Case for diagnosis

Caso para diagnóstico

Paula Maio[1]
Ana Afonso[3]
José Cabeçadas[5]
Candida Fernandes[2]
Fernanda Sachse[4]
Jorge Cardoso[6]

CASE REPORT

We describe a 73-year-old male patient who was previously healthy. Two months prior to presenting to our department, the patient noticed a dermatosis involving the trunk, upper and lower limbs, consisting of multiple pruriginous patches, infiltrated plaques, and nodules with an erythematous surface. The patient did not complain of any additional constitutional symptoms besides pruritus. There were no complaints of weight loss, anorexia or malaise.

Physical examination revealed a dermatosis involving, in a bilateral and symmetrical fashion, the upper torso, abdomen, and proximal upper and lower limbs. The dermatosis consisted of roughly annular patches and plaques, with unequal infiltration, exhibiting an erythematous surface. The lesions were painless on palpation (Figure 1). The examination of lymph node chains was unremarkable. Palpation of the abdomen revealed no mass or organomegaly.

Incisional cutaneous biopsy revealed the presence of diffuse infiltration by intermediate-sized lymphoid cells, with immunohistochemical expression of CD45, CD4, CD56, and CD123 and negativity for myeloperoxidase, CD3, CD2, CD5, and CD7 (Figures 2 and 3).

Axial tomography of the thoraco-abdominal and pelvic regions, blood count, myelogram, bone biopsy, and immunophenotyping of peripheral and medullary blood did not reveal evidence of systemic involvement at the time.

The patient was started on oral prednisolone therapy (40 mg/day), which led to partial remission of the cutaneous lesions and pruritus. However, relapse was observed after the first month of therapy. After six months of follow-up, the patient still showed an exclusively cutaneous involvement, but rapid and fatal progression of the disease occurred after 8 months of follow-up, resulting in the patient's death.

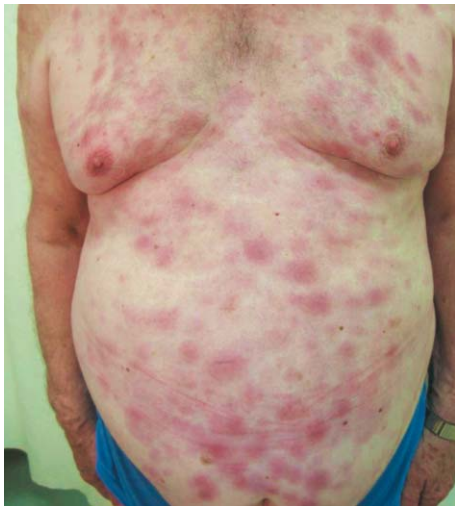

FIGURE 1: Clinical aspects. Multiple erythematous annular plaques

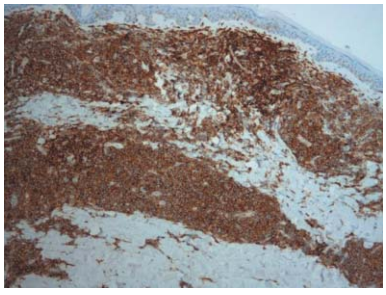

FIGURE 2: Immunohistochemistry. Diffuse infiltration by intermediate-sized lymphoid cells with immunohistochemical expression of CD4

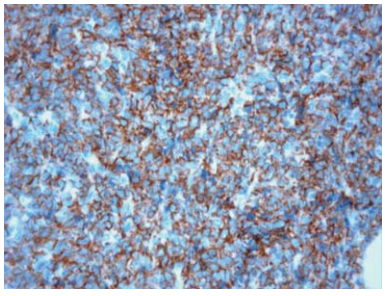

FIGURE 3: Immunohistochemistry showing positivity for CD56

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[1] MD – Hospital physician, Dermatology and Venereology - Hospital Curry Cabral – Centro Hospitalar de Lisboa Central (CHLC) – Lisbon, Portugal.
[2] MD – Hospital assistant - Hospital Curry Cabral – Centro Hospitalar de Lisboa Central (CHLC) – Lisbon, Portugal.
[3] MD – Hospital assistant - Anatomical Pathology Service, Hospital Curry Cabral – Centro Hospitalar de Lisboa Central (CHLC) – Lisbon, Portugal.
[4] MD – Hospital assistant – Dermatology Service, Instituto Português de Oncologia de Lisboa Francisco Gentil – IPOLFG (Francisco Gentil Portuguese Institute of Oncology) – Lisbon, Portugal.
[5] PhD - Hospital assistant - Anatomical Pathology Service, Instituto Português de Oncologia de Lisboa Francisco Gentil – IPOLFG (Francisco Gentil Portuguese Institute of Oncology) – Lisbon, Portugal.
[6] MD - Hospital assistant - Dermatology Service, Hospital Curry Cabral – Centro Hospitalar de Lisboa Central (CHLC) – Lisbon, Portugal.

DISCUSSION

Blastic plasmacytoid dendritic cell neoplasm is a recently classified entity.[1] The first cases were described in 1994, based on WHO recent diagnostic criteria for blastic NK-cell tumor.[2,3] It is more frequently diagnosed in the sixth decade of life, but it may occur at any age, and it seems to predominate in male patients. It is a rare neoplasm with a usually aggressive clinical progression, in spite of the established therapeutic regimens. Patients with this neoplasm have a mean survival time of 14 months.[2]

This neoplasm was formerly known as CD4-positive/CD56-positive hematodermic neoplasm.[3] It was considered to be originated from a myelomonocytic lineage precursor, given the common expression of the surface antigens CD4, CD5, and CD68, and also because of the common deletion of 5q.

Recent studies, however, identified the expression of another antigen, CD123, bringing it closer to the immunohistochemical profile of plasmacytoid dendritic cells. These cells are still very poorly characterized, both biochemically and immunophenotypically. They are present in small amounts in adult bone marrow, lymph nodes, tonsils, and blood plasma.[4-6]

The existence of skin lesions at the time of diagnosis is clinically frequent and, most of the time, is the main reason for the patient to seek clinical observation.[7,8] In the majority of the cases, physical examination reveals the existence of lymphadenopathies and/or splenomegaly. More rarely, involvement of the liver, lungs, eyeballs, and central nervous system may occur. Rapid progression of the disease is typical, with increasing number of lesions and progressive systemic involvement.

Cutaneous lesions are heterogeneous, erythematous or deep purple papules, plaques, nodules or tumors that can ulcerate, rarely reaching dimensions over 10 mm. Laboratory studies usually reveal thrombocytopenia, anemia, and more seldom leucopenia or leukocytosis. At the time of diagnosis, approximately 80% of patients already exhibit medullary involvement.[2,9]

We stress the importance of this case because of the rare absence of systemic involvement at the time of the initial presentation of this entity. ❑

Abstract: Blastic plasmacytoid dendritic cell tumor is a rare, highly aggressive systemic neoplasm for which effective therapies have not yet been established. We describe a 73-year-old man with multiple nodules and patches emerging on the trunk and limbs. Lesional skin biopsy revealed a plasmacytoid dendritic cell tumor with dense dermal infiltrate of tumor cells with blastoid features. No apparent systemic involvement was identified in the initial stage. The patient was treated with prednisone daily, with notorious improvement of the skin lesions, although no complete remission was obtained. During the six-month follow-up period, no disease progression was documented, but fatal systemic progression occurred after that period of time.
Keywords: Dendritic cells; Leukemia; Neoplasms

Resumo: A leucemia de células dendríticas plasmocitóides blásticas é uma entidade de classificação recente. Descreve-se o caso de um homem de 73 anos com dermatose envolvendo manchas, placas infiltradas e nódulos eritematosos na face anterior do tronco, no dorso e nos membros. A biopsia cutânea revelou presença de infiltração difusa por células linfóides de tamanho intermediário. A imunocitoquímica permitiu o diagnóstico de neoplasia de células dendríticas plasmocitóides blásticas. O estadiamento não revelou envolvimento sistêmico na época do diagnóstico. O doente iniciou a terapêutica com prednisolona e apresentou remissão inicial das lesões cutâneas. Posteriormente, observou-se progressão sistêmica da doença, e o doente veio a falecer.
Palavras-chave: Células dendríticas; Leucemia; Neoplasias

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Mailing address:
Paula Maio
Rua da Beneficência, 8. Lisboa, Portugal.
1069-166
E-mail address: paulamaio@gmail.com

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