

## Case Report

# Blastic Plasmacytoid Dendritic Cell Leukemia in a Black Malian

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### INTRODUCTION

**B**lastic plasmacytoid dendritic cell neoplasm (BPDCN) is an acute myeloblastic leukemia (AML) characterized by the clonal proliferation of precursors of plasmacytoid dendritic cells. It is categorized as an acute myeloid neoplasm by the 2008 world health organization classification of neoplasms. Over 90% of cases present with skin lesions in the form of our knowledge; this is the first reported case of BPDCN in black African people.

### CASE REPORT

In January 2014, a 19-year-old Malian female was presented to the Hopital du Mali with anemia, fever, weakness, and weight loss. She had an antecedent of periauricular lymph node and dysphasia three months ago before her admission. Her clinical examination revealed an unestimable wasting, conjunctival pallor, and inguinal and axillaries lymphadenopathy measuring of 2 cm, in diameter and body temperature at 37.4°C. The complete blood count found

### ABSTRACT

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare, clinically aggressive hematologic malignancy, that most commonly manifests as cutaneous lesions. A 19-year-old Malian female was admitted to the Unit of Medicine of Hopital du Mali with anemia, fever, weakness, and weight loss. On physical examination she was wasted, pale, febrile (37.4°C), and had inguinal and axillary lymphadenopathies. The complete blood count found pancytopenia with Hemoglobin level of 4.8 g/dL, Leucocytes count of 1900/ $\mu$ L (neutrophil: 300/ $\mu$ L), and platelets count of 56 000/ $\mu$ L. The ultrasonographic examination found hepatomegaly and splenomegaly. The bone marrow biopsy and flow cytometer analysis were in keeping with a diagnosis of BPDCN. The patient, unfortunately, was lost four months later after her hospital admission due to late diagnosis by septicemia. The early diagnosis and availability of specific drugs for acute leukemia could improve the clinical outcome of patients with BPDCN in Mali.

**KEYWORDS:** Acute Leukemia, black african, dendritic cell, Mali

pancytopenia with hemoglobin level of 4.8 g/dL, (MCV: 79.3 fL, MCHC: 26.1 pg), a leucocytes count of 1900/ $\mu$ L (neutrophil: 300/ $\mu$ L), and platelets count of 56000/ $\mu$ L. The haemostatic and renal function tests were, however, normal. Abdominal Ultrasonographic Scan found hepato-splenomegaly measuring 17.9 cm and 14.9 cm, respectively. The bone marrow aspiration completed by bone biopsy (performed in Montfermeil-France), showed a diffuse infiltration by dendritic cells. This was characterized by small and intermediate sized cells with a moderately abundant cytoplasm and heterogeneous nuclear [Figure 1]. The chromatin is fine without nucleoli. The nucleus is sometime heterogeneous with variables forms with significant mitotic activity. The Reticulin stain found a moderate accentuation of fibrous non myelofibrosis tissue.

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[Figures 1-8] The cells were positive for CD45+, CD4+, CD56+, TCL1, and CD7+; and negatives for CD3, CD10, CD23, CD20, CD30, CD79a, CD163, CD68, CD123, CD303, TiA1, Granzym B, and Perforin at flow cytometry, were performed. The above findings led to a diagnosis of BPDCN. The immunohistochemical detection of EBNA was negative, but we could not confirm the negativity for

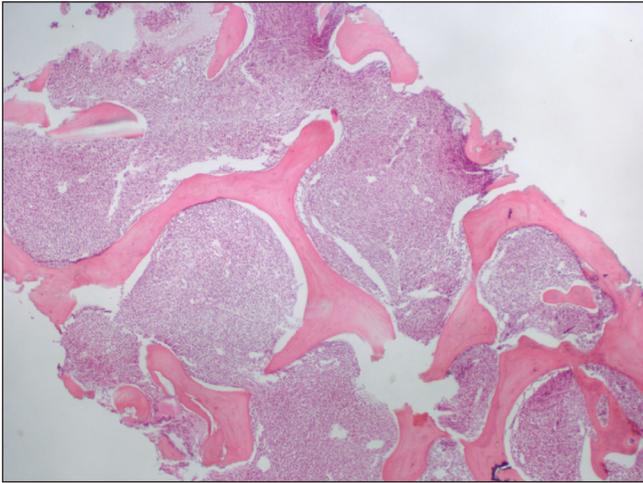
EBV by in situ hybridization with EBER-EBV, which was not interpretative.

