OPEN ACCESS

ISSN: 2835-9496

Akoum R (2026): 1343
Ameri J Clin Med Re, 2026; 6(1): 100250
DOI: 10.71010/2835-9496/ajcmr-e250

Research Article

American Journal of Clinical and Medical Research

Atypical Blastic Dendritic Plasmacytoid Cell Neoplasm: Two Blast Populations and Delayed Skin Manifestations

Riad Akoum*, MD

Medical oncology / Hematology, Lebanese American University Medical Center Rizk Hospital. Beirut, Lebanon

***Corresponding author:** Riad Akoum MD, Email: riad.akoum@laumcrh.com

Citation: Akoum R (2026) Atypical Blastic Dendritic Plasmacytoid Cell Neoplasm: Two Blast Populations and Delayed Skin Manifestations. *Ameri J Clin Med Re*: AJCMR-250. *https://doi.org/10.71010/2835-9496/ajcmr-e250*.

Received Date: 13 April, 2026; **Accepted Date:** 16 April, 2026; **Published Date:** 22 April, 2026

Abstract

Blastic dendritic plasmacytoid cell neoplasm (BDPCN) is a rare hematological malignancy with initial presentation in the skin. Usually the blastic cells are monoclonal CD4-positive/CD56-positive, derived from the plasmacytoid type 2 dendritic cell. These cells are MPO and CD34 negative. We report the case of a 68 year-old patient who presented with pancytopenia revealing the presence of two blastic populations in the bone marrow, the first one is purely myeloblastic (CD34-positive, MPO-positive) and the second one has a plasmacytoid dendritic differentiation. The typical skin lesions of BDPCN were absent at presentation and appeared lately. Is it a biclonal form of the same disease or a coexistence of two different malignancies? We present the clinical, the histopathological, the immunophenotypical and the cytogenetic study results.

Keywords: *Blastic dendritic plasmacytoid cell neoplasm, Acute myeloblastic leukemia, Biclonal and atypical presentation.*

Introduction

Blastic dendritic plasmacytoid cell neoplasm is a rare hematologic malignancy, primarily affecting the elderly patients, often confined to the skin at presentation, it progresses to more generalized disease almost invariably despite initial local and systemic therapy [1]. It represents 0.44% of all hematologic malignancies [2,3]. The skin manifestations are virtually present in all patients. Papules, nodules, plaques and or bruise-like lesions are seen typically prior to any hematologic manifestation [3,4,5]. Localized at onset, they become multiple and more generalized during the course of disease. 60% to 90% of patients develop subsequently mono, bi-, or pancytopenia with or without leukemia and transformation into leukemic phase frequently occurs [6,7,8,9].

No known racial, ethnic or geographic predilection is observed. Though this neoplasm can occur at any age, it affects mostly the elderly men (3.3 men: 1 woman) [1,10]. There is no known etiology. Some relation with MDS has been reported [11,12] and cases with concomitant multiple myeloma and lymphoma have been described [13,14,15]. The diagnosis is based on the skin and bone marrow biopsies and on the peripheral blood and bone marrow flow cytometry. The skin biopsy shows non-epidermotropic infiltration of the dermis by a monotonous population of intermediate sized cells. The cells have scant cytoplasm, absent or indistinct nuclei resembling lymphoblasts or myeloblasts. The flow cytometry immunophenotyping and the immunohistochemistry shows positive staining for CD4, CD56, CD43, TdT, CD123 and TCL1. The cells do not express T-cell and B-cell markers, the MPO (Myeloperoxidase), the CD34 and the CD10. In the leukemic phase, the cells are primitive of intermediate size with blast-like morphology. The TCR genes show germ line configuration [1,2,6,16,17,18].

The cytogenetic abnormalities mostly reported are Del 5q, haploid and tetraploid cells, Del 12p, Del 6q and monosomy 15, 9 [19]. The differential diagnoses are acute myeloblastic leukemia (AML) which is MPO-positive, HTLV1 leukemia/lymphoma which is CD3-positive, cutaneous NK/T-cell lymphoma which is CD4-negative, myelomonocytic AML leukemia cutis which is MPO-positive and undifferentiated carcinoma or malignant melanoma.

BDPN was first described in 1994 by Adachi et al [20]. The expression of CD56 pointed toward NK cells as the cells of origin. BDPN was formerly included in the following categories: Blastic NK-cell lymphoma (WHO), Neegative-CD30 cutaneous lymphoma (EORTC), Agranular CD4-positive NK-cell leukemia, Blastic NK leukemia/lymphoma and Agranular CD4-positive/CD56-positive hematodermic neoplasm (WHO, EORTC). Recent evidences suggest an early plasmacytoid type 2 dendritic cells (DC2) origin [7,21] and it is now clearly categorized in the WHO classification of hematological malignancies [22,23].

