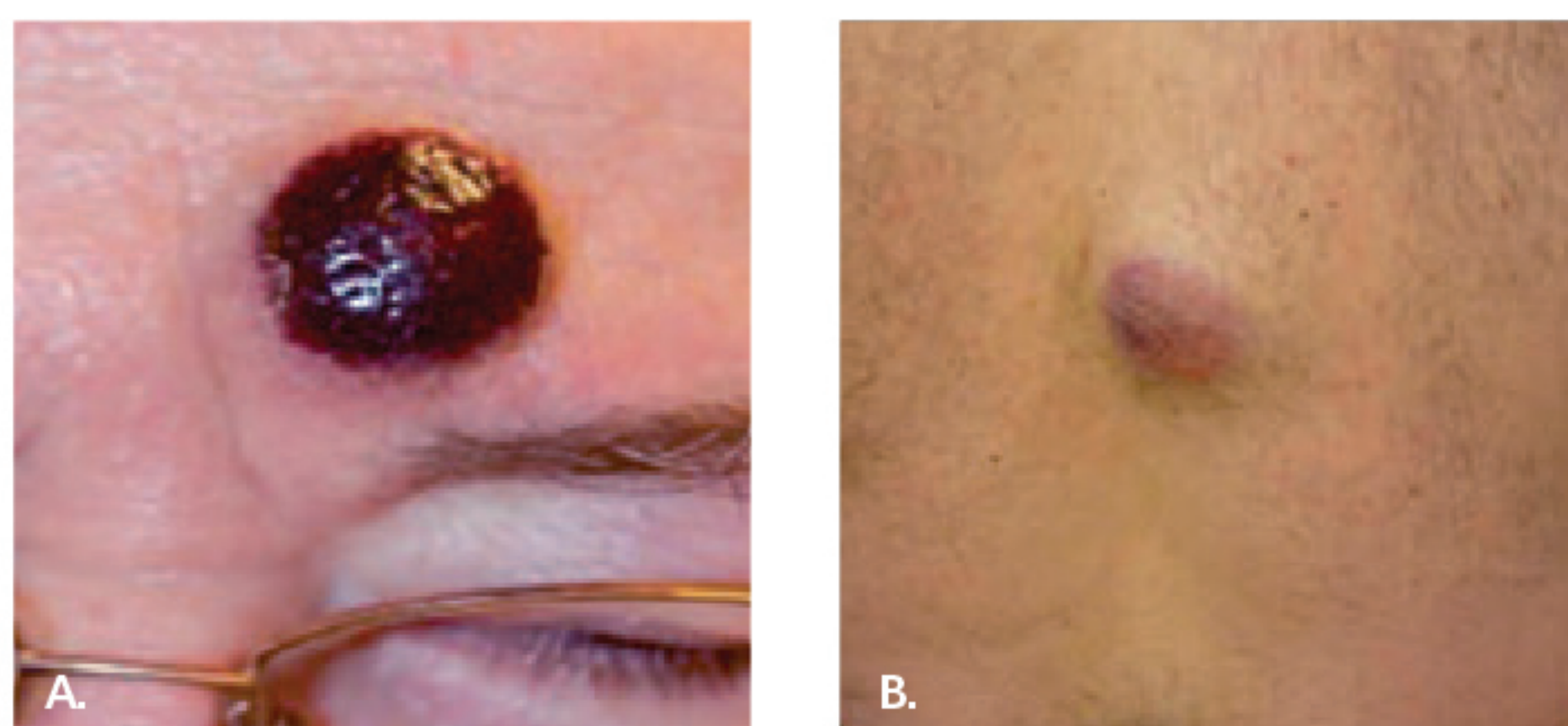
BPDCN PRESENTS AS LEUKEMIC DISEASE IN ~60% TO 90% OF PATIENTS^{4,12}

Patients may present with leukemic signs/symptoms, such as anemia or other cytopenias, due to bone marrow involvement, swollen lymph nodes, or enlarged spleen¹