The patients were managed with intravenous corticotherapy (1 mg/kg/day), antibiotics, and blood transfusion.

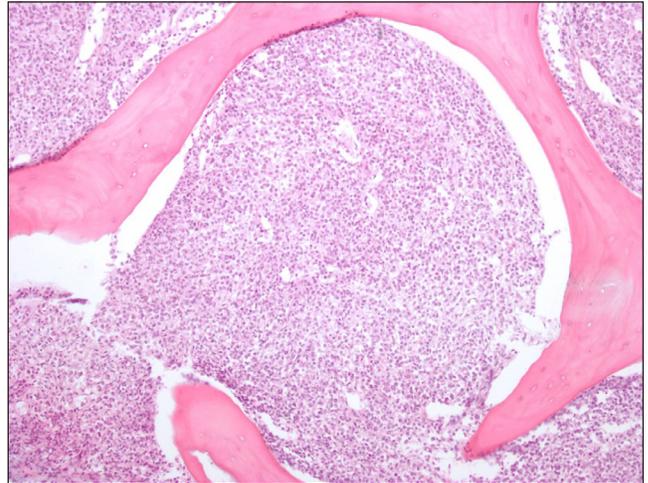
Unfortunately, she died four<sup>[4]</sup> months later due to septicemia. Cytotoxic chemotherapy could not be started because of her late presentation and abrupt evolution.