Case report

We present the case of a 68 year-old man, diabetic and smoker who presented with quick onset of fatigue. He denied fever, chills, weight loss, recurrent infection or bone pain. His physical examination was unremarkable with no splenomegaly, no lymph node enlargement, no skin abnormality and no abnormal mass. The lab tests revealed a pancytopenia with hemoglobin level at 6.5g/dl, platelet count at 15 000/mm3 and white blood cell count at 2500/mm3 with high LDH level, normal creatinine, normal serum protein electrophoresis and immunoelectrophoresis, normal liver function tests and negative direct Coombs test. The CT scan of the thorax, the abdomen and the pelvis were unremarkable. To further asses the pancytopenia, bone marrow aspiration and biopsy were performed and showed the presence of pathological foci of CD38-positive and CD56-positive cells. All stains with B- and T-cell markers were negative. The patient received packed red blood cells and platelets transfusion and was discharged home. He has been readmitted one month later for severe pancytopenia. On examination we noticed the emergence of 3 brown superficial plaques on the back and 2 dark nodules on the chest with the largest measuring 3 cm. His skin lesions rapidly progressed and became generalized [figure 1].

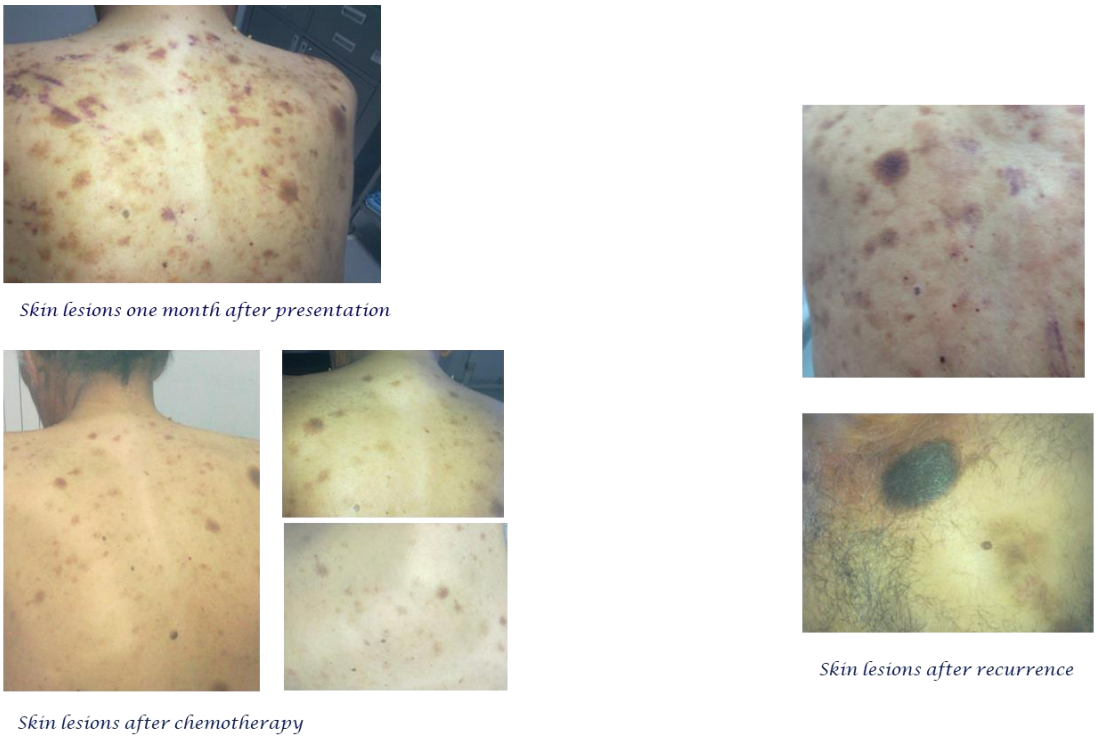

Figure 1: The consecutive skin lesions from the occurrence to the recurrence.

His blood condition required multiple transfusions. A repeated bone marrow biopsy with skin biopsy were performed. Both showed infiltrating blasts. Immunohistochemistry showed positivity for CD38 and CD56 [Figure 2,3,4] while all the B- and T-cell markers showed negative staining as well as CD30, CD117, κ- and λ- light chain and CD79a.

