

Blastic Plasmacytoid Dendritic Cell Neoplasm in a Pediatric case – A Rare Case Report

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Abstract : Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a very rare CD4+ CD56+ myeloid malignancy that is challenging to diagnose and treat. BPDCN predominantly affects skin followed by blood/ bone marrow and later progresses to leukemia. It presents as nonspecific cutaneous lesions with or without extra-cutaneous manifestations, thereby forming a close differential diagnosis for hematodermic malignancies. We present a case of a 2.5-year-old child with blastic plasmacytoid dendritic cell neoplasm which is an extremely rare hematological malignancy with a distinct treatment protocol.

Keywords: BPDCN, blastic plasmacytoid, myeloid, neoplasm, cutaneous, dendritic cell

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I. Introduction

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is an extremely rare hematological malignancy of plasmacytoid dendritic cells. It has an incidence of less than 1 % of all hematological malignancies (1). BPDCN was formerly named blastic natural killer (NK) cell lymphoma, agranular CD4+ NK cell leukemia, blastic NK leukemia/lymphoma, and agranular CD4+ CD56+ hematodermic neoplasm/tumor (2) (3). Upon the discovery of its origin from plasmacytoid dendritic cell, it was renamed in the 2008 World Health Organization (WHO) Classification of Tumours of Haematopoietic and Lymphoid Tissues as BPDCN and classified as myeloid lineage (4). It is currently classified as a separate entity in the 2016 WHO classification of hematolymphoid malignancies (5) (6).

II. Case Report

A 2 ½ year old male child presented to the pediatric OPD, Government Rajaji Medical College and Hospital, Madurai, Tamil nadu with complaints of fever for 10 days, epistaxis and multiple bruise like skin papules.

On examination, the child was febrile and was found to have pallor. He had multiple ecchymotic bruise like skin papules over upper back, bilateral gluteal region, and groin (Fig 1,2). Abdominal examination showed hepatomegaly. Generalized lymphadenopathy and splenomegaly was absent. The peripheral smear showed pancytopenia (Fig 3) with total count of 2900 cells/mm³, RBC – 2.9 million/ µl, Hemoglobin – 5.8g/dl, PCV – 17.9%, platelets – 58,000 / µl and reticulocyte count was 0.4%. Biochemical investigations done showed a RBS – 65 mg%, serum urea, creatinine and serum electrolytes were normal. Chest X ray showed no mediastinal mass. CT and USG abdomen showed no organomegaly. ECHO was within normal limits. Bone marrow aspiration was done which showed 29% blasts. Blasts were large cells with irregular nuclei with finely dispersed chromatin and abundant blue gray cytoplasm that had microvacuoles with pseudopodia like extensions (Fig 4-6). The blood was sent for flow cytometric analysis. The flow cytometry showed blast – 15%, CD 4 – 90%, CD 56 – 81%, HLA DR – 83% aberrant CD 33 – 93%. Other lineage markers namely T-Lymphoid markers (CD2, CD3, CD5, and CD7), B-Lymphoid markers (CD10, CD19, CD20, and CD 21), myelomonocytic markers (CD11B, CD13, CD14, CD16, and CD117) and plasma cell markers (CD138, SmlgM) were absent. Thus, a diagnosis of blastic plasmacytoid dendritic cell neoplasm was made. Treatment was based on the acute lymphoblastic leukemia (ALL) protocol comprising of hyperCVAD (cyclophosphamide, vincristine, adriamycin and decadron) alternating with MTX/Ara-C (methotrexate and cytarabine). The patient achieved remission which was confirmed by a bone marrow aspiration (Fig 7).



Fig – 1,2 Ecchymotic bruise like skin papules over groin and upper back.