**Table 1: Different case reported in the literature**

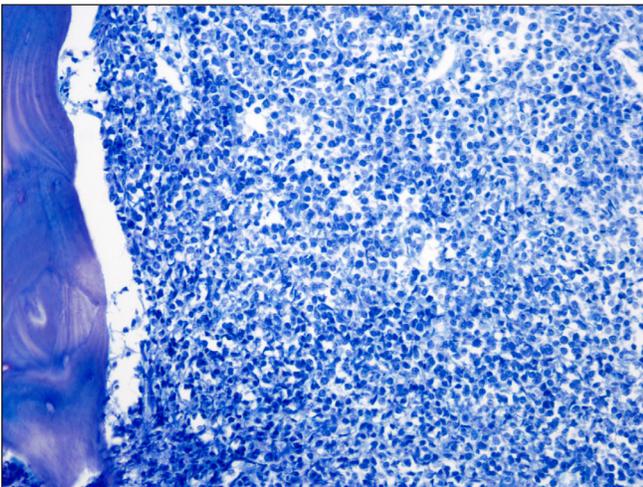
Case	Blood count	Phenotypes positive	Phenotype négative	Treatment regime	conclusion
Sahin_Turkey: Case Reports 13 20 year, woman , fatigue Hematology	LDH 1139, Hb: 127 N: 700, plat: 241	CD33, HLA-DR, CD64, CD45, CD4, CD56, CD38	cCD3, CD5, CD7, CD8, CD10, CD13, CD14, CD20, CD22, CD34, Ccd79a, CD103, CD117, MPO	Arac-Ida and Etop_ auto CSH	Auto CSH is feasible in the absence of related donor. Critère diag: CD4+, CD56+ et CD128+
Lencastre-Posrtugal: Case report 13, Cutaneous lesions, 76 year man, MDS, Prostat, neutropenia Dermatology	LDH 1139, Hb: 127 N: 700, plat: 241	CD4, CD43, CD45, CD56, CD68p, CD4, CD43, CD45, CD123, CD56, HLA-DR : BOM	CD3, CD5, CD20, CD30, CD34, CD38, CD117, TdT, MPO, IgG, PAX5, Ki-67: 60%	6CHOP: RC Relapse 2 months later death in hyperleukocytosis	Allogenic transplantation must be the solution
Lokare US, Hematology reports 14, 62 years man, nodes cutaneous 4cm Hematology	Hb:12,1, Leuk 1,7 plat:96 N:500	CD4, CD56, CD123, BDCA2, CD45, CD43, HLA-DR, CD33, CD99, CD117, CD68R	CD34, TdT, CD20,CD79a, CD3, CD5, CD2, CD7, CD8, CD57, CD30, MPO, CD13, CD14, CD16, CD11c, CD163, Perforin, Granzyme B, CD138, Ki-67: 50%	CODOX-M (Cyclo, vinc, Dox meth) IVAC (Ifos, etop, Cyta): RC AlloCSH with TBI and Alenzutumab	FISH: EBER Neg Allo with attenuate conditioning seam promising
Nomura H, Japon Acta Derma. Skin lesion (46×26) mimicking traumat purpura, 7 years Girl Dermatology	Normal	CD4,CD56, CD123, TCL1	CD3, CD8, CD20, CD79a, TiA1, Granzyme B	CHOP-LASP Allo CSH	RC Very rare in pediatric population, 8+1. Skin :83%
Sakashita K, Japan Pediatr Blood cent-2013. 5 years, F, papule (5 Times 6cm) Hematology	Hb:13,3 Leuc: 5,8 platelet 267, blast 2%, LDH:385	CD4,CD56, CD123, CD2, CD3, CD5, CD7, CD10, CD19, CD33, CD34		ALL L04-16	Skin involvement is aggressive disease, HSCT do not improve OS in pediatric => High risk ALL + CNS prophylaxis HSCT is reserved for second remission. Small size of study population. RC after HSCT for >48 months
Our Case	Hb: 4,8g/dL, (MGV: 79,3 fL, MCHT: 26,1 pg), N: 300) platelets 56 G/L	CD45+, CD4+, CD56+, TCL1 and CD7+ low.	CD3, CD10, CD23, CD20, CD30, CD79a, CD163, CD68, CD123, CD303, TiA1, Granzym B and Perforin	Cortisteroid	Death



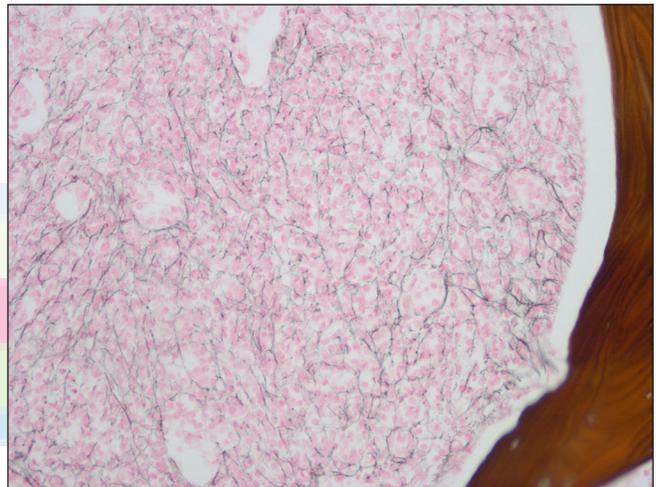
**Figure 1:** Bone marrow biopsy after Hematoxylin Eosin Coloration. View in 40



