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Subcutaneous Panniculitis-Like T-Cell LymphomaAssociated to Systemic Lupus Erythematosus

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Abstract **Case Report**

Subcutaneous panniculitis-like T-cell lymphoma (SPTCL) accounts for lessthan 1% of all Lymphomas that manifest themselves with sub cutaneous nodes, SPTCL may be associated to a systemic disease. We report in this paper the first case described in our department of a 57 years old female patient followed up for 5 years for a Subcutaneous panniculitis-like T-cell lymphoma, treated with dermocorticosteroids after 4 years of therapeutic abstention and who developed a Systemic lupus erythematosus for which she was put under treatment with a very good evolution.

Keywords: patient, SPTCL, dermocorticosteroids, therapeutic abstention.

INTRODUCTION

Panniculitis-like T-cell lymphoma is considered as a rare entity amongst cutaneous lymphomas, with a variable clinical description and a generally favorable prognosis.; this lymphoma may be associated with many autoimmune diseases, but in half the cases and more frequently, it is associated with systemic lupus erythematosus; The diagnosis is anatomopathological and the therapeutic management are not codified, and there is still no consensual therapeutic protocol; the treatment can vary from a simple local corticotherapy to a chemotherapy based on anthracyclines and/or radiotherapy of the cutaneous lesions.

CLINICAL OBSERVATION

The patient is a 57 years old female with a past medical history of treated tuberculosis at the age of 25 years old, a splenectomy at the age of 20 years old and a myomectomie at the age of 54 years old.

The onset of symptoms goes back to 4 years ago with the onset of a 4 cm left jugal swelling (Fig-1), unilateral that became bilateral, appearing to be inflammatory, erythemato-pruritic and desquamative.

The clinical examination during the first consultation showed a patient in very good general condition presenting with bilateral erythemato-squamous and pruritic lesions on the jugal level, the palpation found sub-epidermal sub-centimetricnodes, the lymph node and the rest of the somatic examination were without any pathological findings.

Several skin biopsies with immunohistochemistry were performed, showing a CD20+, CD3+, CD5-, CD8+, CD4-, CD56-, Ki67 subcutaneous lymphoma at 20%.The extension workup did not reveal any other tumor location. The patient refused treatment for 4 years, but with worsening skin lesions, she again requested treatment.

Therapeutically, the patient was put on dermocorticoids with a very good clinical evolution; but she decided to stop the treatment and refused to continue it after 2 months, nevertheless, she remained under surveillance, the evolution was favorable with the disappearance of the cutaneous lesions as well as the sub-epidermal nodules but a persistence of sequelae lesions (Fig-2).

Four months later, the evolution was characterized by the appearance of bilateral areas of alopecia measuring 10 cm each (Fig-3), from which the biopsy came back in favor of a chronic cutaneous lupus.

An immunological assessment was requested and came back in favor of lupus. The patient was

therefore put on a hydroxychloroquine treatment with a very good clinical evolution, especially the appearance of hair on the previously described areas of alopecia at the end of the three months of treatment. The patient is currently on the same treatment.