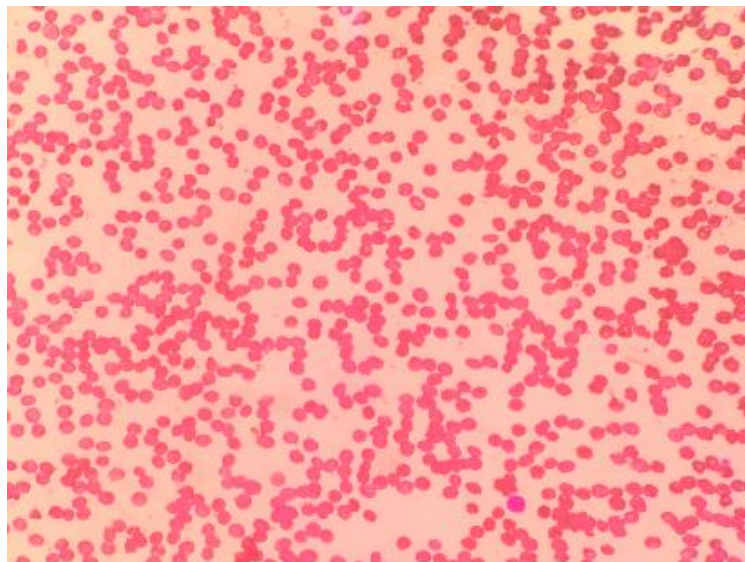


Fig 3 – peripheral smear showing pancytopenia

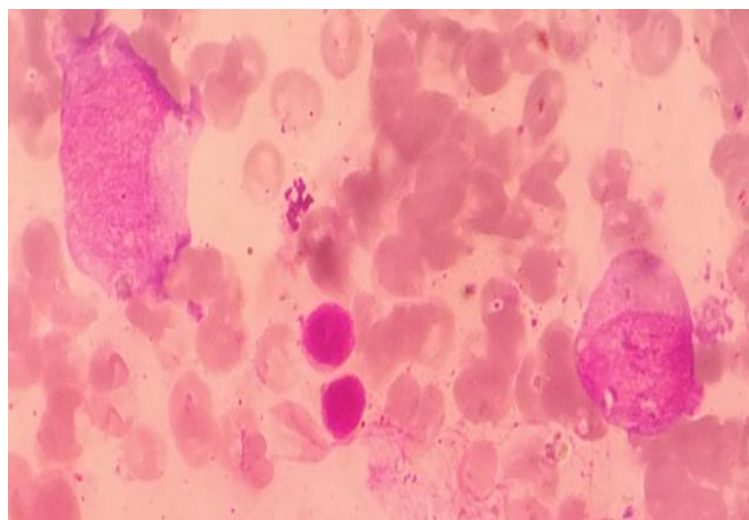


Fig 4: Bone marrow aspirate showing large agranular blasts with abundant cytoplasm

