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BLASTIC PLASMACYTOID DENDRITIC CELL NEOPLASM: A CASE REPORT OF A RARE LEUKEMIA

Oncology

Dr. Suresh Babu M. C	Professor and HOD, DM Medical Oncology, Kidwai Memorial Institute of Oncology
Dr Manjunath S Hiremani*	Senior Resident, DM Medical Oncology, Kidwai Memorial Institute of Oncology *Corresponding Author
Dr Lokesh K. N	Professor, DM Medical Oncology, Kidwai Memorial Institute of Oncology
Dr. Rudresha A. H	Associate Professor, DM Medical Oncology, Kidwai Memorial Institute of Oncology
Dr. L K Rajeev	Associate Professor, DM Medical Oncology, Kidwai Memorial Institute of Oncology
Dr. Smitha C Saldanha	Associate Professor, DM Medical Oncology, Kidwai Memorial Institute of Oncology
Dr. Giri G V	Associate Professor, DM Medical Oncology, Bangalore Medical College and Research Institute

ABSTRACT

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare, highly aggressive hematologic malignancy that typically affects elderly males and accounts for <1% of all hematologic cancers. It most often presents with cutaneous lesions and rapidly progresses to systemic disease with poor survival outcomes. We report a rare case in an adolescent male. A 17-year-old presented with bruise-like cutaneous patches over the chest and back, accompanied by fever and pancytopenia. Bone marrow evaluation revealed atypical blasts, and flow cytometry demonstrated positivity for CD2, CD4, CD12, CD33, CD38, CD56, CD58, CD117, CD123, and HLA-DR, confirming the diagnosis of BPDCN. Cytogenetics showed a normal male karyotype. The patient was treated with HYPERCVAD chemotherapy, achieving hematological and cytological remission after two cycles, and is subsequently planned for allogeneic bone marrow transplantation. This case highlights the diagnostic challenge and aggressive nature of BPDCN, especially in adolescents where it is exceedingly rare. Early recognition, prompt initiation of ALL-like induction chemotherapy, and consideration of consolidative allogeneic stem cell transplantation are crucial to improve outcomes.

KEYWORDS

BPDCN, HYPERCVAD, ALL-like Induction Chemotherapy, Allogenic Transplant

INRODUCTION

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare and aggressive hematologic malignancy comprising only 0.44% of all hematologic malignancy.[1] It can occur in all age groups, but predominantly seen in elderly males with median age of presentation being 67 years. It commonly presents with nonspecific cutaneous lesions, often accompanied by peripheral blood and bone marrow involvement, and can progress rapidly to systemic involvement and death within a year.[2] The diagnosis of BPDCN can be challenging due to its clinical and biological heterogeneity, as well as its overlapping features with other hematologic malignancies.[3] Here we discuss a rare case presenting rarely in AYAs.

CASE

History

A 17 year old male from Chamarajanagar, Karnataka presented with complains of skins lesions over front and back of the chest, not associated with itching, tenderness, any bleeding manifestation . No h/o any drug exposure or similar history in family.

H/o fever one week history insidious onset, intermittent, high grade with chills and predominantly nocturnal fever. Associated with headache, throbbing type, bifrontal, intermittent, non radiating type.

On evaluation outside, he was found to have pancytopenia, he received supportive care and referred to us for further management. Rest other fever profile tests done outside were negative (PS FOR MP, WIDAL, DENGUE).On presentation to KIDWAI, he still had complains of fever, skin lesions persisting. He was further evaluated here.

The patient does not have history of similar complaints in the past and does not have any personal or family history of cancer, diabetes, hypertension, cancer, or tuberculosis. The patient appeared to be moderately nourished and built upon general inspection. On examination he had pallor and bruise like patches on anterior part of chest and back as shown in Figure 1.

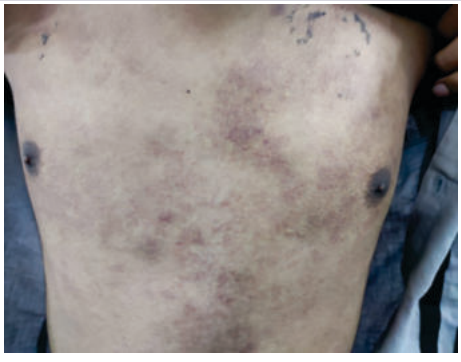

Figure 1: Multiple Ill-defined, Bruise-like, Violaceous Patches and Plaques Scattered Over the Anterior Chest Wall.