~85% TO 90% OF PATIENTS WITH BPDCN PRESENT WITH SKIN LESIONS^{3,7,10}

Deep purple nodular lesions^{4,5,10,13}

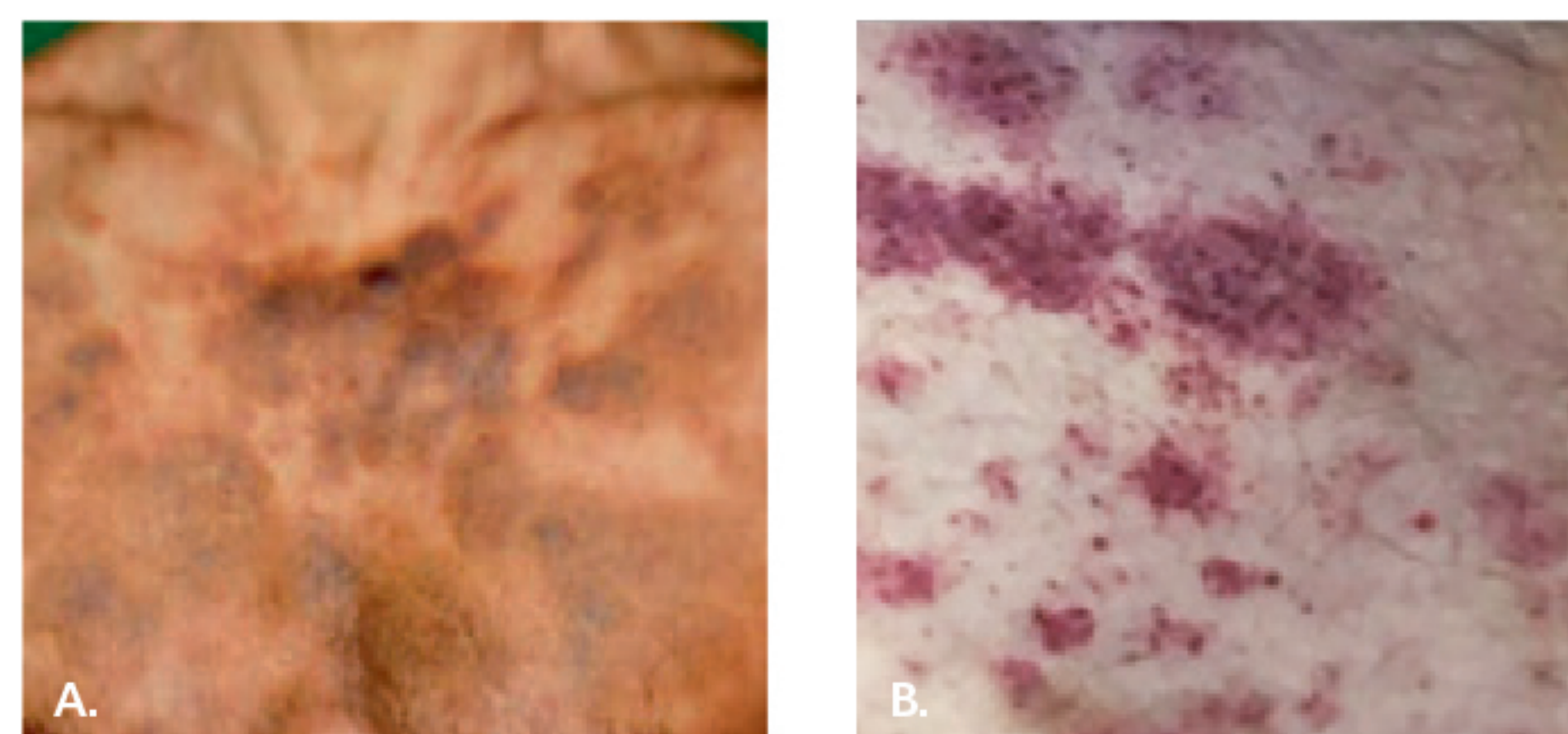
- Most common cutaneous manifestation in BPDCN patients
- Vary in appearance and distribution, localized to various body areas, particularly the trunk, limbs, and head



A. and B. Julia F, et al., Blastic plasmacytoid dendritic cell neoplasm: clinical features in 90 patients, *British Journal of Dermatology*. 2013;169: 579-586, published by John Wiley and Sons. © 2013 The Authors BJD © 2013 British Association of Dermatologists