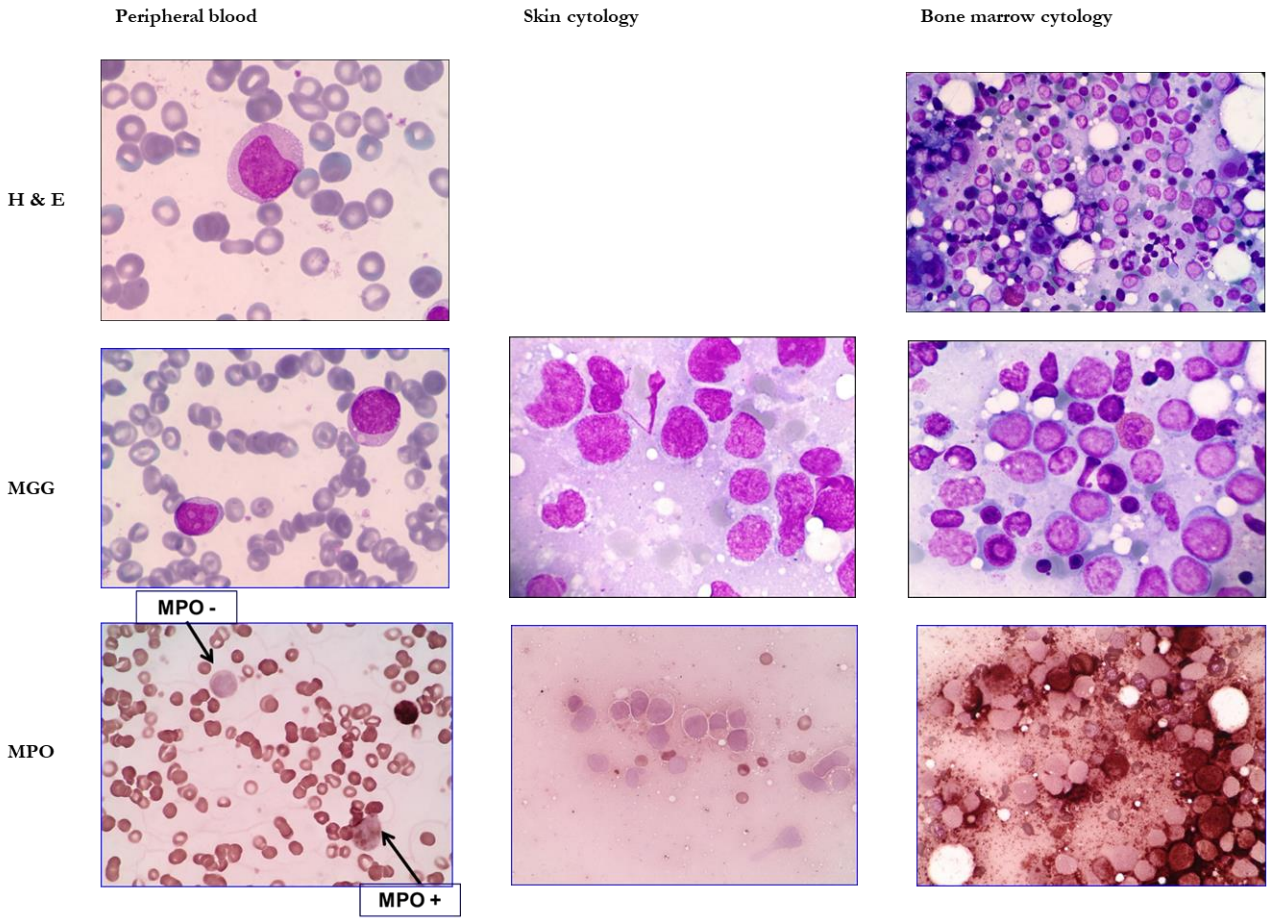

Figure 2: The peripheral blood smear showing a blast with monocytoid aspect (H&E) and two blasts (MPO+ and MPO-). The skin cytology showing the MPO- blasts. The bone marrow cytology with the two MPO type blasts. [H&E: Hematoxilin and Eosine stain, MGG: May Grunwald Giemsa stain, MPO: Myeloperoxidase stain.]

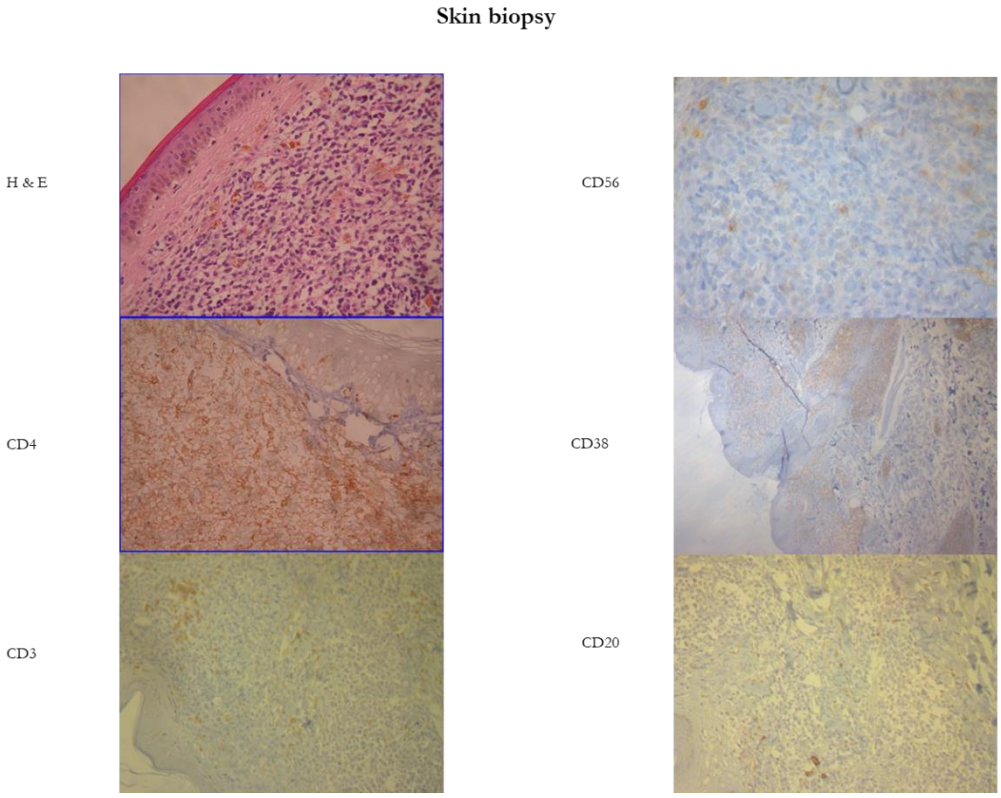

Figure 3: The skin biopsy showing positive staining for CD38, CD56, CD4 and negative staining for CD3 and CD20.

