

CASE REPORT

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Primary cutaneous anaplastic large cell lymphoma initially clinically considered to be pyoderma gangrenosum

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Abstract

Primary CD30+ anaplastic large cell lymphoma, which is generally seen in adults, is the second most common cutaneous T-cell lymphoma after mycosis fungoides. The lesions are characterized by red-brown plaque or nodules often with ulceration. Although usually solitary, there may occasionally be multiple lesions. Here in this case report, a 44-year old male patient with multifocal primary cutaneous anaplastic large cell lymphoma is presented. Primary cutaneous anaplastic large cell lymphoma is a rare disease especially when it is multifocal and has no systemic involvement like in this case. Therefore this case is found valuable to share.

Keywords: Cutaneous lymphoma, anaplastic, primary

Introduction

The term of primary CD30+ lymphoproliferative diseases encompasses CD30+ primary cutaneous anaplastic large cell lymphoma and lymphomatoid papulosis headings. Both of them show resemblance in terms of clinical, histopathological and immunopathological characteristics [1]. Ulceration showing red-brown plaque or nodules is often clinically observed in anaplastic large cell lymphoma [2]. The primary cutaneous form has a good prognosis and 5- year survival rate is approximately 90%. Few cases have been reported to spread to the extracutaneous tissues such as lymph nodes, bone marrow, lungs and the central nervous system [3].

Case Report

A 44-year old male patient applied to our polyclinic with the complaint of wounds on his body which has been ongoing for 8 years. At first, the wounds had started in the neck region and later, it had spread to other regions of the body. As a result of the biopsy which was performed at an external center, a diagnosis of pyoderma gangrenosum had been made and the patient has had dapsone, cyclosporine, prednisolone and infliximab treatments. The patient was unresponsive to these treatments and after that he applied to our polyclinic. In the dermatological examination, dark brown, indurated, ulcerated lesions 1-5 cm in size and many atrophic scars of varying sizes were observed on the face, trunk, legs (Figure 1a,1b,1c,1d).

The patient was hospitalized to our clinic for further research concerning the pre-diagnoses of cutaneous lymphoma, tumoral stage mycosis fungoides and pyoderma gangrenosum.



Figure 1A. In the dermatological examination, dark brown, indurated, ulcerated lesions 1-5 cm in size and many atrophic scars of varying sizes



Figure 1B. In the dermatological examination, dark brown, indurated, ulcerated lesions 1-5 cm in size and many atrophic scars of varying sizes

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Figure 1C. In the dermatological examination, dark brown, indurated, ulcerated lesions 1-5 cm in size and many atrophic scars of varying sizes

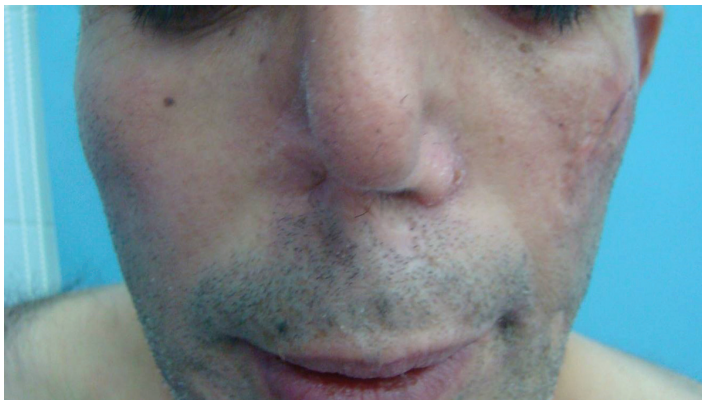


Figure 1D. In the dermatological examination, dark brown, indurated, ulcerated lesions 1-5 cm in size and many atrophic scars of varying sizes

A skin biopsy was performed and it revealed a mild parakeratosis, moderate acanthosis, lymphocyte exocytosis, minimal focal spongiosis, one ulceration focus and epidermotropism in the epidermis.

In the perivascular, perineural and periadnexial regions, there was patchy lymphoid tumoral infiltration with round-oval cells showing nuclear polymorphism whole through the dermis. In some blood vessels, subendothelial lymphoid infiltration were recognized also. There were extensive areas of necrosis in the tumoural region and in the subcutaneous fatty tissue (Figure 2).

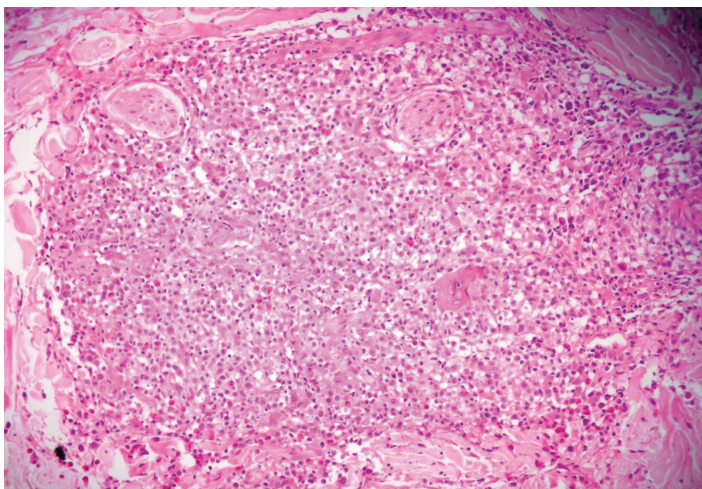


Figure 2. Necrosis in the tumoural region and in the subcutaneous fatty tissue

Immunohistochemically; lymphoid cells showed diffuse staining (+3) with Leukocyte Common Antigen (LCA), neoplastic lymphoid cells showed the same degree staining with CD3, 70 % of the neoplastic lymphoid cells were CD4 (+), 30 % of them were CD8 (+). Blastic cells were CD30 (+) (<75%), mast cells were CD117 (+), some of the reactive lymphoid cells were CD7 (+) and ALK was negative (Figure 3).

