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Plasmacytoid Dendritic Cell-Derived Leukemia: A Case Report

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ABSTRACT

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare and aggressive hematologic malignancy (<1% of acute leukemias), predominantly affecting elderly patients. It typically presents with cutaneous lesions associated with bone marrow and peripheral blood involvement. We report an atypical pediatric case in an 11-month-old infant without skin lesions. The complete blood count revealed cytopenias with leukocytosis, and bone marrow examination showed 89% blasts. Immunophenotyping demonstrated a CD4+, CD56+, CD123+, HLA-DR+ profile, consistent with BPDCN. Diagnosis relies primarily on immunophenotyping due to the absence of exclusive specific markers. Management is not standardized, but ALL-type regimens and allogeneic stem cell transplantation may improve prognosis. This case highlights the diagnostic challenges in pediatric presentations and the importance of a multidisciplinary approach.

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KEYWORDS:
Blastic plasmacytoid dendritic cell neoplasm · BPDCN · Immunophenotyping · CD4 CD56 CD123 · Cytopenia

INTRODUCTION

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare and aggressive form of acute leukemia (accounting for approximately 1% of acute leukemias), with cutaneous and extracutaneous involvement, most commonly affecting lymph nodes, peripheral blood, and bone marrow [1].

In 1995, the disease was known as CD4+ natural killer (NK) cell acute leukemia. Following the discovery of its plasmacytoid origin in 2008, the World Health Organization (WHO) introduced the designation "blastic plasmacytoid dendritic cell neoplasm" to better classify this group of hematologic malignancies [2].

It is a rare condition predominantly described in elderly adults (median age at diagnosis: 70 years) and remains exceptional in the pediatric population. We report a pediatric case illustrating this atypical presentation.

CASE REPORT

An 11-month-old male infant, the only child, presented with bone pain. His medical history revealed no consanguinity; the pregnancy had been uneventful; the child had motor developmental delay; there was no history of allergies, medication use, toxic exposure, or herbal remedies.

On general examination, the patient was tachycardic (169 bpm), tachypneic (SpO_2 = 91%), and afebrile. Mucocutaneous examination revealed generalized pallor.

Neurologically, the patient was conscious with no meningeal signs. Musculoskeletal examination revealed bone pain with bilateral lower limb edema. Mild splenomegaly was noted without hepatomegaly.

Skin examination showed generalized mucocutaneous pallor without purpura or hemorrhagic bullae.

Imaging studies (chest X-ray and thoraco-abdomino-pelvic CT scan) revealed no abnormalities.

The complete blood count demonstrated a normocytic hypochromic anemia (Hb: 6.3 g/dL; MCV: 99.7 fL; MCH: 25 pg), leukocytosis at 18,970/µL with lymphocytosis at 15,760/µL, an absolute neutrophil count of 2,900/µL, and thrombocytopenia at 58,000/µL.

Metabolic, hepatic, and renal panels were within normal limits: sodium 136 mEq/L, potassium 2.3 mEq/L, chloride 98 mEq/L, corrected calcium 76 mg/L, phosphorus 27 mg/L, creatinine 3.6 mg/L, AST 27 IU/L, ALT 12 IU/L, and LDH 369 IU/L.

Bone marrow aspirate revealed a hypercellular marrow with 89% blastic cells of heterogeneous size, featuring nuclei with reticulated nucleolated chromatin and abundant basophilic cytoplasm, occasionally showing cytoplasmic