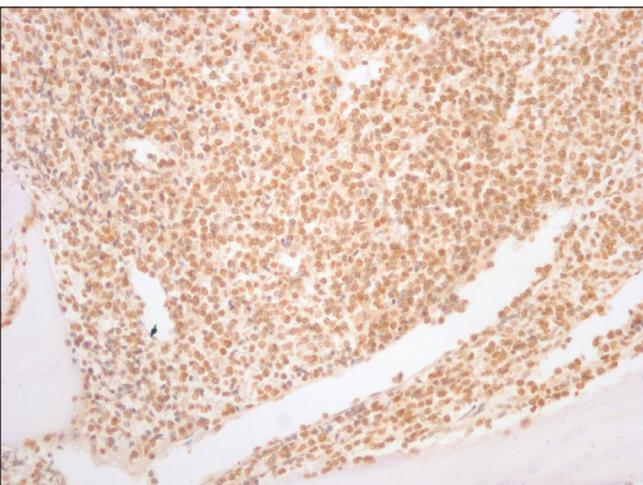
**Figure 2:** Bone marrow biopsy after Hematoxylin Eosin Coloration. View in 100



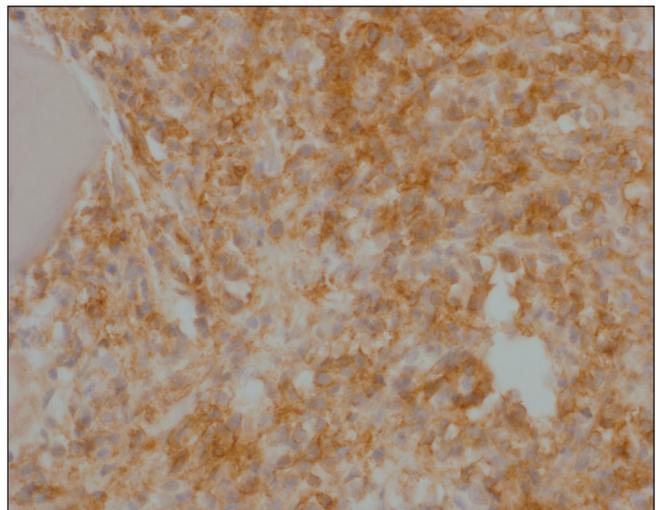
**Figure 3:** Bone marrow biopsy after May Grunewald Giemsa coloration. View in 200



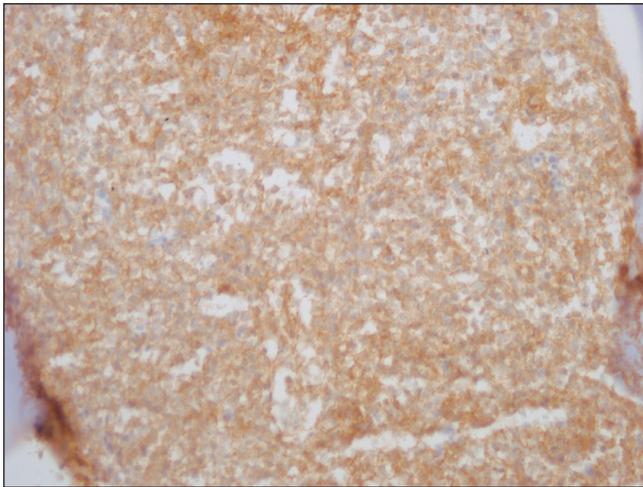
**Figure 4:** Reticulin coloration. View in 200



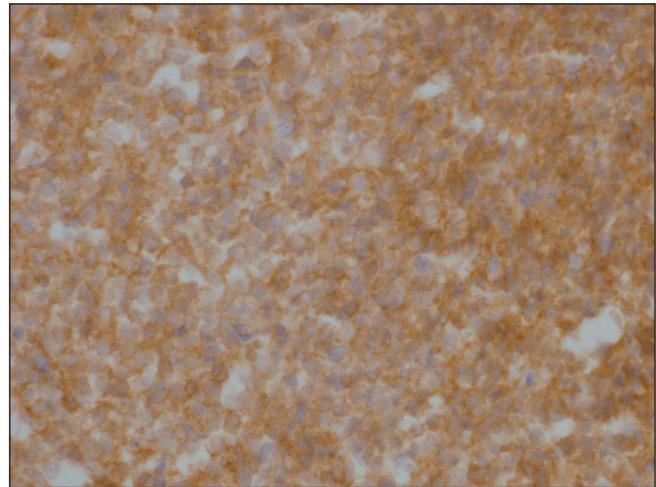
**Figure 5:** Cells positives for EBER test. View in 200