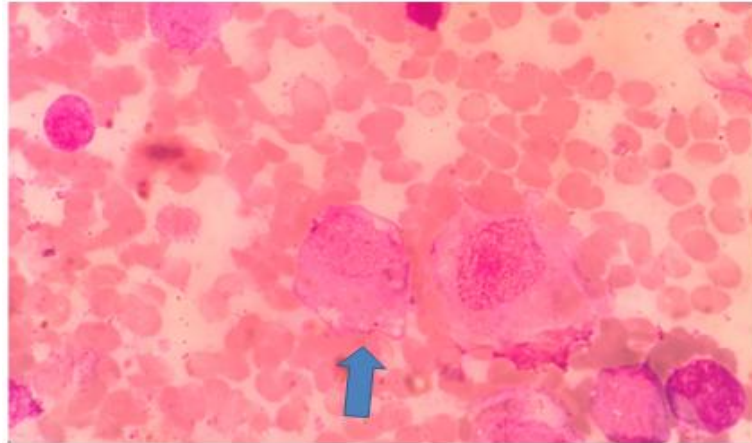


Fig. 5 –blasts with cytoplasmic microvacuoles

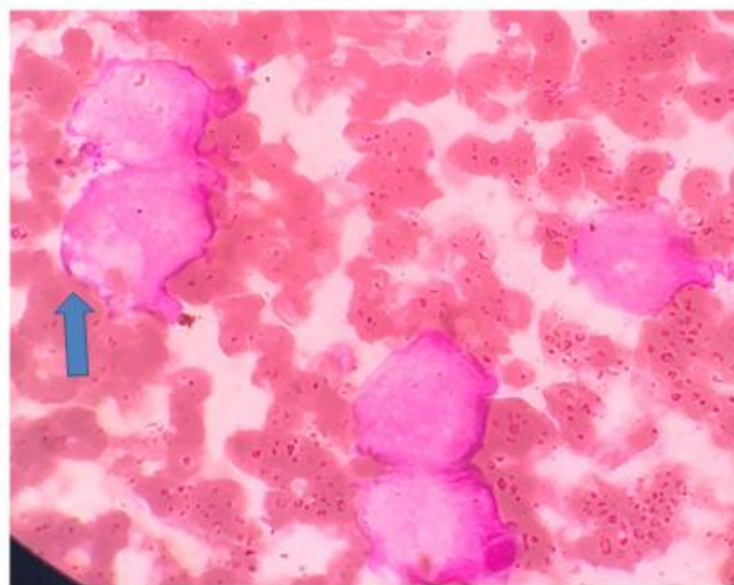


Fig 6 : Blasts with cytoplasmic pseudopodia like extensions

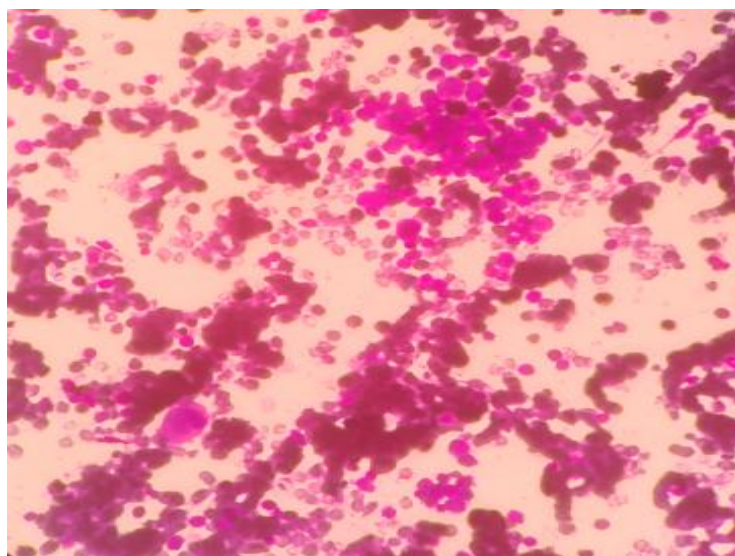


Fig 7: Bone marrow aspirate with erythroid, myeloid precursors and a megakaryocyte

III. Discussion

BPDCN is an extremely rare hematological malignancy of plasmacytoid dendritic cells producing IFN α . It has an M: F ratio – 3.3:1. Patients are mostly elderly – 61-67 years though it occurs rarely in children (7). There are currently no clues about etiology. BPCDN mostly presents with skin manifestations (64- 100% of cases). Blood and bone marrow involvement also occur (60-90% cases) (8). The peripheral blood and bone marrow aspirates show agranular blasts with fine chromatin and abundant blue gray cytoplasm with microvacuoles and pseudopodia. Diagnosis of BPCDN is based on flowcytometry/IHC with CD45+, CD4+, CD56+, HLA-DR+ immunophenotype. Other lineages – myeloid,lymphoid,myelomonocytic are negative .CD123 , CD303, TCL 1A are the recent markers that are very specific in the diagnosis of BPCDN. It also occurs in association with CMML, MDS and AML (9) (10). Differential diagnoses that must be considered are - AML with Monocytic differentiation [MPO +, CD117 +, CD 34+, CD 11+, CD 14+], Mature plasmacytoid dendritic cell proliferation (MPDCP) [CD 56 -], T / NK Extra Nodal Lymphoma [CD8 +, CD 30+/-], Cutaneous Peripheral T lymphoma [CD8+, CD2 +, CD5 +, CD7 +]. No standardized treatment protocols have been devised so far. Current treatment strategy involves the ALL protocol. Prognosis is dismal – 10 to 19.8 months with subsequent drug resistance(11).Age has an adverse effect on survival with paediatric patients faring better(12) (13).Hematopoietic stem cell transplant after the achievement of first remission is the best way for longer survival (14).

IV. Conclusion

Blastic plasmacytoid dendritic cell neoplasm is an aggressive and extremely rare hematological neoplasm. It presents a diagnostic challenge due to its varied presentation and diverse skin manifestations. A complete work up and thorough immunophenotyping of all hematodermic neoplasms are required to rule out this rare malignancy as the treatment strategies and prognosis are distinctly different.

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