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Title

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Journal

Dermatology Online Journal, 25(2)

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Publication Date

2019

DOI

10.5070/D3252042892

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Peer reviewed

Blastic plasmacytoid dendritic cell neoplasm: an early presentation

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Abstract

A blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a cutaneous lymphoma derived from a plasmacytoid dendritic precursor cell that exhibits aggressive clinical behavior. Herein, we report a 46-year-old woman with a complaint of a painless nodule on the back, associated with pruritus. The nodule grew and new growths appeared over six months of evolution. The histopathological examination of one of the left upper limb lesions showed a dense lymphoid cell infiltrate with atypia in the superficial and deep dermis. Immunohistochemistry showed positivity for CD45, S-100 protein, CD123, and TCL 1. About two months after the initial evaluation, the patient was admitted to the Emergency Hospital of Marituba-PA with dyspnea. She progressed to cardiorespiratory arrest and death within 12 hours of admission. There is still no consensus for the treatment of BPDCN. Intensive therapy for acute leukemia can be useful, but allogeneic bone marrow transplantation has a greater chance of long-term survival.

Keywords: blastic plasmacytoid dendritic cell neoplasm, cutaneous nodule

Introduction

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) makes up approximately 0.7% of all primary cutaneous lymphomas, with overall incidence of 0.04 cases per 100,000 [1, 2]. This entity has a hematopoietic origin and is derived from the plasmacytoid dendritic precursor cell. Aggressive

clinical behavior is generally observed. The skin is the most affected organ (90% of cases), [3], but bone marrow, lymph nodes, and spleen can also be affected [1, 3, 4]. It is more typically manifested in elderly people (mean age 65 years old), but rare in children, having only 36 cases reported [5, 6].

Clinically, it presents with disseminated erythematous and violaceous nodules of the trunk and extremities [6, 7]. The diagnosis is made by visualizing lymphocytic atypia in the histopathological examination of the skin and the diagnosis is confirmed with immunohistochemistry demonstrating positivity of CD123 and TCL-1; usually the diagnosis is delayed and there are no uniform treatment guidelines [3, 5].

We report the clinical and morphological aspects of a patient with BPDCN diagnosed by the dermatology service of the State University of Pará. We emphasize the importance of early diagnosis in a rare disease with an aggressive clinical behavior.

Case Synopsis

A 46-year-old woman, IV phototype, presented to the dermatology department with a complaint of a painless nodule on the back, associated with pruritus. Over the course of 6 months, the nodule had grown and new lesions appeared disseminated throughout the body.

At first, the patient had presented to her physician in a good general state, without focal or systemic lymph node involvement. She denied fever or other



Figure 1. Erythematous and violaceous tumor of stone consistency, presenting irregular surface, approximately 10cm in the largest diameter and multiple small nodules, of hardened consistency, all painless.

systemic symptoms. After one month from the beginning of the clinical presentation, a new eruption of reddish nodules occurred. These were painless and located on, the upper limbs, abdomen, and lower limbs, along with an increase in size of the initial nodule.

At dermatological examination, a painless, erythematous and violaceous, infiltrated 10×10cm tumor of stony consistency on the back (**Figure 1**) was observed. In addition, multiple smaller nodules measuring on average 2×3cm were located on the upper limbs, trunk, abdomen, and lower limbs.

The patient had mild anemia (Hb=10) without other laboratory alterations. The histopathological examination of one of the left upper limb nodules showed a dense lymphoid cell infiltrate with atypia in the superficial and deep dermis (**Figure 2**). Immunohistochemistry showed positivity for CD45, S-100 protein, CD123, and TCL 1. The set of findings allowed the diagnosis of BPDCN. The patient did not comply with follow up recommendations.

After two months from the initial dermatological evaluation, the patient was admitted to the Emergency Hospital of Marituba-PA with dyspnea. She progressed to cardiorespiratory arrest and death within 12 hours of admission.

Case Discussion

The first report of BPDCN demonstrated the expression of CD56+ and CD4+ in tumor cells by immunohistochemistry. It was designated blastic NK lymphoma, owing to the probable origin of NK cells expressing CD56+ antigen. However, the positivity of CD4 and CD56 are not specific markers for NK cells. Therefore, a French Study Group on Cutaneous Lymphomas preferred the term, agranular CD4+/CD56+ hematodermal neoplasm. However, later it was demonstrated by flow cytometry that tumor cells also express CD123 and TCL-1, specific markers for plasmacytoid dendritic cells. Since then, these cells have been considered as the precursor cell of this neoplasm [2, 8, 9]. In the present report positivity for CD123 and TCL-1 confirms the diagnosis of NBCDP.

This condition is a rare and aggressive neoplasm, typically occurring in elderly patients with a mean age between 60 and 70 years old. However, it can occur in any age group. It affects more men than women, in a ratio of 3:1 [3]. Our patient does not match the common demographic findings, since she is female and has an age of less than 60 years.

Cutaneous lesions are present in all patients, being the initial clinical manifestation in 94% of cases and the only clinical manifestation in 57% of patients [3, 10].

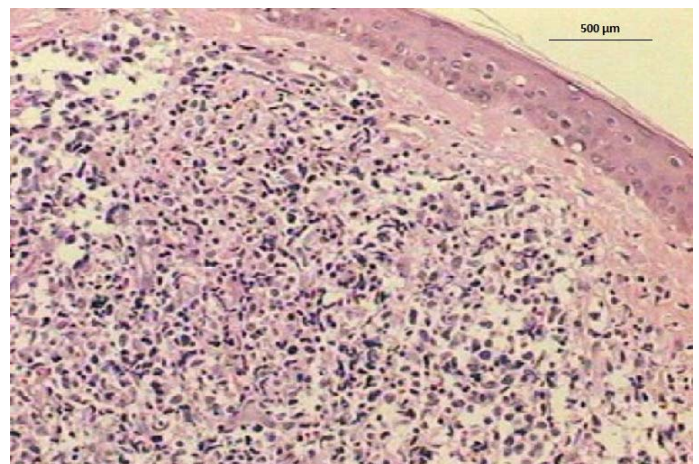


Figure 2. Infiltration by malignant neoplasm of immature cells. Neoplastic cells exhibit scarce cytoplasm, irregular nuclei, and sometimes evident nucleoli. H&E.

Classically, the disease begins with a solitary, erythematous/violaceous nodule located on the trunk or extremities in 50% of cases. New lesions appear and can be nodules or ecchymotic plaques, with fine scaling and size variations [3, 5, 10]. In the present case, the patient showed the classic clinical pattern of the appearance of cutaneous lesions.

Lymph node involvement (40-50%) and bone marrow and peripheral blood involvement (60-90%) also can occur. However, at diagnosis, these findings are absent in 70% of the cases [1, 4, 7]. On the other hand, splenomegaly (20%) and fulminant leukemia (5-25%) are uncommon [3, 4, 7].

There is still no consensus for the treatment of BPDCN. Intensive therapy for acute leukemia (CHOP - cyclophosphamide, hydroxyoxubicin, oncovin, prednisone) seems to increase the rate of complete remission [10]. However, only myeloablative

treatment with allogeneic bone marrow transplantation has resulted in a greater chance of long-term survival. Novel targets and agents are being studied, but no specific agent has been approved [10, 11].

Conclusion

Blastic plasmacytoid dendritic cell neoplasm is an extremely aggressive disease with rapid evolution. Early diagnosis is crucial for the adequate support. However, there is still no consensus for optimal treatment, especially in elderly patients. New active and tolerable drugs are necessary.

Potential conflicts of interest

The authors declare no conflicts of interests.

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