Diffuse bruise-like or hyperpigmented red-brown macules^{4,6,10}

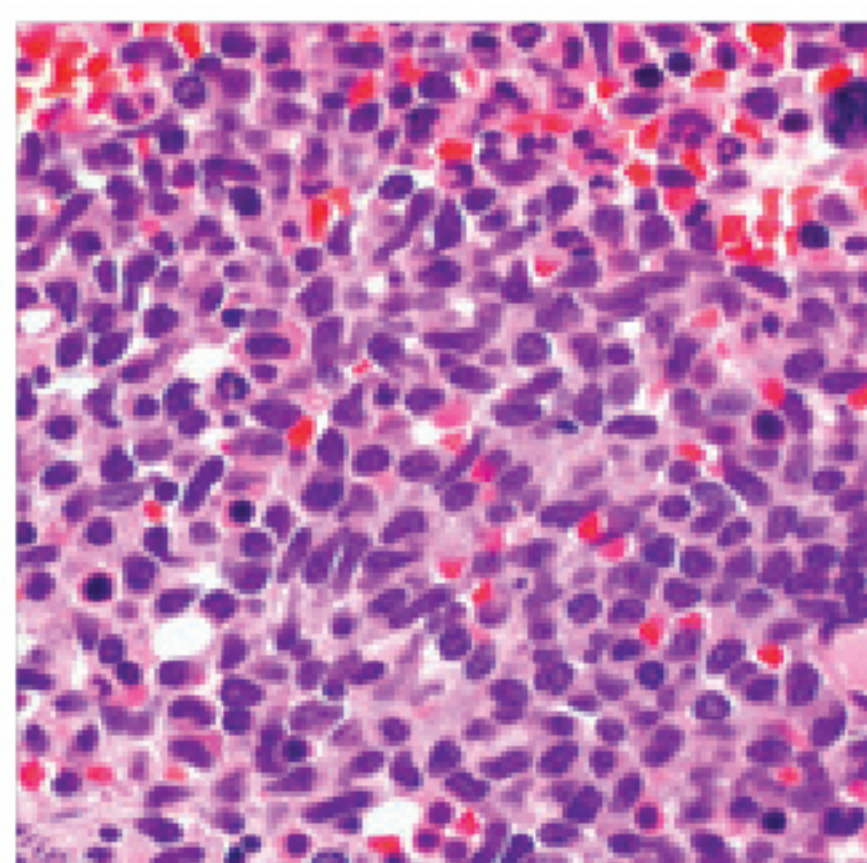
- May be small and/or diffuse, 1 or several patches confined to various body areas, often nonpruritic
- Some patients present with disseminated and mixed lesions



A. Julia F, et al., Blastic plasmacytoid dendritic cell neoplasm: clinical features in 90 patients, *British Journal of Dermatology*. 2013;169: 579-586, published by John Wiley and Sons. © 2013 The Authors BJD © 2013 British Association of Dermatologists B. Republished with permission from *American Society of Hematology*.

BIOPSIES ARE CRITICAL IN IDENTIFYING BPDCN¹⁴

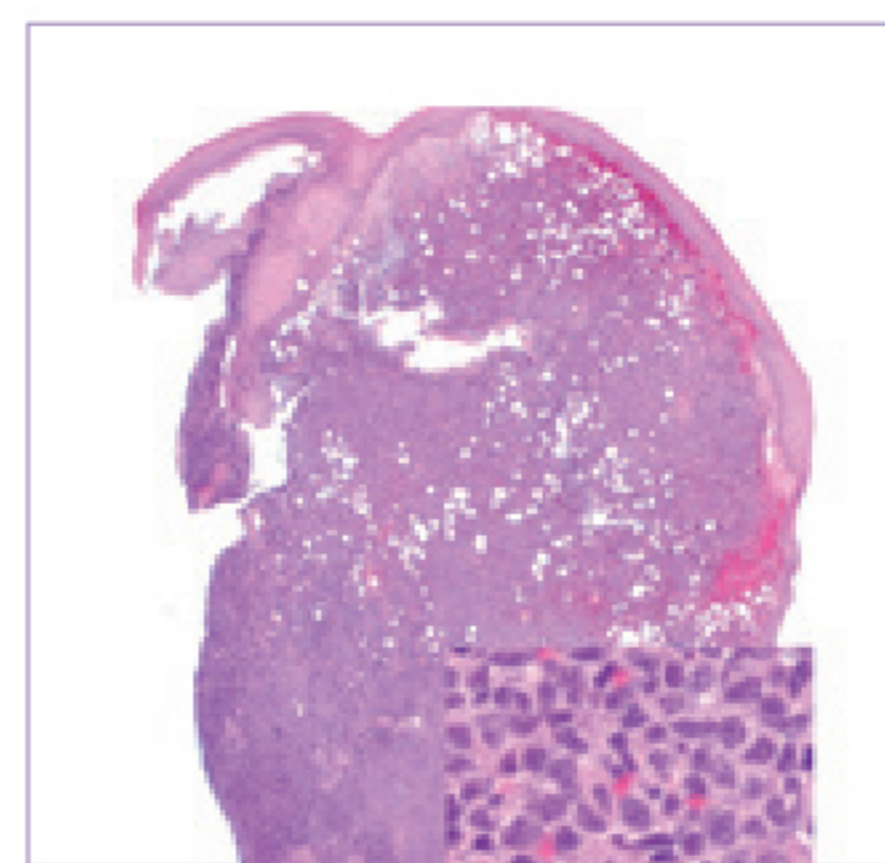
Bone marrow



Core biopsy showing diffuse infiltrate by BPDCN (H&E stain, x600)⁴

Wasif Riaz, Ling Zhang, Pedro Horna, et al, *Cancer Control*, Volume 21, Issue 4 p. 11, copyright © 2014 by SAGE Publications. Reprinted by Permission of SAGE Publications, Inc.

