

## Case Report

# Blastic Plasmacytoid Dendritic Cell Neoplasm: A Rare Case Report

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## ABSTRACT

The rare haematologic malignancy dubbed blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a pathological condition characterised by the proliferation of immature plasmacytoid dendritic cells, typically presenting with cutaneous lesions and systemic symptoms that incidentally present an aggressive clinical course. A 68-year-old male presented with erythematous skin nodules characterised by a proliferation of immature plasmacytoid dendritic cells, and consistent with a diagnosis of BPDCN. Histological examination of skin biopsy revealed a neoplasm composed of a monomorphous population of cells co-expressing cluster of differentiation (CD) CD123, CD56 and focal CD4, further confirming the BPDCN diagnosis. Bone marrow evaluation showed no significant involvement. Initial treatment with steroids led to partial regression of skin lesions, but recurrence necessitated chemotherapy using a modified Mitoxantrone, Chlorambucil, Prednisolone (MCP) 841 regimen, which led to significant clinical improvement initially. This case accentuates the diagnostic value and critical role of immunohistochemistry in diagnosing BPDCN. Early diagnosis and initiation of therapy are critical in prognosis, and emerging targeted therapies offer new hope in the treatment of this disease.

**Keywords:** Blastic plasmacytoid dendritic cell neoplasm, CD123, Cutaneous lesions, Immunohistochemistry

## INTRODUCTION

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare and aggressive haematologic malignancy that often presents with the proliferation of immature cells with plasmacytoid dendritic cell differentiation.<sup>[1]</sup> BPDCN's estimated incidence approximates to only around 0.04 cases/100,000 individuals, with fewer than 200 cases reported globally.<sup>[1]</sup> Although it often presents with cutaneous lesions, it can also compromise other organs such as the bone marrow, central nervous system (CNS) and lymph nodes. The disease is recognised for its challenging diagnosis and poor prognosis. Its cutaneous clinical manifestations are diverse, ranging from isolated brown or violaceous nodules or patches to disseminated mixed lesions. Non-cutaneous presentation, especially bone marrow involvement, correlates with poorer overall survival.<sup>[2]</sup> The diagnosis of BPDCN is rendered on the grounds of clinical findings, histopathological features and characteristic immunophenotyping. We present a case of BPDCN in a 68-year-old man, detailing its clinical presentation, diagnostic evaluation and therapeutic management. It also simultaneously attempts to emphasise the diagnostic challenges accompanying BPDCN due to its rarity and heterogeneous clinical presentation.

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## CASE REPORT

A 68-year-old male presented with multiple erythematous to violaceous lesions and skin nodules all over the body for 4 months. On examination, varying-sized firm nodules ranging in size from 0.5 cm to 2 cm were found. He also exhibited constitutional symptoms such as fatigue, weight loss and low-grade fever. The liver and spleen were not palpable on physical examination and there was no generalised lymphadenopathy. On laboratory investigations, he was found to have a haemoglobin level of 9.5 g/dL and a lactate dehydrogenase level of 107 IU/L. Peripheral smear examination showed normocytic, normochromic anaemia with polychromatic cells. The patient subsequently tested negative for the Breakpoint Cluster Region - Abelson (BCR-ABL) translocation.

An excision biopsy was performed on a 2 × 1 cm skin nodule from the patient's chest [Figure 1]. The histological examination revealed a neoplasm located in the dermis composed of a monomorphous population consisting of medium-sized cells with hyperchromatic, vesicular nuclei [Figure 2a and b]. Immunohistochemical staining was performed to further characterise the neoplastic cells.

The immunohistochemical profile of the lesion demonstrated diffuse positivity for CD123 and CD56, with focal positivity for CD4 [Figure 3a and b]. The Ki-67 proliferation index was noted to be 35–40. CD45, myeloid markers (CD117), B cell markers PAX5, CD10, Bcl2, CD20, CD19, CD1a and Terminal deoxynucleotidyl transferase (Tdt) were negative.

These features substantiate a diagnosis of BPDCN. Subsequent bone marrow evaluation with immunohistochemistry revealed only trilineage haematopoiesis without significant or identifiable plasmacytoid dendritic cell populations. No significant evidence of myeloproliferative or lymphoproliferative disorders was observable.



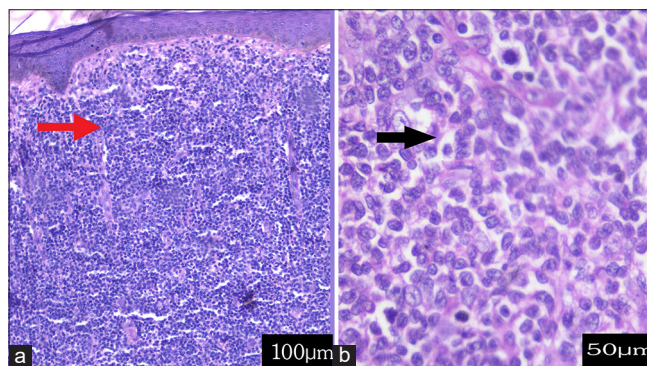
**Figure 1:** Erythematous to violaceous skin nodules in chest.

The clinical presentation, histological features and immunohistochemical findings corroborated the BPDCN diagnosis. Although World Health Organization (WHO) does not recommend a formal BPDCN staging system, baseline status is still noted. Our patient presented with preserved general health. He was empirically started on corticosteroids, which led to regression of the cutaneous lesions. However, upon tapering the steroids, the lesions began to reappear. Subsequently, chemotherapy was initiated using a modified MCP-841 regimen (Vincristine and Adriamycin), resulting in significant clinical improvement.

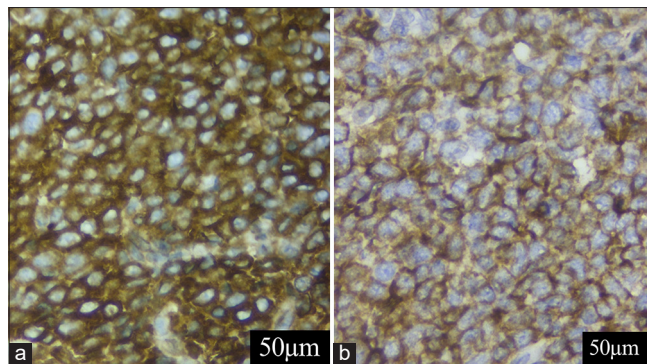
At a later follow-up after 7 months, the patient's symptoms worsened, with the development of neurological manifestations. Cerebrospinal fluid analysis confirmed CNS involvement by BPDCN through flow cytometry. Despite further management, the patient's condition deteriorated, and he eventually succumbed to the disease.

## DISCUSSION

The predominant demographic afflicted with BPDCN,



**Figure 2:** (a) Haematoxylin and eosin staining of a skin nodule biopsy from chest shows ( $\times 10$  magnification) a dermal neoplasm (red arrow). (b) Haematoxylin and eosin staining of dermal neoplasm composed of monomorphous population of medium-sized cells with hyperchromatic, vesicular nuclei (black arrow). ( $\times 40$  magnification).



**Figure 3:** (a) Immunohistochemical staining (IHC) demonstrated diffuse positivity for CD123. (b) IHC staining demonstrated diffuse positivity for CD56.

a recently described highly invasive haematologic malignancy, is older adults, with a diagnosis age range of 62–70 years.<sup>[1]</sup> It is distinguishable by atypical proliferation of immature plasmacytoid dendritic cells, which are involved in immune responses to viral infections. Plasmacytoid dendritic cells of the symptomatic individuals typically express CD123 and CD56. In addition, they can show patchy expression of CD7, Tdt and CD4 while being negative for expression of myeloid lineage markers (CD117, CD34, MPO, CD13) and the expression of B and T lymphoid markers (e.g., CD3, CD20, PAX5, CD19) and a moderately high Ki-67 index. As in our case, neoplastic cells exhibited blastoid morphology and expressed markers such as CD123, CD56 and CD4 (focal), while lacking myeloid or lymphoid lineage markers.

BPDCN often presents with cutaneous lesions, which are erythematous, nodular or purplish in appearance. Our patient also had a similar presentation. Although skin involvement is common, bone marrow involvement is variable and when present, it may contribute to anaemia, thrombocytopenia or leukopenia. The differential diagnosis includes other haematologic malignancies, such as acute myeloid leukaemia (including acute blastoid leukaemia), lymphoma or myeloid sarcoma, the possibility of which was eliminated based on the absence of lineage-specific markers and the existence of the unique immunophenotypic profile of BPDCN.<sup>[3]</sup>

The prognosis for BPDCN remains poor, resulting in a median survival of approximately 14 months. The disease is highly resistant to conventional therapies. Advances in the understanding of BPDCN have led to the development of novel therapies targeting CD123 (a marker expressed on plasmacytoid dendritic cells).<sup>[4]</sup>

This case points to the importance of considering a provisional diagnosis of BPDCN in patients presenting with skin lesions and systemic symptoms. BPDCN is a highly invasive haematopoietic tumour that requires a high index of suspicion for timely diagnosis and management. Immunohistochemistry becomes a salient tool in confirming the diagnosis and distinguishing BPDCN from other haematologic and cutaneous malignancies. Given the poor prognosis, early diagnosis and enrollment in clinical trials for novel therapies are critical to improving patient outcomes. Each newly diagnosed case should undergo a complete initial evaluation, including a bone marrow examination, to optimise treatment strategies and improve prognosis.

## CONCLUSION

BPDCN is a rare and aggressive malignancy requiring high clinical suspicion. Immunohistochemistry is essential for diagnosis, and early evaluation with timely therapy can improve outcomes. Greater awareness is vital due to its variable presentation and poor prognosis.

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**Declaration of patient consent:** The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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