Investigations

Routine Blood Investigations :

Parameter	Report (with units)	Reference Range
Hemoglobin (Hb)	4.8 g/dL	13.0 – 17.0 g/dL (male)
Total Leukocyte Count (TLC)	1500	4,000 – 11,000 /µL
Absolute Neutrophil Count (ANC)	30	1,500 – 8,000 /µL
Absolute Lymphocyte Count (ALC)	1400	1,000 – 4,000 /µL
Platelets	96,000 /µL	150,000 – 450,000 /µL
Peripheral Smear	RBC, TLC, Platelets reduced. No atypical cells.	
Renal Function Test (RFT)	Normal	Within normal limits
Liver Function Test (LFT)	Normal	Within normal limits
Serum Sodium (Na)	138 mmol/L	135 – 145 mmol/L
Serum Potassium (K)	3.9 mmol/L	3.5 – 5.0 mmol/L

Serum Calcium	9.2	8.5 – 10.5 mg/dL
Serum LDH	220	140 – 280 U/L
Serum Uric Acid	2	3.5 – 7.2 mg/dL (male)
Vitamin B12	2000 pg/mL	200 – 900 pg/mL
Iron Profile	128 1229.12	Serum Iron: 60–170 µg/dL; Ferritin: 30–400 ng/mL (male)

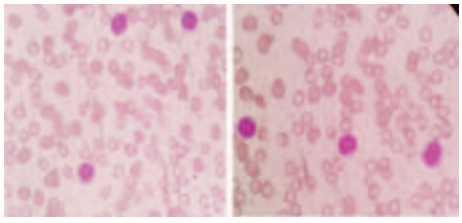

Figure 2: Bone Marrow Smear Showed Atypical Cells That Showed High Nuclear-cytoplasmic (NC) Ratio, Prominent Nucleoli, and Slightly Vacuolated Cytoplasm.

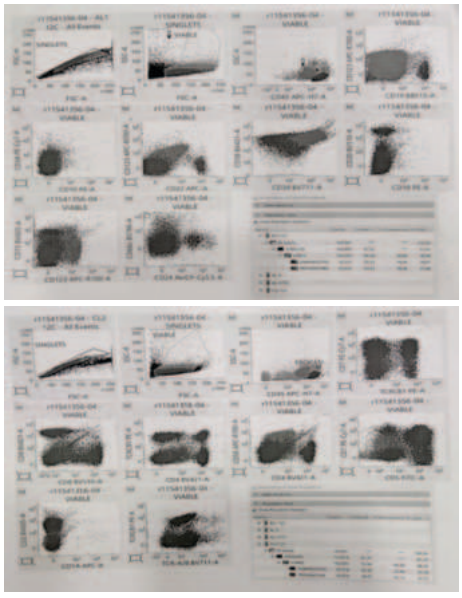

Figure 3: Flow Cytometry of Bone Marrow Showed Blasts Positive for Cluster of Differentiation (CD) 2, CD4, CD12, CD38, CD58, CD56, CD 33, CD 117 and CD123, and HLA-DR Indicative of BPCDN

On evaluation, the patient was found to have severe anemia with hemoglobin of 4.8 g/dL, thrombocytopenia with platelets of 96,000/µL, and neutropenia with an absolute neutrophil count of around 30. Biochemical parameters including renal function tests and serum electrolytes were within normal limits (sodium 138 mmol/L, potassium 3.9 mmol/L). Vitamin B12 was elevated at 2000 pg/mL, while viral markers were negative and ECG showed a normal sinus rhythm. Bone marrow examination revealed atypical blasts characterized by a high nuclear-to-cytoplasmic ratio, prominent nucleoli, and slightly vacuolated cytoplasm. Flow cytometry of the bone marrow confirmed the abnormal population of blasts, which were positive for CD2, CD4, CD12, CD33, CD38, CD56, CD58, CD117, CD123, and HLA-DR, consistent with a diagnosis of Blastic Plasmacytoid Dendritic Cell Neoplasm. Cytogenetic analysis demonstrated a normal male karyotype, and the mutation panel did not reveal any pathogenic alterations.