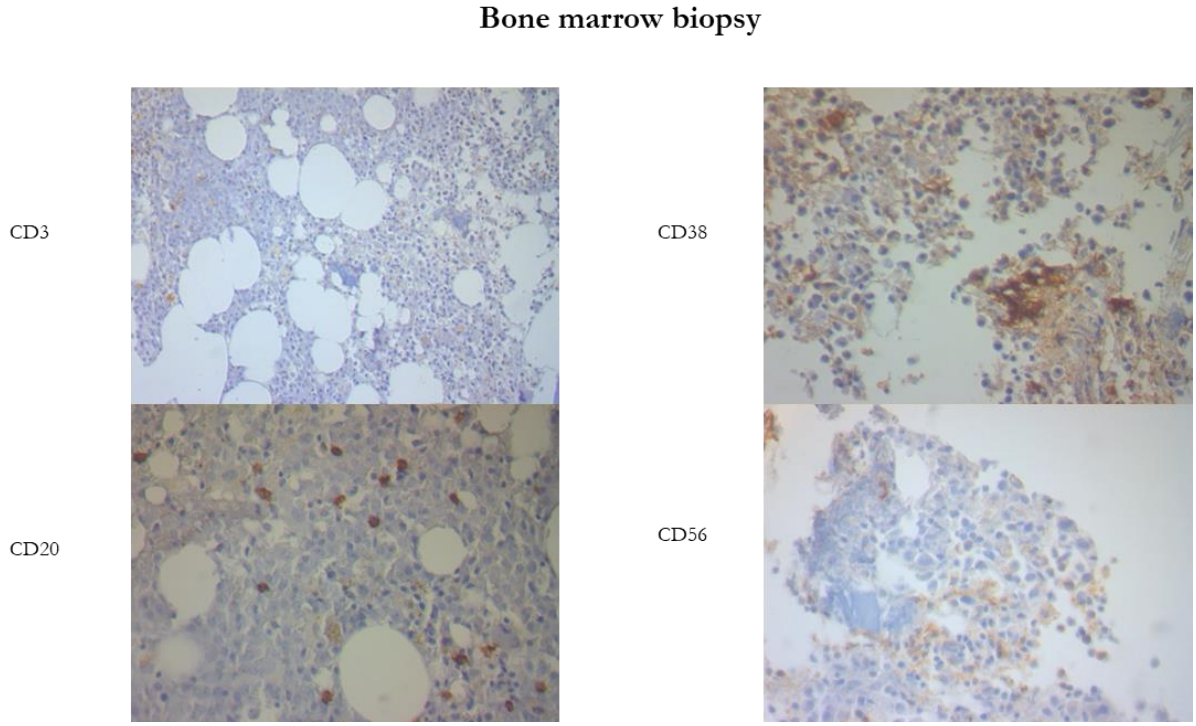

Figure 4: The bone marrow biopsy showing positive staining for CD38 and CD 56.

The bone marrow aspirate film showed 20% blastic cells most probably of myeloid origin. The bone marrow immunophenotyping revealed the presence of 2 blastic populations. The first one (P1) expressed: CD13-positive/ HLA DR-positive, CD33-positive, CD34-positive/CD117-positive, CD38-positive, MPO-positive and CD4-negative/CD56-positive. The second population (P2) presenting atypical immunophenotypic profile with plasmacytoid dendritic cells: CD13-negative/HLA DR-positive, CD33-low, CD34-negative/CD117-negative, CD38-positive, MPO-negative and CD4-positive/CD56-positive. However, in the skin biopsy the cells were expressing exclusively the P2 immunoprofile [figures 5,6,7]. The cytogenetic investigations analyzed thirty mitotic bone marrow cells, 25 cells showed 46,XY, 4 cells showed 46,XY plus 10q26 and 1 cell showed 47,XY+8.[Figure 7].

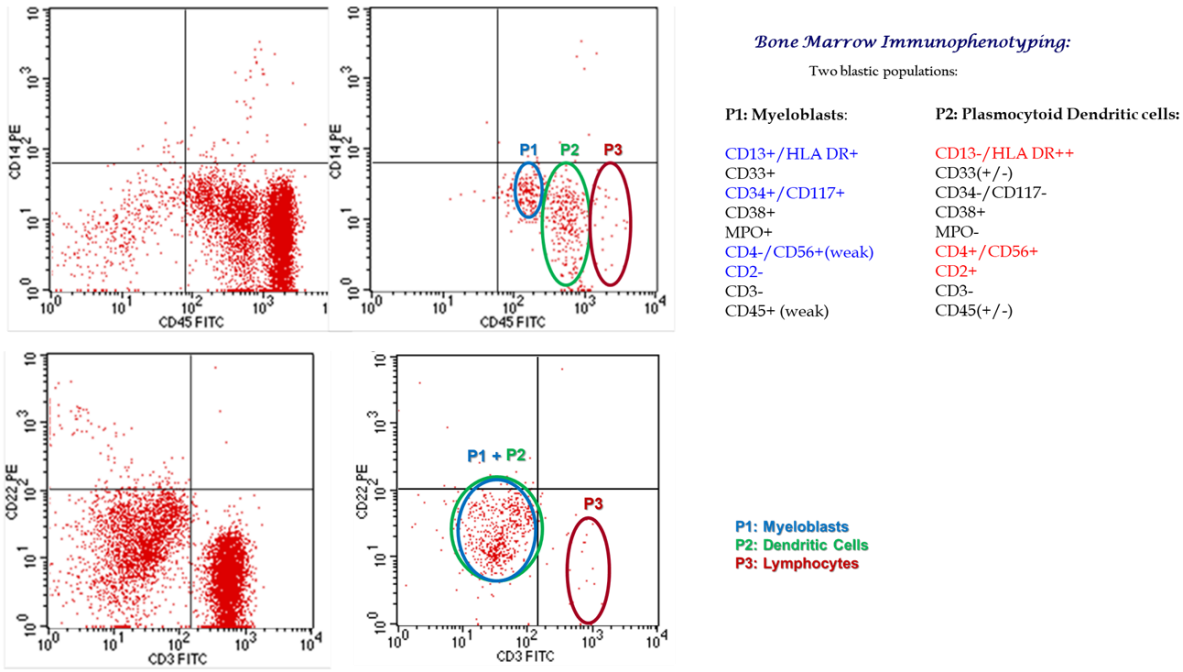

Figure 5: The flow cytometry results of the bone marrow cells with the two distinct populations.

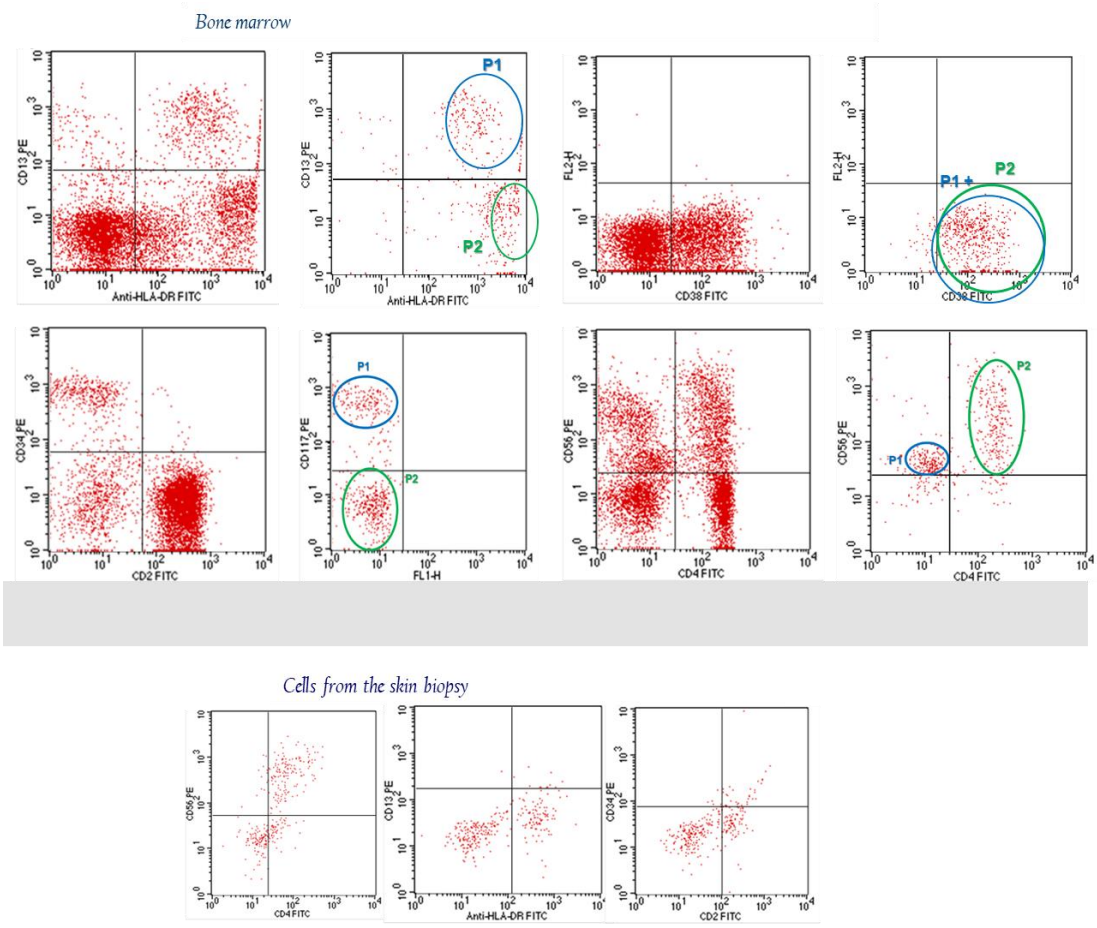

Figure 6: The flow cytometry results of the bone marrow cells and the skin cells.