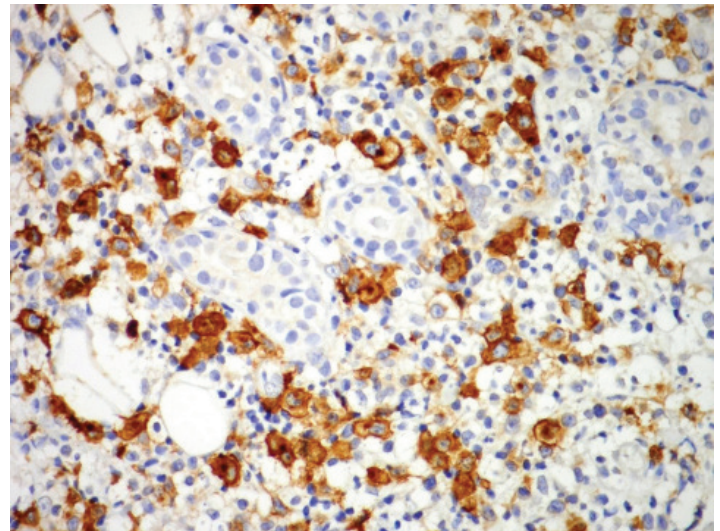


Figure 3. Blastic cells were CD30 (+) (<75%), mast cells were CD117 (+), some of the reactive lymphoid cells were CD7 (+) and ALK was negative

In the direct immunofluorescence, fine granular mild staining (+1) with IgM was observed at the dermoepidermal junction and IgA, C3c, C1q stainings were negative. Investigations for systemic involvement like hemogram, peripheral blood smear, ultrasonography of the neck and the whole abdomen, pulmonary radiography, abdominal and thoracic tomographies revealed normal results.

Now that the lesion itself originated from the skin and considering all the morphologic and immune findings, the patient was diagnosed as having primary cutaneous anaplastic large cell lymphoma.

Discussion

Anaplastic large cell lymphoma was first described by Stein et al in 1985, as a lymphoma with large CD30 positive anaplastic lymphocytes. It is 2-3 times more common in males than females. In the literature, cases with the liver, spleen, skin, testis, stomach, bone and bone marrow involvement apart from the lymph node involvement are reported [4].

Peripheral T-cell lymphomas have been classified in 4 main groups as leukemic, nodal, extranodal and cutaneous form by the World Health Organisation (WHO). Primary skin lymphomas show histological similarities to systemic lymphomas, but they have different clinical characteristics, prognosis and they are classified differently [5]. Primary cutaneous anaplastic large cell lymphoma is a T-cell lymphoma which has at least 75 % CD30 (+) anaplastic lymphoid cells [4,6]. Anaplastic large cell lymphoma constitutes 1%-3% of all cutaneous lymphomas [7].

Clinically, nodules ranging in diameter from 0.5-15 cm are often observed in a specific anatomic region. Ulceration of the lesions is frequently seen. While systemic involvement is rare in patients

with solitary lesions, it is more often observed in patients with multiple lesions [6,8]. In our case, there were many indurated, dark brown, encrusted, ulcerated lesions 1-5 cm in size on the face, trunk and legs.

In the histopathologic examination, there were large blast-like cells with horseshoe-shaped pleomorphic nuclei and more than 75% of the cells had CD30 expression. Anaplastic lymphoma kinase (ALK) gene expression is often determined in primary systemic anaplastic large cell lymphoma [9]. In our case, ALK gene expression was negative.

The mean 4-year survival rate in primary cutaneous anaplastic large cell lymphoma has been reported as 90%. Early diagnosis is important in respect of preventing a progressive course and reducing the mortality rate, as well as in affecting the lifespan and the quality of life directly. Advanced age, increased level of lactate dehydrogenase and bone marrow involvement are factors affecting the prognosis of T-cell lymphomas [4,8].

Although chemotherapy, surgical therapy and radiotherapy can be used alone or in combination, there is no standard treatment approach. Other treatment options include oral bexarotene, interferon alpha, 13-cis-retinoic acid, methotrexate, intralesional methotrexate, etoposide, anti-CD30 antibodies and stem cell transplantation [9,10].

As there is a high possibility of systemic involvement in multifocal anaplastic large cell lymphoma limited to the skin, aggressive treatment is recommended, although there is as yet no consensus on using the combined chemotherapy [8,9]. As ALK was negative and no systemic involvement was determined in our case, methotrexate treatment was planned.

At the beginning of the disease, it can be difficult to make a diagnosis of primary cutaneous lymphoma, as histopathological and skin findings have a non-specific course. However, early diagnosis is important in respect of preventing the progressive course of the disease.

In the early stages of the disease in the current case, the diagnosis of pyoderma gangrenosum was considered but no response was obtained from the treatments. As a result of the second skin biopsy,

the correct diagnosis was made. Therefore, although rarely seen, cutaneous lymphomas are a disease group which should be kept in mind in cases with non-specific, ulcerated, nodular skin lesions.

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