Blastic Plasmacytoid Dendritic Cell Neoplasm

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Introduction

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare neoplasm of plasmacytoid dendritic cells (PDC) which very often affects the skin and shows a leukemic phase in the majority of the patients.

Clinical Features

In the skin, BPDCN presents with nodules or contusiform, bruise-like disseminated patches mostly on the trunk and head (Fig. 70.1). Oral mucosal involvement is commonly found. At the time of diagnosis or shortly after, leukemic spread with circulating malignant cells and bone marrow involvement occurs in 70 % of the patients. The central nervous system may become involved, whereas spread to lymph nodes is rare. In some patients, BPDCN develops in the context of preceding myelodysplastic syndrome.

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Pathology

In fully developed skin lesions, there is a dense dermal monomorphous infiltrate of blasts. The infiltrate is separated from the epidermis by a grenz zone (Fig. 70.2). The blastic tumor cells display nuclei with fine-dispersed chromatin and a sparse cytoplasm (Fig. 70.3). Extravasated erythrocytes are a characteristic finding which leads to the contusiform clinical aspect of the skin lesions. In early lesions, only subtle perivascular infiltrates composed of smaller blasts, lymphocytes, and histiocytes may be present (Fig. 70.4).

The tumor cells display a characteristic phenotype with expression of CD4, CD56, CD123, CD303, and TCL-1. Apart from CD123, expression of additional PDC markers such as BDCA-2, BCL11a, and CD2AP can be demonstrated. There is variable expression of TdT. Incomplete phenotypes with loss of markers have been observed. There is no reactivity for B- and T-cell markers except for very rare cases expressing CD3. T-cell receptor genes and immunoglobulin genes are not rearranged. The 9p21.3 (CDKN2A/CDKN2B), 13q13.1-q14.3 (RB1), 12p13.2-p13.1 (CDKN1B), 13q11-q12 (LATS2), and 7p12.2 (IKZF1) regions are commonly deleted in BPDCN.

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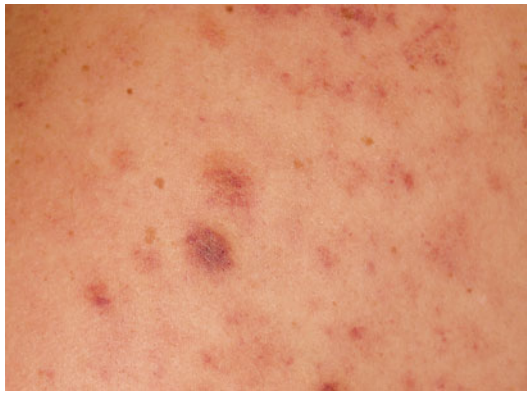

Fig. 70.1 BPDCN: Clinical manifestation with contusiform macules on the trunk

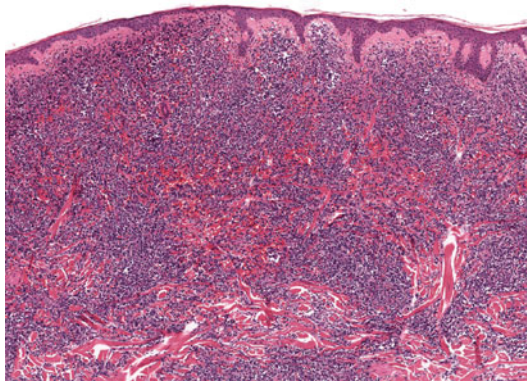

Fig. 70.2 BPDCN: Dense dermal infiltrates of blasts

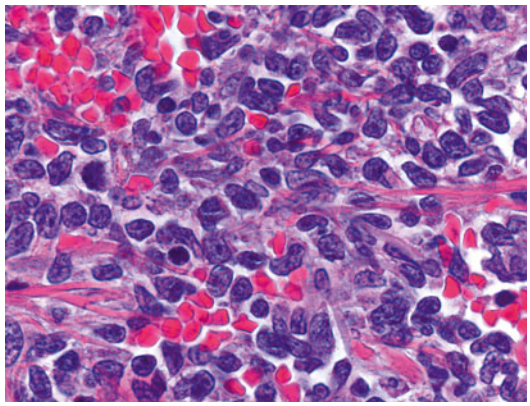

Fig. 70.3 Blasts representing neoplastic Plasmacytoid dendritic cells. Note the extravasated erythrocytes

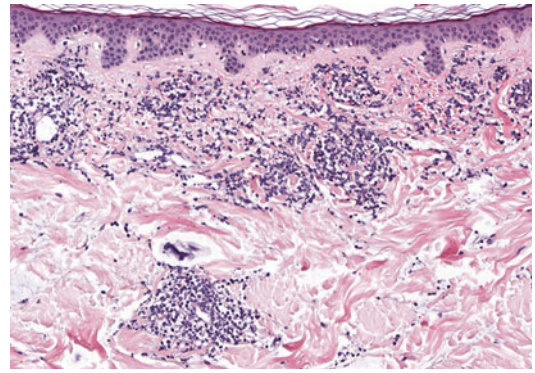

Fig. 70.4 BPDCN: Superficial perivascular infiltrates of blasts, lymphocytes, and histiocytes in a patient with leukemic spread

Differential Diagnosis

Acute and chronic myeloid and myelomonocytic leukemia (AML/AMML/CML) show overlapping clinical, histological, and phenotypic features, but TCL-1 is not expressed by tumor cells in AML/AMML, and extravasated erythrocytes are not a typical finding of specific skin infiltrates of AML/AMML. Large B-cell lymphomas and neuroendocrine carcinomas can be distinguished by their phenotypic profile. Reactive accumulations of PDC may occur in the context of AML and may mimic BPDCN.

Prognosis

BPDCN exhibits a poor prognosis with an aggressive course and a median survival of 12 months to 2 years despite initial response to treatment. CD303 expression and high proliferative index (Ki-67) were significantly associated with longer survival. Age over 40 years and biallelic loss of locus 9p21.3 indicate a shorter survival.

Treatment

Multiagent chemotherapy and allogeneic bone marrow transplantation are the first-line treatment and may in some patients result in long-term survival.

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