Pseudo-Rosette-Forming Blastic Plasmacytoid Dendritic Cell Neoplasm

Psödorozet Oluşturan Blastik Plazmasitoid Dendritik Hücre Neoplazmı

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To the Editor,

We read the recent article on blastic plasmacytoid dendritic cell neoplasm (BPDCN) [1] with interest and would like to add another interesting case to the list with an intriguing finding in the bone marrow. A 64-year-old male patient presented with complaints of fever and easy fatigability. Moderate pallor and hepatosplenomegaly were present. Laboratory investigations showed pancytopenia with hemoglobin of 72 g/L, total leukocyte count of 0.9×10^9/L, and platelet count of 45×10^9/L. Bone marrow examination revealed 92% medium-sized blastoid cells with fine chromatin, inconspicuous nucleoli, and agranular cytoplasm with pseudopodia (Figure 1A). Myeloperoxidase and periodic acid-Schiff staining was negative. Bone marrow biopsy showed sheets of tumor cells forming peculiar pseudo-rosette formations at places (Figure 1B). Immunophenotyping identified a dim CD45-positive blast population, which was also positive for CD4, CD56, CD123, CD33, CD38, and HLA-DR. The blasts were negative for CD34, B-cell markers, T-cell markers (surface and cytoplasmic CD3, CD4, CD8, CD5, and CD7), myeloid markers (CD13, CD117, and myeloperoxidase), monocytic markers (CD14, CD64, and CD36), and markers of immaturity (TdT, CD34) (Figures 1C-1F). The patient was diagnosed with blastic plasmacytoid dendritic cell neoplasm (BPDCN). His general condition deteriorated and he died due to progressive illness.

BPDCN is a rare and aggressive hematological malignancy associated with a high frequency of skin and/or bone marrow infiltration and leukemic dissemination [2]. Absence of lineage-specific markers in association with high levels of expression of plasmacytoid dendritic cell markers such as CD123, CD4, CD56, and CD45RA is pathognomonic of BPDCN [2]. Rosette formation by hematopoietic neoplasms is known [3,4]; however, this is a unique case of pseudo-rosette formation by BPDCN in bone marrow, emphasizing that hematopathologists should consider BPDCN in the differential diagnosis of pseudo-rosette-forming tumors in the bone marrow.

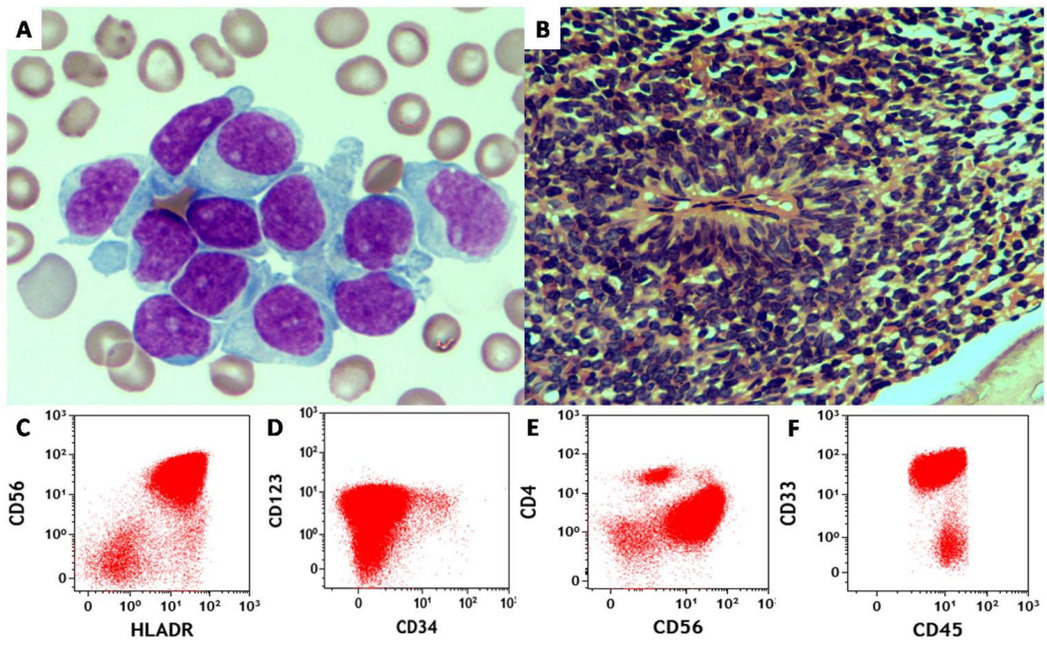

Figure 1. A) Bone marrow aspirate smear showing blastoid cells (Giemsa stain, 1000x). B) Bone marrow biopsy showing pseudo-rosette formation by tumor cells (hematoxylin and eosin stain, 400x). C-F) Flow cytometric dot plots showing characteristic surface immunophenotypic marker expression profile of blastic plasmacytoid plasma cell neoplasm.

Keywords: Blastic plasmacytoid dendritic cell neoplasm, Pseudo-rosette, Bone marrow

Anahtar Sözcükler: Blastik plazmositoid dendritik hücre neoplazmı, Pseudorosette, Kemik iliği

Informed Consent: Informed consent was obtained from the patient included in the study.

Authorship Contributions

Concept: S.N.; Design: S.N.; Data Collection or Processing: P.S.; Analysis or Interpretation: S.N.; Literature Search: P.S.; Writing: P.S.

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Reply

To the Editor,

We have read the letter "Pseudo-Rosette-Forming Blastic Plasmacytoid Dendritic Cell Neoplasm," which presents a very fine example of blastic plasmacytoid dendritic cell neoplasia (BPDCN).

There is one published case in the literature of CD4+ and CD56+ blastic NK cell lymphoma with pseudo-rosettes [1]. Although this case was diagnosed with a name other than BPDCN, the clinical, morphological, and immunophenotypic features are very similar to BPDCN, except for TdT positivity. BPDCN is a relatively new name and blastic NK cell lymphoma was one of BPDCN's older names before BPDCN was defined and accepted as a new and separate entity [2].

Pseudo-rosette formation is a very well-known infiltration pattern for many lymphomas, especially central nervous system lymphomas, T/NK cell lymphomas, etc. However, except for this particular case, even in larger case series, pseudo-rosettes are not defined as a common or a diagnostic pattern for BPDCN [3,4]. We did not see pseudo-rosettes in our small case series of BPDCN [5]. The patterns of the tumors in our series were mostly diffuse in the dermis in skin biopsies and interstitial in bone marrow biopsies, as expected.

Since we do not have enough information about this very rare tumor, diagnostic and treatment options are very limited and controversial. Therefore, it is very valuable to learn any features of this rare entity.

Best Regards,

Ahu Senem Demiröz, Cuyan Demirkesen, Ayşe Salihoğlu, Nükhet Tüzüner

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