**Figure 6:** Cells positives for CD56 test. View in 400



**Figure 7:** Cells positives for CD4 test. View in 200



**Figure 8:** Cells positives for CD4 test. View in 400

## DISCUSSION

BPDCN leukemia is a very rare hematopoietic malignancy. The global incidence is estimated around 3 for 100,000 persons. The male/female ratio is 3/1. To our knowledge this is the first reported case in Mali. BPDCN is mostly reported in the pediatric and elderly population.<sup>[1]</sup> Generally, this rare and aggressive lymphoid cells neoplasm is associated with skin involvement.<sup>[2-6]</sup> The clinical presentation of BPDCN varies from none to disseminated cutaneous lesions associated with peripheral lymphadenopathy, hepatosplenomegaly, and systemic symptoms such as weakness and weight loss<sup>[7]</sup> [Table 1].

However, some rare cases without skin presentation have been reported. This index patient does not present any cutaneous lesion. The skin involvement seems a poor criteria for BPDCN evolution.<sup>[8]</sup> *Yu S. et al.* also reported a case of BPDCN without skin involvement, but the patient presented with a supraclavicular lymph node,<sup>[9]</sup> which is similar to ours.

In the case reported by *Yu S et al.*, the patient was 70 years old without cytopenia, but had atypical blast in peripheral blood contrary to the our reported case who had pancytopenia. The diagnosis was made from histology of a skin biopsy and confirmed by immunohistochemistry. The major criteria for diagnosis of a typical BPDCN is the expression of CD4, CD56, and CD123 antibody and more than 20% among marrow-infiltration blast cells.<sup>[7]</sup> However, some CD4 Negative cases were reported.<sup>[7]</sup> In this index case, cells did not express CD123, but other antibodies such as CD45, low CD7 and TCL1 were detected. The lack of evidence for association with EBNA infection and the negativity for TIA1, Granzym B, and Perforin can rule out T/NK lymphocyte cell origin. The CD31 is a marker of vascular

tissues, which is present in platelet and other blood cell such as lymphocyte, monocytes, immature dendritic cell, neutrophils, NK cell, endothelial cell, and plasma cells. It seems to play a potential role in hemorrhagic events, aggressive dissemination and chemo-resistance occurring in BPDCN.<sup>[10]</sup> Unfortunately, CD31 was not determined in this case due to its non-availability.

There are many standardized treatment regimens, and many protocols with different outcomes have been reported. Some cases with complete remission were reported under acute myeloblastic or lymphoblastic leukemia chemotherapy regimen<sup>[7,11]</sup> or chronic leukemia treatment regimen.<sup>[1]</sup>

The autologous or allogeneic stem cell transplantation (auto-SCT, allo-SCT) seems to improve the prognosis, but further studies are needed to confirm the place and the indication of this treatment strategy.<sup>[11]</sup> In pediatric population, *Sakashita et al.* reported that allo-SCT *after acute lymphoblastic leukemia (ALL)-type chemotherapy can be recommended in first complete remission or relapse.*<sup>[8]</sup>

The clinical outcome of our case was fatal due to late presentation and non-availability of specific acute leukemia chemotherapeutic drugs. It is thus recommended that acute leukemia diagnosis and management be improved in developing countries such as Mali. Similarly, there is a need to develop cooperation between centers in developed countries and developing countries to improve the clinical outcome of this type of neoplasm.

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## Conflicts of interest

There are no conflicts of interest.

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