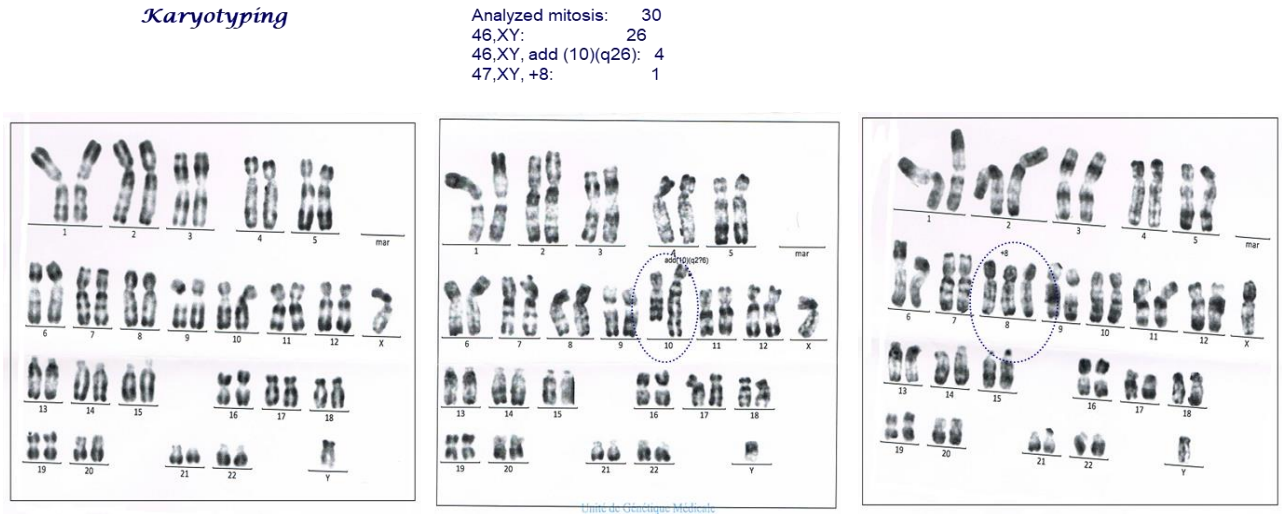

Figure 7: The cytogenetic test results.

We started a combination chemotherapy including Doxorubicine, Vincristin, Cyclophosphamide and Dexamethasone which lead to a partial remission of the skin lesions [figure 1] and a hematological complete response with no more need for transfusions. After 8 months of normal complete blood counts, the skin spots began to multiply showing black, brown and red plaques and the hematological picture began to deteriorate.

Discussion

The normal plasmacytoid monocytes, also called plasmacytoid type 2 dendritic cells (DC2) are derived from a common myeloid/NK precursor stem cells. They are present in the umbilical cord, the blood, the bone marrow and the T-cell zone of lymph node. They are small to medium-sized cells with blastoid morphology (size between lymphocyte and monocyte). They have excentric nuclei and agranular, pale eosinophilic cytoplasm. They are strongly positive for CD123 (IL-3α), TCL2 (T cell leukemia 1) and TCLA. Blastic plasmacytoid dendritic cell neoplasm is a very rare aggressive malignancy of cutaneous and hematopoietic tropism. It was characterized by the co-expression of CD4 and CD56 and the absence of common myeloid, B- and T-cell expression. Recently, the use of CD123 has supported the positive diagnosis. Typically BDPCN manifests as cutaneous lesions and later progresses to involve the bone marrow and the peripheral blood. Our patient had reverse presentation that initially involved the bone marrow and later manifested in the skin. Interestingly, our patient had two distinct blast populations, myeloblastic and plasmacytoid. The last one could have developed later in the course of disease [1,2,6,10,18].

Given the rarity and the indolent course of this disease, the definitive diagnosis is usually delayed. The typical form requires a triple positivity for CD4, CD56 and CD123 marker staining along with negative staining for myeloid and lymphoid markers. In our case, the presence of 2 malignant cell populations in the bone marrow might have contributed to the delay in the definitive diagnosis and could explain the atypical presentation. The concomitant occurrence of two hematological malignancies is rare [13, 14, 15, 24, 25, 26, 27].it should be suspected in situations with unusual or unexpected findings. Simultaneous occurrence of two different lymphoid malignancies, multiple myeloma with myeloid malignancy or lymphoma with myeloid malignancy have been described [13, 14]. However, the coexistence of acute myeloblastic leukemia with BDPCN is still a matter of controversy. Our results imply that leukemic cells infiltrating the bone marrow are derived from the myeloid lineage. There are two distinct forms of neoplasms derived from pDCs; the one is blastic plasmacytoid dendritic cell neoplasm (BPDCN) and the other is mature pDCs proliferation coexisted with myeloid tumors. Huang et at [24] suggested that there are many kinds of plasmablastic dendritic cells (pDC) and there is a broader spectrum of pDC-associated neoplasms than those currently recognized; blastic-PDCN and mature-PDCN. However, AML and MDS with different immunophenotypes not related to plasmacytoid dendritic one are different entities. In a relatively similar case to ours Wang et al [26] reported that the second myeloid population of blasts in the bone marrow is no other than a mature plasmacytoid dendritic cell proliferation. Moreover, Rigolin et al [27] reported that different populations of pathological dendritic cells may coexist in the same patient. Consequently, the presence of the second population seen on flow cytometry of the bone marrow cells in our patient may be a part of the hematological and immunophenotypic picture of BDPCN. The aberrant karyotype found in our patient displayed unspecific abnormalities. However aberrations on chromosome 8 were more frequently observed [26].

Although patients with BDPCN may initially show a response to Doxorubicin-based chemotherapy like in this case, relapses are common and the median overall survival remains approximately one year. Intensification along with allogeneic hematopoietic stem cell transplantation could offer better results.

Conclusion

This report shows the morphologic, the histopathologic, the immunophenotypic and the cytogenetic characteristics of an atypical case of BDPCN with the hematologic manifestations preceding the skin lesions and the coexistence of two different blast populations in the bone marrow. One population consists of plasmacytoid dendritic cells which is also present in the skin lesions and the other marrow population consists of myeloblastic cells not present in the skin lesions.

References

1. Cazzato G, Capuzzolo M, Bellitti E, De Biasi G et al. Blastic Plasmocytoid Dendritic Cell Neoplasm (BPDCN): Clinical Features and Histopathology with a Therapeutic Overview. Hematol Rep. 2023 Dec 8;15(4):696-706. doi: 10.3390/hematolrep15040070. PMID: 38132278; PMCID: PMC10742669.
2. Owczarczyk-Saczonek A, Sokołowska-Wojdyło M, Olszewska B, Malek M et al. Clinicopathologic retrospective analysis of blastic plasmacytoid dendritic cell neoplasms. Postepy Dermatol Alergol. 2018 Apr;35(2):128-138. doi: 10.5114/ada.2017.72269. Epub 2018 Apr 24. PMID: 29760611; PMCID: PMC5949541.
3. Pagano L, Valentini CG, Pulsoni A, Fisogni S et al. Blastic plasmacytoid dendritic cell neoplasm with leukemic presentation: an Italian multicenter study. Haematologica. 2013 Feb;98(2):239-46. doi: 10.3324/haematol.2012.072645. Epub 2012 Oct 12. PMID: 23065521; PMCID: PMC3561431.
4. Ng AP, Lade S, Rutherford T, McCormack C et al. Primary cutaneous CD4+/CD56+ hematodermic neoplasm (blastic NK-cell lymphoma): a report of five cases. Haematologica. 2006 Jan;91(1):143-4. PMID: 16434387.
5. Julia F, Dalle S, Duru G, Balme B et al. Blastic plasmacytoid dendritic cell neoplasms: clinico-immunohistochemical correlations in a series of 91 patients. Am J Surg Pathol. 2014 May;38(5):673-80. doi: 10.1097/PAS.0000000000000156. PMID: 24441662.
6. 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 May;127(5):687-700. doi: 10.1309/FY6PK436NBK0RYD4. PMID: 17439829.
7. Petrella T, Comeau MR, Maynadié M, Couillault G et al. 'Agranular CD4+ CD56+ hematodermic neoplasm' (blastic NK-cell lymphoma) originates from a population of CD56+ precursor cells related to plasmacytoid monocytes. Am J Surg Pathol. 2002 Jul;26(7):852-62. doi: 10.1097/00000478-200207000-00003. PMID: 12131152.
8. Adnan A, Powell PR, Staples CJ, Evanson B, Parekh PK. Blastic Plasmacytoid Dendritic Cell Neoplasm: A Case Series and Review. Am J Dermatopathol. 2021 May 11. doi: 10.1097/DAD.0000000000001968. Epub ahead of print. PMID: 33989217.
9. Dantas FE, de Almeida Vieira CA, de Castro CC, Neto GC, Matos DM et al. Blastic plasmacytoid dendritic cell neoplasm without cutaneous involvement: a rare disease with a rare presentation. Acta Oncol. 2012 Jan;51(1):139-41. doi: 10.3109/0284186X.2011.602110. Epub 2011 Aug 28. PMID: 21871006.
10. Garnache-Ottou F, Vidal C, Biichlé S, Renosi F, Poret E et al. How should we diagnose and treat blastic plasmacytoid dendritic cell neoplasm patients? Blood Adv. 2019 Dec 23;3(24):4238-4251. doi: 10.1182/bloodadvances.2019000647. PMID: 31869411; PMCID: PMC6929390.
11. Chamoun K, Loghavi S, Pemmaraju N, Konopleva M et al. Early detection of transformation to BPDCN in a patient with MDS. Exp Hematol Oncol. 2018 Oct 6;7: 26. doi: 10.1186/s40164-018-0117-6. PMID: 30323983; PMCID: PMC6174068.
12. Ma L, Delforge M, van Duppen V, Verhoef G, Emanuel B et al. Circulating myeloid and lymphoid precursor dendritic cells are clonally involved in myelodysplastic syndromes. Leukemia. 2004 Sep;18(9):1451-6. doi: 10.1038/sj.leu.2403430. PMID: 15284864.
