A CASE OF BLASTIC PLASMACYTOID DENDRITIC CELL NEOPLASM WITH UNUSUAL PRESENTATION

Running title: A case of BPDCN.

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Blastic Plasmacytoid Dendritic Cell Neoplasm (BPDCN) was categorized in 2008 World Health Organization (WHO) classification of hematological diseases under acute myeloid leukemia and related precursor neoplasms1, however in the revised edition 2017 it is a separate entity2. BPDCN predominantly affects elderly patients with a mean age between sixth and seventh decade with a male predilection (M: F=3:1); however they can present even in children3,4. It has an aggressive behavior and rapid systemic dissemination. We herein report a case of BPDCN in an adolescent patient with unusual presentation.

A 13-year-old male presented with fever and weakness for 10 days. The patient had a history of left testicular swelling for which he underwent orchidectomy two months back. The routine hematological investigations revealed: Hemoglobin (Hb) - 9.1 g/dl, Total leucocyte count (TLC) - 5 x 10^3/μL and Platelet (PLT) - 80 x 10^3/μL and 18% blasts on differential count. Bone marrow aspirate showed 90% blasts with lymphoid morphology (Figure 1A). Flow cytometric immunophenotyping (FCM) analysis of bone marrow
revealed 80% blasts with CD45dim and low side scatter. The cells were also positive for CD4,CD56,CD38,CD123,CD7 and negative for CD19,CD20,CD22,CD3,CD8,CD1a,CD34,CD36 TdT, CD61,CD235a,CD13,CD33,CD14,CD64,CD36,CD117,cMPO,cCD79A and cCD3 (Figure 1J,K,L,M,N,O,P). Bone marrow biopsy revealed near total replacement by monomorphic cells which on Immunohistochemistry(IHC), were positive for CD4, CD43 and CD7 and negative for CD3 and CD68 (Figure1B, C,D,E,F). The paraffin blocks of the testicular mass were reviewed and revealed a tumor comprising of small monomorphic round cells with scanty cytoplasm, round to focally indented nuclei with prominent nuclear membrane and inconspicuous nucleoli. Few mitotic figures were seen. The seminiferous tubules were enclosed and were being indented by the tumor throughout the tissue section. These cells were positive immunohistochemically for CD4, CD43, CD56, CD123 and negative for CD3, CD68 and CD8 (Figure 1G, H, I). Based on histopathological findings, (IHC) and FCM analysis the patient was diagnosed with “BPDCN”.

BPDCN closely resembles precursor of plasmacytoid dendritic cell by immunophenotyping, gene expression profiling and biologic function. Skin manifestations are the main clinical presentation of typical cases of BPDCN seen in 64%-100% cases of adults and the diagnosis is made on skin biopsy. Extracutaneous involvement often occurs in the lymph nodes, peripheral blood, bone marrow, skeleton, gastrointestinal tract, CNS, lung, mediastinum and pancreas. Our patient, a young male presented with testicular involvement and without any cutaneous manifestation which is extremely rare. 2 months later the patient presented with leukemia like symptoms with bone marrow involvement.

FCM is an useful tool and the cells express CD4,CD43,CD45RA,CD56,CD123,CD303,CD304, TCL1,CLA, CD38,CD99, and CD31. Recently the TCF4(E2-2)transcription factor represents a faithful diagnostic marker. The immunonegativity of immature cells for CD14,CD15,CD36,CD64 and CD68 excludes possibility of acute myeloid leukemia with monocytic differentiation. Cytogenetic studies may show genomic abnormalities or a normal karyotype.

From a pathologist perspective the immunophenotyping profile should be kept in mind before considering a case of BPDCN as an Acute undifferentiated leukemia or Acute myeloid leukemia with monocytic differentiation.

References:


