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CASE REPORT

Cutaneous blastic plasmacytoid dendritic cell neoplasm: Report of a case and review of the literature

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ABSTRACT

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare and aggressive hematologic malignancy that usually occurs in elderly individuals and manifests with skin lesions followed by involvement of the lymph nodes, bone marrow, and peripheral blood. We report our experience of establishing the diagnosis of a case of BPDCN in an 86-year-old man. The related literature is reviewed, and the differential diagnosis and treatment modality of this rare entity are discussed.

Introduction

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare hematologic malignancy that originates from the precursors of plasmacytoid dendritic cells. It was originally called a CD4/CD56 hematodermic neoplasm[1] and subsequently regarded as a blastic variant of NK/T cell lymphoma, owing to the expression of CD56, in the third edition of the "WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues".[2] In the 2008 edition of the WHO classification, this entity was renamed as BPDCN and listed under the category of acute myeloid leukemia (AML) and related precursor neoplasms because it was found to be a neoplasm derived from the precursors of plasmacytoid dendritic cells.[3] It is an aggressive hematologic malignancy that occurs mainly in elderly individuals,[4] but childhood cases have been reported.[5] Our experience of the diagnosis of a cutaneous case of BPDCN is reported, and the current literatures is reviewed.

Case Report

An 86-year-old man with two asymptomatic nodular plaques on his scalp was referred to our hospital under a diagnosis of extranodal NK/T cell lymphoma because of its CD56 immunopositivity. Upon examination, the patient had two erythematous and violaceous nodular plaques measuring 5 cm on his parietal and temple scalp that had been present for 5 months and 2 months, respectively (Figure 1A). Another 1.5-cm nodule was found on his back (Figure 1B). The nodules were not ulcerated. The tumor on the parietal scalp was rebiopsied. Microscopically, the tumor showed diffuse infiltrates of monomorphous, medium-sized, blast-like cells with round to oval nuclei, small nucleoli, and limited cytoplasm throughout the dermis extending into the subcutis (Figures 2A and 2B). No epidermotropism was observed, and a Grenz zone was present below the epidermis. Pilosebaceous structures were not involved. Brisk mitosis was observed, and there was hemorrhage in the superficial dermis, but no necrosis or angioinvasion was found.

Immunohistochemically, the tumor cells were positive for CD4, CD33, CD56, CD68 (KP1), CD123, and TCL-1 (Figure 2), and negative for CD3, CD20, CD10, Bcl-6, CD30, ALK, myeloperoxidase, lysozyme, and granzyme B (Figure 2). The immunophenotype of CD4, CD56, CD123, and TCL-1 with negative granzyme B and myeloperoxidase established the diagnosis of BPDCN. EBER *in situ* hybridization was not performed because the histopathological and immunohistochemical studies did not support a diagnosis of extranodal NK/T cell lymphoma. A whole-body computed tomographic study revealed no hepatosplenomegaly, but mild swelling of the submandibular, neck, and axillary lymph nodes was noted, and a core needle biopsy of the submandibular lymph nodes showed similar blast-like cells expressing CD4, CD56, and CD123. Laboratory tests showed a white

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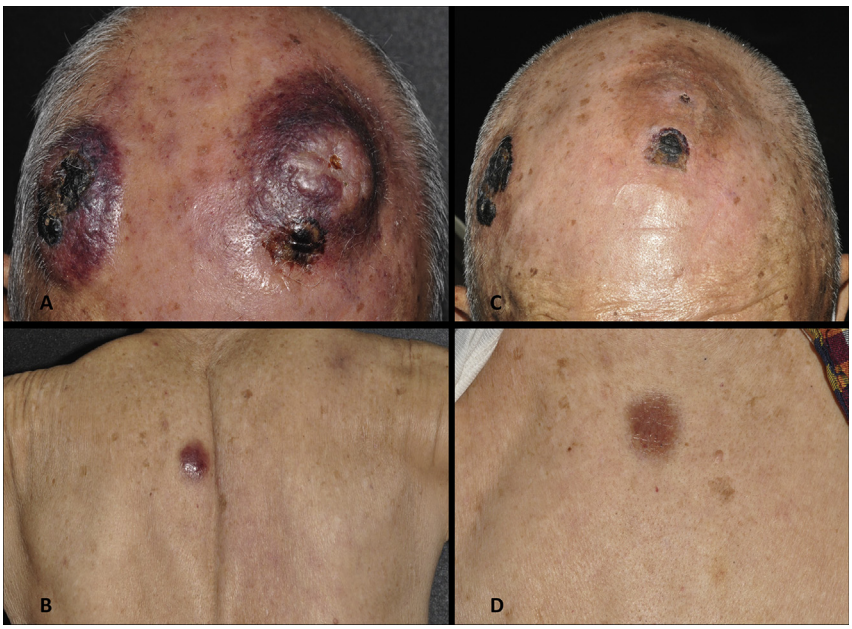