Skin



Punch biopsy of a skin lesion showing BPDCN (H&E stain, x40)

Inset: medium-sized malignant cells spare the epidermis (H&E stain, x1000)⁴

Wasif Riaz, Ling Zhang, Pedro Horna, et al, *Cancer Control*, Volume 21, Issue 4 p. 11, copyright © 2014 by SAGE Publications. Reprinted by Permission of SAGE Publications, Inc.

CD123, CD4, AND CD56 COMPRISE A SIGNATURE MARKER TRIAD THAT IS KEY IN IDENTIFYING BPDCN^{1,8*}

The CD123 marker:

- Is highly expressed on BPDCN cells (~95%) and negligibly expressed on healthy cells^{3,9,15}
- Can be identified through skin biopsy or bone marrow biopsy of malignant cells¹⁴
- Can be both a diagnostic marker and a therapeutic target in BPDCN^{2,3,14}
- Expression of MPO excludes BPDCN⁷

AML = acute myeloid leukemia; MPO = myeloperoxidase.

*BPDCN diagnosis can include other markers, such as TCL1, TCF4, and CD303 (BDCA2).^{1,11}

Include BPDCN in your differential diagnosis for AML and other hematologic malignancies^{1,14}

References: 1. Pagano L, Valentini CG, Grammatico S, Pulsoni A. Blastic plasmacytoid dendritic cell neoplasm: diagnostic criteria and therapeutic approaches. *Br J Haematol*. 2016;174(2):188-202. 2. Pemmaraju N. Novel pathways and potential therapeutic strategies for blastic plasmacytoid dendritic cell neoplasm (BPDCN): CD123 and beyond. *Curr Hematol Malig Rep*. 2017;12(6):510-512. 3. Laribi K, Denizon N, Besançon A, et al. Blastic plasmacytoid dendritic cell neoplasm: from origin of the cell to targeted therapies. *Biol Blood Marrow Transplant*. 2016;22(8):1357-1367. 4. Riaz W, Zhang L, Horna P, Sokol L. Blastic plasmacytoid dendritic cell neoplasm: update on molecular biology, diagnosis, and therapy. *Cancer Control*. 2014;21(4):279-289. 5. Goyal A, Carter JB, Duncan LM. Blastic plasmacytoid dendritic cell neoplasm. In: Carter JB, Goyal A, Duncan LM, eds. *Atlas of Cutaneous Lymphomas: Classification and Differential Diagnosis*. Cham, Switzerland: Springer International; 2015:193-203. 6. Reichard KK. Blastic plasmacytoid dendritic cell neoplasm: how do you distinguish it from acute myeloid leukemia? *Surg Pathol Clin*. 2013;6(4):743-765. 7. Sullivan JM, Rizzieri DA. Treatment of blastic plasmacytoid dendritic cell neoplasm. *Hematology Am Soc Hematol Educ Program*. 2016;2016(1):16-23. 8. Pemmaraju N, Konopleva M. *The Hematologist* website. <http://www.hematology.org/Thehematologist/Ask/8927.aspx>. Published August 28, 2018. Accessed December 11, 2020. 9. Pagano L, Valentini CG, Pulsoni A, et al. Blastic plasmacytoid dendritic cell neoplasm with leukemic presentation: an Italian multicenter study. *Haematologica*. 2013;98(2):239-246. 10. Julia F, Petrella T, Beylot-Barry M, et al. Blastic plasmacytoid dendritic cell neoplasm: clinical features in 90 patients. *Br J Dermatol*. 2013;169(3):579-586. 11. Ceribelli M, Hou ZE, Kelly PN, et al. A druggable TCF4- and BRD4-dependent transcriptional network sustains malignancy in blastic plasmacytoid dendritic cell neoplasm. *Cancer Cell*. 2016;30(5):764-778. 12. Herling M, Jones D. CD4+/CD56+ hematodermic tumor: the features of an evolving entity and its relationship to dendritic cells. *Am J Clin Pathol*. 2007;127(5):687-700. 13. Hirner JP, O'Malley JT, LeBoeuf NR. Blastic plasmacytoid dendritic cell neoplasm: the dermatologist's perspective. *Hematol Oncol Clin North Am*. 2020;34(3):501-509. 14. Facchetti F, Cigognetti M, Fisogni S, Rossi G, Lonardi S, Vermi W. Neoplasms derived from plasmacytoid dendritic cells. *Mod Pathol*. 2016;29(2):98-111. 15. Frankel AE, Woo JH, Ahn C, et al. Activity of SL-401, a targeted therapy directed to interleukin-3 receptor, in blastic plasmacytoid dendritic cell neoplasm patients. *Blood*. 2014;124(3):385-392.



Follow us on twitter @BPDCNinfo

Visit BPDCNinfo.com for more information.

Copyright 2021 - Stemline Therapeutics, Inc.
All rights reserved. 1/2021 US-NON-00663

Stemline