Treatment History

Based on the above features, he was diagnosed as a case of BPDCN and Treatment in the form of HYPERCVAD was started. After 2# of HYPERCVAD (PHASE A AND B), patient is in hematological remission and planned for Allogenic BMT.

DISCUSSION

Blastic Plasmacytoid Dendritic Cell Neoplasm (BPDCN) is an aggressive and rare hematologic malignancy originating from precursors of plasmacytoid dendritic cells. It constitutes less than 1% of all hematologic malignancies and typically affects older adults with a median age of 67 years. However, it can also occur in younger populations, as evidenced in our case involving a 17-year-old male. BPDCN's natural history is marked by its aggressive progression and poor prognosis, with a median overall survival of 12 to 16 months. The disease often rapidly advances to systemic involvement, leading to a high relapse rate within the first year after diagnosis. It was formally categorized by the World Health Organization in 2008 as a distinct entity under hematopoietic and lymphoid tumors.[4]

The clinical presentation of BPDCN commonly includes cutaneous lesions, which are often the first noticeable symptoms. These lesions can appear as bruise-like patches, nodules, or plaques, and may be accompanied by systemic symptoms such as fever, pancytopenia, and, in advanced stages, lymphadenopathy and hepatosplenomegaly. Our case reported with skin lesions on the chest and back, along with systemic symptoms. Similarly, the case by Hirata et al. (2023) initially presented with erythema nodosum-like symptoms, which were later diagnosed as BPDCN upon further biopsy.[5] Granroth et al. (2023) also documented cutaneous presentations, underscoring the importance of recognizing these early signs for timely diagnosis.[9]

Diagnosing BPDCN is notably challenging due to its rarity and the overlapping features with other hematologic malignancies. BPDCN can be confused with other malignancies such as acute myeloid leukemia (AML), leukemia cutis, and myeloid sarcoma due to similarities in clinical and pathological features. Specific immunohistochemical markers like CD123, CD4, and CD56 are crucial for distinguishing BPDCN from these conditions.[6] As it is a rare disease, many clinicians and pathologists may not be familiar with its characteristics, leading to delayed or incorrect diagnoses. Increased education and awareness efforts are necessary to improve diagnostic accuracy.

Treatment for BPDCN has traditionally involved chemotherapy regimens similar to those used for acute lymphoblastic leukemia (ALL) and acute myeloid leukemia (AML).[10] In our case we utilized the HYPERCVAD regimen, achieving hematological and cytological remission. A retrospective study by Pagano et al. showed significantly better outcomes with ALL-type regimens, both in terms of response rate and overall survival.[4] The use of ALL-type induction chemotherapy followed by allogeneic stem cell transplantation has shown promise, particularly in patients without prior recurrences. [7]

Recent advances in treatment include targeted therapies such as Tagraxofusp (SL-401), a CD123-directed cytotoxin, which has shown promising results in clinical trials.[8] Granroth et al. (2023) reported the use of venetoclax combined with hypomethylating agents, providing new therapeutic options for BPDCN patients.[9] These advancements are critical for improving patient outcomes given the aggressive nature of BPDCN and its poor prognosis with traditional chemotherapy alone.

Future research in BPDCN should focus on developing more effective targeted therapies with fewer side effects, understanding the molecular and genetic basis of the disease to identify potential biomarkers, and conducting large-scale, multi-center trials to establish standardized treatment protocols. Additionally, age-specific research is necessary, as highlighted by the young age of the patient in the our case. Addressing these research areas will be crucial for improving the prognosis and quality of life for BPDCN patients.

CONCLUSION

This case highlights the unusual presentation of Blastic Plasmacytoid Dendritic Cell Neoplasm in an adolescent, emphasizing the need for high clinical suspicion in atypical age groups. Prompt diagnosis through immunophenotyping and early initiation of ALL-like chemotherapy with consolidation by allogeneic stem cell transplantation remain the best strategies for durable remission in this aggressive disease.

Future research should focus on developing effective targeted therapies with fewer toxicities, exploring and establishing standardized treatment protocols through large multicenter studies. Moreover, studies addressing the molecular and genetic basis of BPDCN, as well as age-specific research in younger patients, are essential to improve both survival outcomes and quality of life.

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