Figure 1 The clinical appearances of two tumor nodules with erythematous and dark hemorrhagic surface (A) on the scalp and (B) another smaller nodule on the back. (C, D) The tumors regressed after 2 months of oral prednisolone treatment.

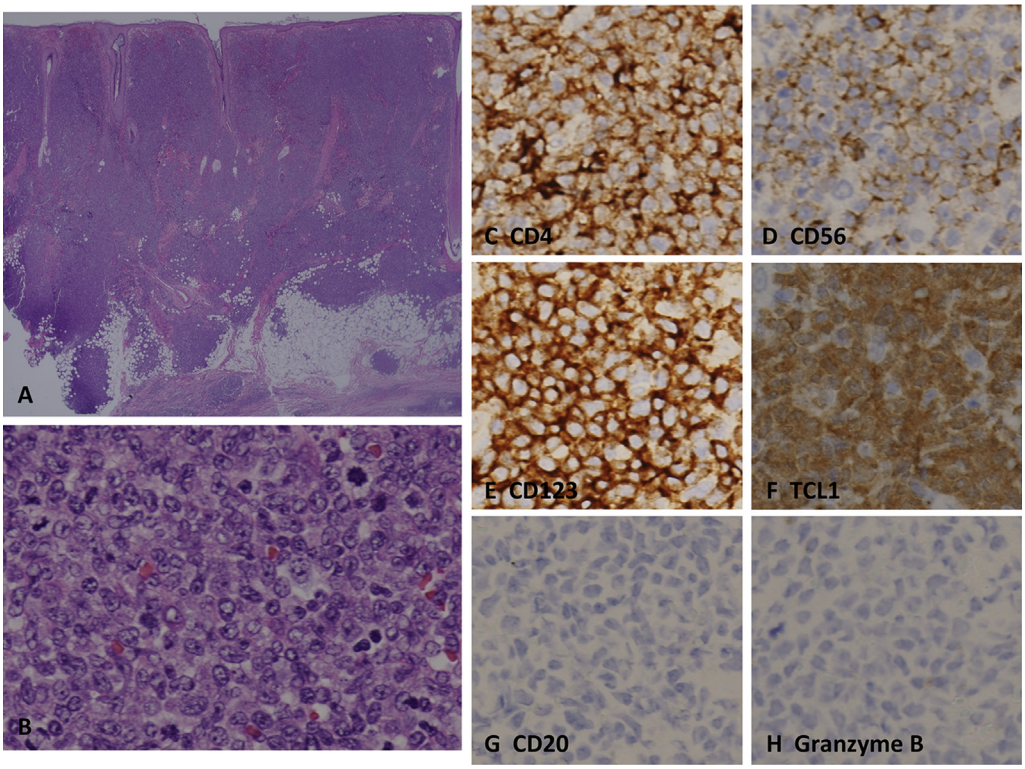

Figure 2 (A) A scanning photomicrograph of the skin tumor shows diffuse dermal and subcutaneous monotonous infiltrates with a Grenz zone (hematoxylin and eosin stain, 20×). (B) Higher magnification of the infiltrates shows medium-sized blast-like cells with brisk mitosis (hematoxylin and eosin stain, 400×). The tumor cells are positive for (C) CD4, (D) CD56, (E) CD123, and (F) TCL1, but negative for (G) CD20 and (H) granzyme B (400×).

blood cell count of 3.9×10^9/L with normal ranges of the differential count. No blast cells were observed. The hemoglobin level was 12.1 g/dL, the platelet count was 135×10^9/L, the alanine aminotransferase level was 11 U/L, and the lactic acid dehydrogenase level was 195 U/L. We did not perform chromosomal analysis or a T-cell receptor gene rearrangement study, and the patient refused a bone marrow biopsy. Because of his advanced age, the patient was treated with palliative oral prednisolone (30 mg/d). We have followed the patient for 6 months, and the tumors lave been progressively regressing (Figures 1C and 1D).

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