

Blastic Plasmacytoid Dendritic Cell Neoplasm: Report of a Case

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Abstract

Blastic plasmacytoid dendritic cell neoplasm (BPDCN), also known as CD4+/CD56+ hematodermic neoplasm, is a rare, highly aggressive hematopoietic malignancy characterized by cutaneous, lymph node and bone marrow involvement, high risk of a leukemic dissemination, and a poor prognosis. It has been recognized as an independent entity in the World Health Organization (WHO) 2008 classification for cutaneous lymphomas and is thought to be derived from the precursors of plasmacytoid dendritic cells. This neoplasm most commonly affects middle-aged or elderly patients with predominant skin or soft tissue involvement. We report a case of a 87-year-old man with a purple-red plaque on his face and multiple purple-red nodules on his trunk. Histopathology and immunohistological staining confirmed the diagnosis of blastic plasmacytoid dendritic cell neoplasm.

Keywords: Blastic plasmacytoid dendritic cell neoplasm; CD4+/CD56+ hematodermic neoplasm; Dermopathology; Immunohistochemistry

Case Report

A 87-year-old man presented with a 6-month history of multiple, violaceous nodules and plaques on the skin of his face and trunk. Initially, the patient presented with a purple-red plaque on his face with a gradual increase in size (Figure 1). Upon physical examination, there was no lymphadenopathy or hepatosplenomegaly. Laboratory tests revealed that complete blood count, blood chemistry and urine analysis were normal. Anti-HIV, tuberculin skin (purified protein derivative), rapid plasma regain (RPR) and *Treponema pallidum* particle agglutination (TPPA) tests were negative. Furthermore, serology for Epstein-Barr virus showed no signs of an ongoing infection.

Histological examination of nodules (from face and trunk) demonstrated an intense hemato-lymphoid infiltration in the dermis and in the subcutaneous tissue (Figure 2). The phenotype of the cells was CD4+, CD 45+, CD 56+ (Figure 3a), Bcl-2 (Figure 3b), CD3-, CD8-, CD5-, CD45RO-, CD20-, CD30-, CD 21-. A strong positiveness for ki67 (MIB-1) was showed (Figure 4). A diagnosis of precursor CD56 hemato-lymphoid neoplasia was made.

A bone marrow biopsy showed a diffuse atypical infiltration that consisted of polymorphic, intermediate-size cells, which sometimes had a small nucleolus (Figure 5a). Stains were positive for CD4 and CD56 (Figure 5b). These cells were negative for CD2, CD3, CD5, CD10, CD20, CD30, and CD68. *In situ* hybridisation (ISH) for Epstein-Barr virus was negative and the diagnosis was blastic NK cell lymphoma. The patient was treated with CHOP regimen (cyclophosphamide, doxorubicin, vincristine, prednisone). This treatment regimen achieved partial remission but the patient died six months after the diagnosis.

Discussion

A CD4+/CD56+ hematodermic neoplasm (plasmacytoid dendritic cell neoplasm), formerly designated primary cutaneous natural killer (NK)/T cell lymphoma, is an aggressive and rare hematologic neoplasm recently recognized by the WHO-EORTC classification consensus for cutaneous lymphomas [1]. The neoplasm tends to affect elderly patients [2], but the disease can occur at any age, including during childhood and infancy [3]. Adachi et al. [4] reported the first case of plasmacytoid dendritic cell neoplasm; since then, about 150 cases have been described [5-8]. All patients presented dermal



Figure 1: Multiple, violaceous nodules and plaques on the skin of face and trunk.

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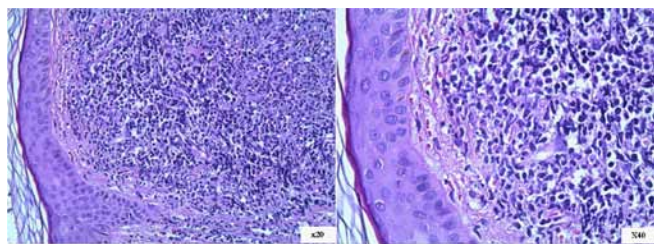


Figure 2: Diffuse monomorphic infiltration in the dermis (H&E, x20). Medium-sized blastic cells with irregular nuclei, fine chromatin, one or several small nucleoli and scant cytoplasm (H&E, x40).

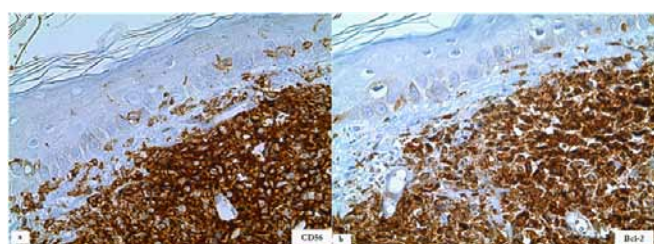


Figure 3: The neoplastic cells show immunoreactivity for CD56 (a) and Bcl-2 (b). (x200).

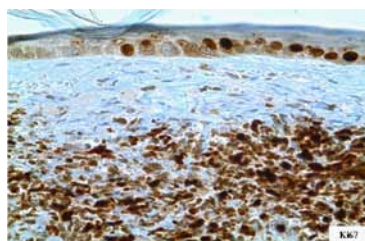


Figure 4: The immunohistochemical stains showed a strong positivity for ki67 (MIB-1).

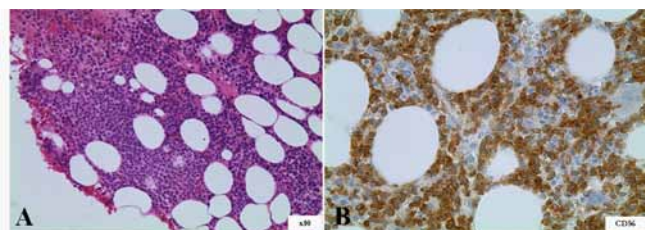


Figure 5: A bone marrow biopsy showed a diffuse atypical infiltration that consisted of polymorphic, intermediate-size cells, which sometimes had a small nucleolus (H/E x20) (a). Stains were positive for CD56 (b).

lesions involving the scalp, face, trunk, arms and legs with or without concurrent extracutaneous disease with a mean age of 55.3 years and a

male/female ratio of 3.05:1 [6]; moreover the bone marrow is involved in 65.2% of patients [6]. In the WHO-EORTC classification, the disease was named CD4+/CD56+ hematodermic neoplasm [1]. The clinical course of BPDCN is aggressive and generally dire at best. Although there is no consensus on the optimal treatment for this neoplasm, a systemic treatment should be started at the outset regardless of whether the disease is localized or disseminated.

The clinical course in most cases has been characterized by an initial response to chemotherapy followed by relapse and subsequent death. As the outcomes of many therapeutic attempts have been reported to be quite variable, there is no standard treatment for BPDCN. Usually the duration of a response to topical steroid, single agent chemotherapy, radiotherapy and polychemotherapy like CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone) or HyperCVAD (cyclophosphamide, vincristine, doxorubicin, dexamethasone alternating with high dose methotrexate and high dose cytarabine) has been reported [7-9].

In the beginning, response to chemotherapy is good; unfortunately relapses and drug resistance frequently occur [9]. The prognosis of BPDCN is poor, with a median survival period of 14 months, and 2- and 5-year overall survival rates of 33% and 6%, respectively [10]. In our case, the patient relapsed quickly and died eight months after despite aggressive chemotherapy.

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