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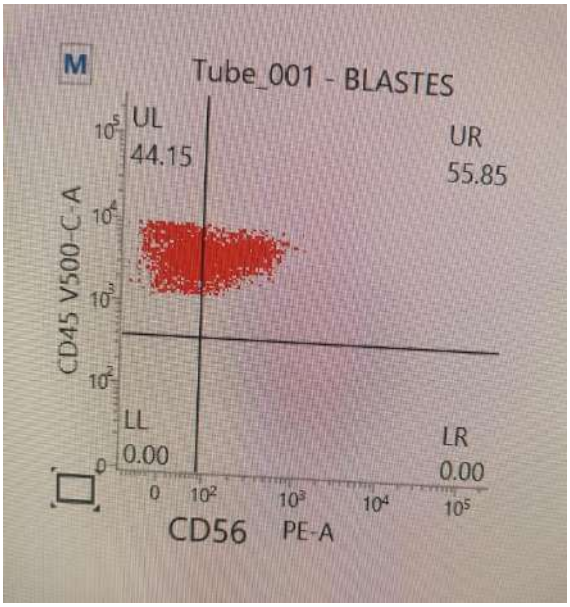
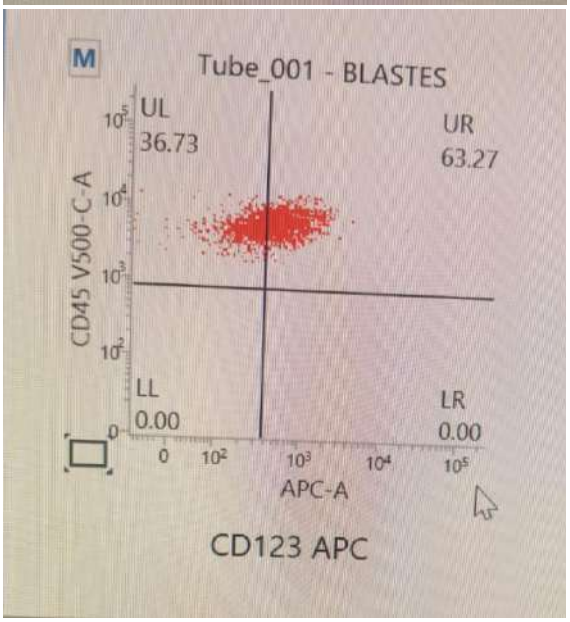
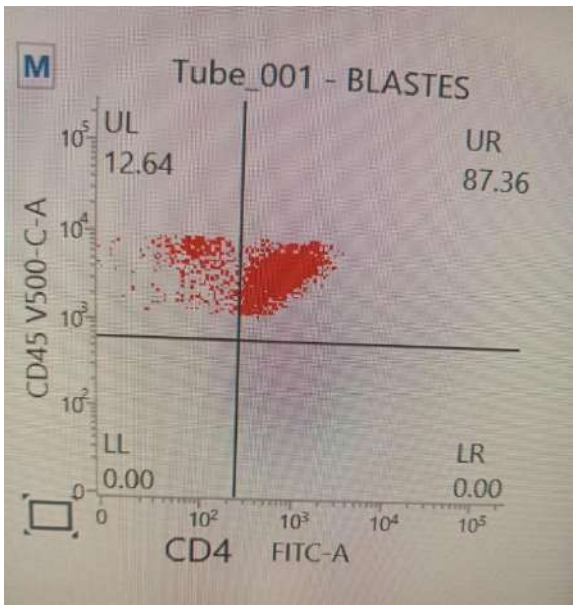
projections (Figure 1). Cytochemical staining for myeloperoxidase (MPO) on smear was negative.

Bone marrow immunophenotyping identified 80% blasts with dim CD45 expression, positive for: CD33+, MPO+, CD4+, CD56+, HLA-DR+, and CD123+, and negative for CD34, CD14, CD117, CD3, and CD7.







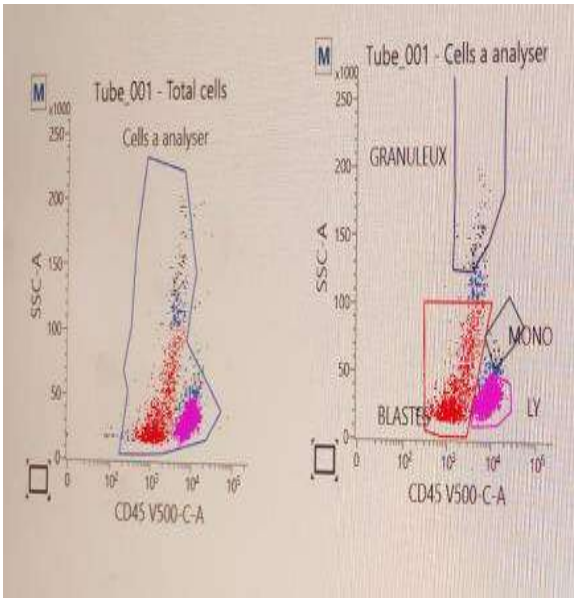


Figure 1. Patient immunophenotyping

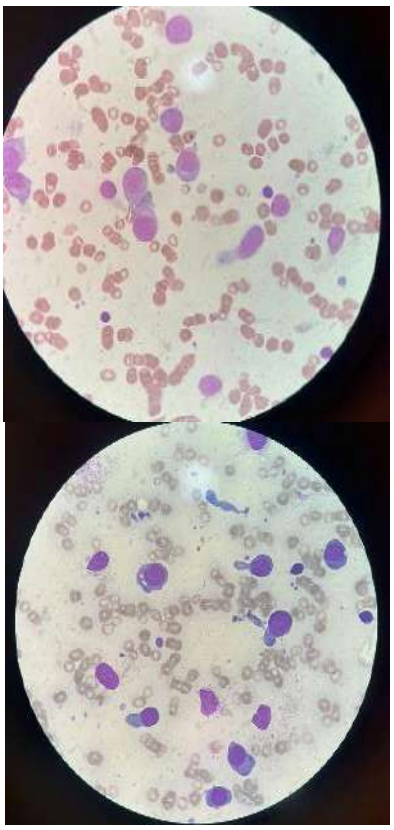


Figure 2. BPDCN blasts with cytoplasmic projections.

DISCUSSION

BPDCN is an exceedingly rare hematologic malignancy, accounting for less than 1% of acute leukemias [1].

Its pathophysiology remains incompletely understood. BPDCN derives from plasmacytoid dendritic cells, the precursors of antigen-presenting dendritic cells. An association with myelodysplasia has been described, raising the question of its potential role in pathogenesis [1].