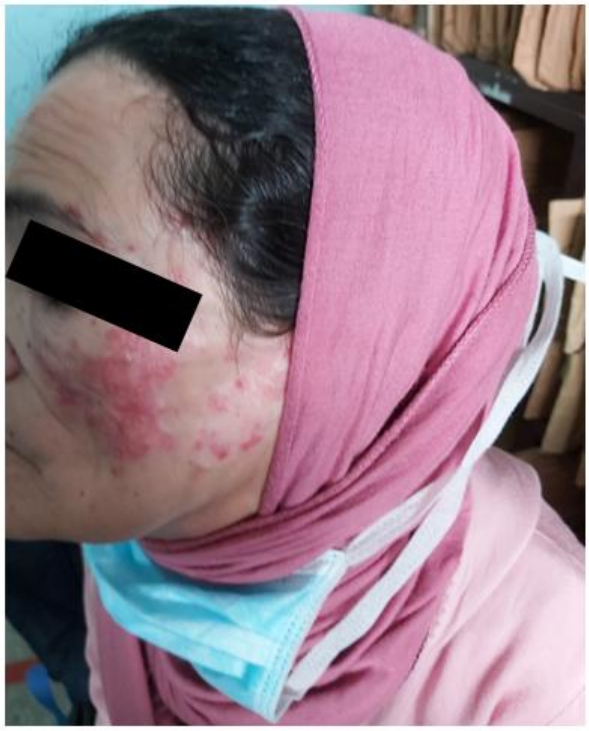

Fig-1: Lesions before treatment

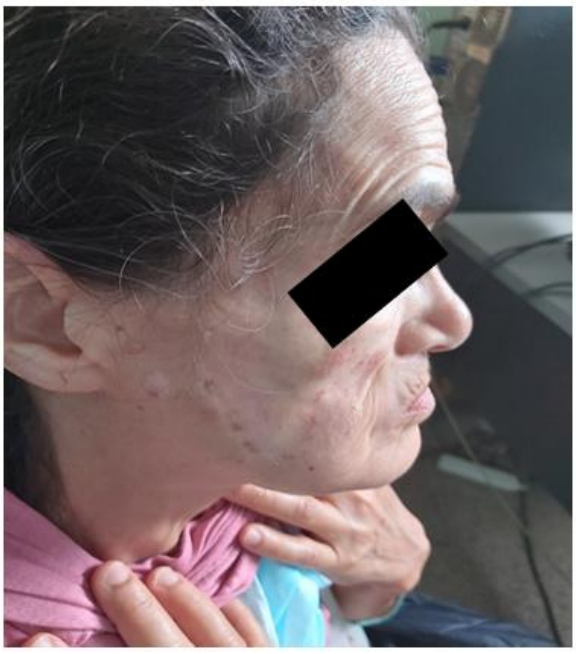

Fig-2: Lesions after dermocorticoids

Fig-3: Areas of alopecia measuring 10 cm

DISCUSSION

The panniculitis-T like cell lymphoma is a rare entity that less than 1% of lymphomas [1]; this lymphoma belongs to lymphomas that have a cutaneous tropism, and that infiltrates the subcutaneous tissues [2]. The clinical description is essentially made of subcutaneous nodules or plaques, localized especially on the face as what has been described by cassis et al in 2004 and Kosari et al., in 2014 [3, 4] in the absence of other lesions; moreover the other most frequent localizations are the trunk and the extremities. In our observation, it is a purely facial localization, the examination of the trunk and extremities is normal.

Panniculitis-T like cell lymphoma (SPTL) was first described in 1991 by Gonzalez's team [5], and in the EORTC as SPTL as cutaneous T-cell lymphoma, and finally by the WHO; the clinical features are dominated by the appearance of cutaneous nodules, the appearance of general signs including fever and night sweats as well as a weight loss, usually, quantified by the patient [6].

A review of the Japanese literature including 22 cases of SPTL, as well as the EORTC study group, in a study of 83 cases of SPTL, described the same clinical signs, specifying that the patients had an average age of 36 years old, the median delay in diagnosis was 47 months, ranging up to 10 years, and the skin lesions described were skin nodules, but also deep patches, of which the most frequent locations were on the legs in more than 50 % of the patients, and less frequently the face representing 25 % of the lesions. In our case, the subcutaneous lesions were located bilateraly on the jugal area.

Biologically, the most frequent abnormalities found in patients and described in the Japanese or the European series are blood count abnormalities such as anemia, leuko-neutropenia or even pancytopenia. The biological depiction may be confusing with a macrophage activation syndrome.

The role of autoimmunity seems to be important in the pathophysiology of the disease; An autoimmune disease is present in about half of the patients with a Tpanniculitis like lymphoma with a predominance of the association with lupus [8].

In both series, an autoimmune disease was associated with SPTL: Gougerot-Sjögren's syndrome [9], Type I Diabetes but also Lupus and Raynaud's syndrome[10]. In our case it is a cutaneous lupus; our patient developed areas of alopecia and the investigations led to the diagnosis of a LED.

The diagnosis of SPTL is based on the anatomopathological study of the sampled subcutaneous tissue which objectifies a lymphomatous infiltration by cells of irregular size with a hyperchromatic nucleus, the presence of vacuolated hystiocytes, with a possibility of vascular damage, the lymphomatous cells are CD8+, they also express granzyme B and perforin as well as TIA1, CD56 is negative finally CD123 and is in the majority of the cases negative [11-14].

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The therapeutic management is very variable, it is different according to the clinical stage and the extension workup as well as the associated pathologies; radiotherapy of the lesions or corticotherapy can be the first-line treatment but we can also indicate an anthracycline-based chemotherapy [7], our patient benefited from a treatment with dermocorticosteroids with a very good response and especially the disappearance of the subcutaneous nodules, in addition she was put under treatment for the lupus with a very good evolution of the alopecia lesions.

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