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Pediatric Case of Blastic Plasmacytoid Dendritic Cell Neoplasm: A Rare Entity

Blastik Plazmasitoid Dendritik Hücreli Neoplazi Tanısı Alan Pediyatrik Bir Hasta: Nadir bir Durum

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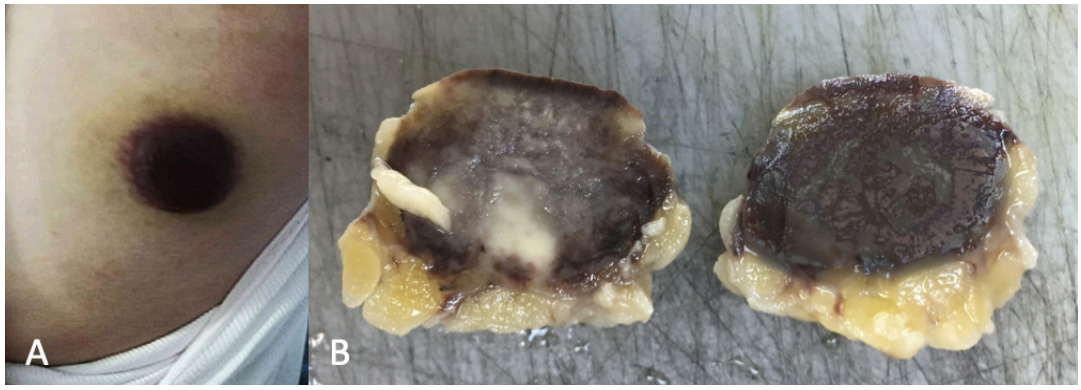

Figure 1. A) Indurated purplish-brown nodular lesion between left breast and axilla. **B)** Grossly, the lesion measured 2.5x2.1x2.1 cm, was soft and entirely solid, and had ill-defined borders and a gray to brown smooth cut surface with glossy appearance.

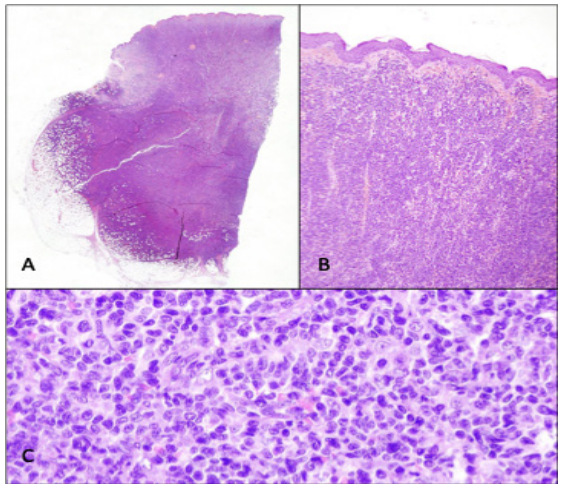

Figure 2. A) Involvement of the dermis and the subcutaneous fat. **B)** Abundant erythrocyte extravasation among neoplastic cells and in the grenz zone, concordant with bruise-like clinical appearance in the early phase and purplish appearance of the late nodule (H&E, 10x (A), 40x (B)). **C)** Irregular nuclear contours, pleomorphic nuclei, finely dispersed chromatin, indistinct nucleoli, and scant amount of cytoplasm (H&E, 400x).

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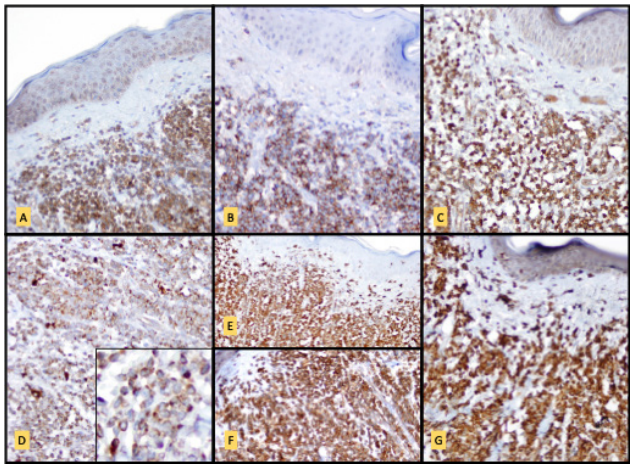

Figure 3. Positive staining with CD4 **(A)**, CD56 **(B)**, CD123 **(C)**, CD68 **(D)**, S100 **(E)**, CD33 **(F)**, and Bcl2 **(G)**. Staining for CD68 was in a dot-like pattern (D, inner photo) (immunohistochemistry, 200x (A, B, C, D, G), 100x (E, F), 400x (D, inner photo).

An 11-year-old female patient was referred with a 4-month history of skin lesion between the left breast and axilla. Initially, the lesion had appeared as a violaceous bruise-like macule, which enlarged progressively, became elevated, and then turned into a purplish-brown nodule (Figure 1). Complete blood count, blood chemistry, and coagulation test results were unremarkable. No B symptoms were noticed. The patient had mild hepatomegaly but no lymphadenopathy or splenomegaly. Ultrasonographic findings of the tumor were compatible with a benign angiomatous lesion. Histopathological examination of the excisional biopsy demonstrated dermal non-epidermotropic, monomorphous, diffuse infiltrate of medium-sized cells reminiscent of lymphoblasts or myeloblasts (Figure 2). Immunohistochemically, neoplastic cells were diffusely positive for CD4, CD56, CD123, and Bcl-2 while most lineage-specific markers, including myeloid (CD117, MPO), B-cell (CD10, CD20, CD79a), and T-cell (CD3, CD5) antigens, were negative, as were CD34, TdT, CD30, CD99, cytotoxic markers (TIA-1, lysozyme), and in situ hybridization results for Epstein-Barr virus (EBER). The majority of the cells were positive for CD45RA, CD68 (dot-like), CD33, and S100 (Figure 3). Following the diagnosis of blastic plasmacytoid dendritic cell neoplasm, systemic investigations including bone marrow biopsy and central nervous system imaging were performed. The patient, with skin-limited disease, received chemotherapy according to the Berlin-Frankfurt-Münster high-risk acute lymphoblastic leukemia protocol. After induction chemotherapy, she achieved complete remission. Unfortunately, she died in the third month of treatment due to refractory *Pseudomonas* sepsis despite all supportive treatments.

Keywords: Pediatric leukemia, Acute leukemia, Other leukemias, Dendritic cell

Anahtar Sözcükler: Pediyatrik lösemi, Akut lösemi, Diğer lösemiler, Dendritik hücre

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