In 2001, the disease was initially classified as a blastoid NK-cell lymphoma. In 2005, it was reclassified as an agranular CD4+ CD56+ hematodermic tumor based on its characteristic immunophenotypic profile and frequent presentation as cutaneous lesions. In 2008, the WHO included it as a subgroup of acute myeloid leukemias (AML). In 2016, it was recognized as a distinct myeloid neoplasm specific to blastic plasmacytoid dendritic cells, and in 2022, BPDCN was classified as a distinct category within acute myeloid leukemias [3].

The disease predominantly affects elderly patients (mean age at diagnosis: 60–70 years) with a strong male predominance (sex ratio 2–7:1) [4, 5].

The incidence of BPDCN in pediatric patients is extremely low, and it may be confused with other aggressive hematologic malignancies [6, 7]. No clinical, laboratory, or imaging findings have been consistently associated with prognosis. Given the limited number of cases, inclusion in collaborative or multicenter studies currently appears unlikely.

In more than 90% of cases, the disease manifests with isolated (in one-third of cases) or generalized cutaneous lesions, appearing as brownish-red to violaceous plaques or nodules of variable size (from a few millimeters to more than 10 centimeters), disseminated or grouped within a given territory [8]. There is no predilective topography; however, trunk and scalp involvement appears more frequent. Prolonged exclusively cutaneous localization (more than 6 months) is rare, as the disease rapidly disseminates to blood, bone marrow, lymph nodes, and extranodal sites. Central nervous system involvement at diagnosis is found in approximately 10% of cases; general patient condition is most often preserved at diagnosis [9].

Some pediatric patients present with isolated bone marrow involvement at diagnosis, while others may initially present with cutaneous erythema and lymphadenopathy, often associated with fulminant disseminated intravascular coagulation (DIC), before rapidly developing bone marrow and CNS disease. These presentations can vary within the pediatric age group, though this is difficult to characterize given the limited number of reported cases. Unlike adult patients, pediatric patients frequently exhibit multiorgan involvement, DIC, and tumor lysis syndrome [10].

On blood count, most patients, as in our case, present with cytopenias related to bone marrow infiltration. Thrombocytopenia is present in 78% of cases at diagnosis, while anemia and neutropenia are found in approximately one-third of cases [1].

Bone marrow infiltration at diagnosis is present in 87% of cases at variable levels, with blasts featuring round or oval, often nucleolated nuclei, fine chromatin, and slightly basophilic cytoplasm that is sometimes granular, displaying a heterogeneous structure with characteristic microvacuoles of plasmacytoid dendritic cells and pseudopod formation [1].

Immunophenotyping is essential for the diagnosis of BPDCN. Diagnosis relies on the identification of a CD4+ CD56+ blastic population in skin biopsy, peripheral blood, bone marrow, or cerebrospinal fluid [4], combined with the absence of strong lineage-specific markers for myeloid, B-lymphoid, and T-lymphoid lineages (cytoplasmic MPO, cytoplasmic CD3, cytoplasmic CD79a, and monocytic markers). In this context, expression of markers associated with the pDC lineage — CD123 (IL-3 receptor), CD303, CD304, and TCL1 — confirms the diagnosis (Figure 1) [5]. Additionally, blasts strongly express HLA-DR and CD36, while CD34 is typically absent. Isolated expression of lineage-associated markers such as CD33, CD2, CD7, and CD22 is relatively frequent (CD33 was positive in our case) and should not exclude this diagnosis. The differential diagnosis with CD4+ CD56+ AML-M5 or undifferentiated acute myeloid leukemia may arise; in this context, CD123 expression alone is insufficient to confirm a diagnosis of BPDCN [8].

Two-thirds of BPDCN patients have cytogenetic abnormalities at diagnosis. In most cases, karyotypes are complex and may involve up to six different chromosomal

regions, including chromosome 13. Loss of the Y chromosome, however, is not recurrent in BPDCN. In reported cases, no association with Herpesviridae — particularly Epstein-Barr virus (EBV) — has been established, in contrast to what is observed in mature NK-cell disorders [1].

There is no consensus regarding the management of BPDCN patients; some studies have shown rapid clinical response with AML- or ALL-type chemotherapy regimens [5, 6]. To date, allogeneic hematopoietic stem cell transplantation remains the only approach to have achieved sustained cytological remission beyond 60 months of follow-up [10].

Given the small number of pediatric BPDCN patients and the lack of randomized clinical trials or even robust case series, treatment regimens used for pediatric BPDCN vary considerably across practitioners and institutions [11].

In pediatric BPDCN patients, high-risk ALL-based treatment protocols using multi-drug combinations with maintenance therapy, including CNS prophylaxis, have yielded encouraging survival outcomes [12].

CONCLUSION

The diagnosis of BPDCN is not always straightforward, neither from a hematological standpoint nor by flow cytometry, due to the absence of validated specific markers and the variability in presentation relative to the consensus definition.

This case illustrates an atypical presentation of acute plasmacytoid dendritic cell leukemia in an infant — a condition classically described in the elderly. The absence of cutaneous lesions, otherwise considered a hallmark clinical sign, made diagnosis more challenging, underscoring the importance of a multidisciplinary approach and early recourse to immunophenotypic analysis for the accurate identification of this rare pediatric entity.

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