13. Lu-qun, W., Hao, L., Xiang-xin, L. et al. A case of simultaneous occurrence of acute myeloid leukemia and multiple myeloma. BMC Cancer 15, 724 (2015). https://doi.org/10.1186/s12885-015-1743-6
14. Bhar VS, Gupta V, Sharma M, Dhawan R et al. Rare Coexistence of Acute Monoblastic Leukemia with Chronic Lymphocytic Leukemia. Case Rep Hematol. 2018 Nov 6;2018: 6452843. doi: 10.1155/2018/6452843. PMID: 30524762; PMCID: PMC6247670.
15. Jeong D, Lee AC, Shin K, Kim J et al. Hema-seq reveals genomic aberrations in a rare simultaneous occurrence of hematological malignancies. Cell Rep Methods. 2023 Oct 23;3(10):100617. doi: 10.1016/j.crmeth.2023.100617. Epub 2023 Oct 17. PMID: 37852254; PMCID: PMC10626221.
16. Feuillard J, Jacob MC, Valensi F, Maynadié M et al. Clinical and biologic features of CD4(+)CD56(+) malignancies. Blood. 2002 Mar 1;99(5):1556-63. doi: 10.1182/blood.v99.5.1556. PMID: 11861268.
17. Cronin DM, George TI, Reichard KK, Sundram UN et al. Immunophenotypic analysis of myeloperoxidase-negative leukemia cutis and blastic plasmacytoid dendritic cell neoplasm. Am J Clin Pathol. 2012 Mar;137(3):367-76. doi: 10.1309/AJCP9IS9KFSVWKGH. PMID: 22338048.
18. Martín JM, Nicolau MJ, Galán A, Ferrández-Izquierdo A, Ferrer AM, Jordá E. CD4+/CD56+ haematodermic neoplasm: a preculsor haematological neoplasm that frequently first presents in the skin. J Eur Acad Dermatol Venereol. 2006 Oct;20(9):1129-32. doi: 10.1111/j.1468-3083.2006.01743.x. PMID: 16987271.
19. Leroux D, Mugneret F, Callanan M, Radford-Weiss I et al. CD4(+), CD56(+) DC2 acute leukemia is characterized by recurrent clonal chromosomal changes affecting 6 major targets: a study of 21 cases by the Groupe Français de Cytogénétique Hématologique. Blood. 2002 Jun 1;99(11):4154-9. doi: 10.1182/blood.v99.11.4154. PMID: 12010820.
20. Adachi M, Maeda K, Takekawa M, Hinoda Y et al. High expression of CD56 (N-CAM) in a patient with cutaneous CD4-positive lymphoma. Am J Hematol. 1994 Dec;47(4):278-82. doi: 10.1002/ajh.2830470406. PMID: 7526680.
21. Niakosari F, Sur M. Agranular CD4+/CD56+ hematodermic neoplasm: a distinct entity described in the recent World Health Organization-European Organization for Research and Treatment of Cancer classification for cutaneous lymphomas. Arch Pathol Lab Med. 2007 Jan;131(1):149-51. doi: 10.5858/2007-131-149-ACHNAD. PMID: 17227117.
22. Cree IA. The WHO Classification of Haematolymphoid Tumours. Leukemia. 2022 Jul;36(7):1701-1702. doi: 10.1038/s41375-022-01625-x. Epub 2022 Jun 22. PMID: 35732830; PMCID: PMC9252902.
23. 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 May;127(5):687-700. doi: 10.1309/FY6PK436NBK0RYD4. PMID: 17439829.

24. Huang Y, Wang Y, Chang Y, Yuan X et al. Myeloid Neoplasms with Elevated Plasmacytoid Dendritic Cell Differentiation Reflect the Maturation Process of Dendritic Cells. Cytometry A. 2020 Jan;97(1):61-69. doi: 10.1002/cyto.a.23953. Epub 2019 Dec 26. PMID: 31876105.
25. Kotchetkov R, Ellison E, McLean J, Pressnail B, Nay D et al. Synchronous dual hematological malignancies: new or underreported entity? Hematology. 2018 Oct;23(9):596-599. doi: 10.1080/10245332.2018.1466428. Epub 2018 Jun 7. PMID: 29874974.
26. Wang M, Chen YJ, Wang LR, Wang YZ, Lu J et al. Plasmacytoid Dendritic Cells Proliferation Coexisted with Acute Myeloid Leukemia. Chin Med J (Engl). 2018 Aug 5;131(15):1866-1867. doi: 10.4103/0366-6999.237404. PMID: 30058585; PMCID: PMC6071469.
27. Rigolin GM, Della Porta M, Bigoni R, Tieghi A et al. Dendritic cells in acute promyelocytic leukaemia. Br J Haematol. 2001 Sep;114(4):830-3. doi: 10.1046/j.1365-2141.2001.03023.x. PMID: 11564070.