

Subcutaneous Panniculitis-Like T-Cell Lymphoma of the Cheek Mimicking a Lupus Erythematosus Panniculitis

Maouni Safae^{1*}, El Anzi Ouiam¹, Sialiti Sanae¹, Zenati Kaoutar², Meziane Mariam¹, Benzekri Leila¹, Nadia Ismaili¹, Senouci Karima¹ and Hassam Badredine¹

¹Department of Dermatology-Venereology, Morocco

²Department of Anatomopathology, Morocco

*Corresponding author: Maouni Safae, Department of Dermatology-Venereology, Morocco

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Abstract

Subcutaneous Panniculitis-like T-lymphoma (SPTL) is a rare non-epidermotropic cutaneous T cell lymphoma, which perform 1% of all non-Hodgkin's lymphomas. The lesions are usually localized in the upper limbs and the trunk. the localization on the cheeks is unusual. The diagnosis of SPTCL depends on the pathologic examination of skin biopsy, but the manifestations of SPTCL are varied and mimic autoimmune disorders. We report a case of a subcutaneous panniculitis-like t-cell lymphoma with an unusual localization in the cheek, mimicking a lupus erythematosus panniculitis, evolving since 2 years in a 40-year-old woman.

Keywords: Lymphoma; Panniculitis; Cheeks; Lupus

Introduction

Subcutaneous Panniculitis-like T-lymphoma (SPTL) is a rare non-epidermotropic cutaneous T cell lymphoma. The lesions are usually localized in the upper limbs and the trunk. the localization on the cheeks is unusual [1]. The diagnosis of SPTCL depends on the pathologic examination of skin biopsy, but the manifestations of SPTCL are varied and mimic autoimmune disorders. We report a rare case of a subcutaneous panniculitis-like t-cell lymphoma of the cheek evolving since 2 years in a 40-year-old woman.

Observation

A 40-year-old woman presented with painless skin lesions in the both cheeks. The lesions appeared 2 years earlier as a nodule that has evolved into plaque. Clinical examination revealed a large rounded infiltrated plate in the cheek, bilateral, measuring 5 cm on the right and 3 cm on the left, flesh-colored with cuculiform depression. She had no other symptoms, organomegaly, or lymphadenopathy. Histopathological examination of a deep skin biopsy revealed a lymphoid infiltrate dissociating adipocyte, made of atypical cells with round nucleus and delicate chromatin. immunohistochemistry of atypical cells was positive for CD3 and

CD8 and negative for CD20, CD4, CD5 and CD56. Positron emission tomography (PET) of the whole body showed subcutaneous thickening of the cheeks, with localization in the vertebral bone without regional lymph node. Biological examinations have not revealed signs of macrophage activation syndrome. With all this finding, the diagnosis of subcutaneous panniculitis-like T-cell lymphoma (SPTCL) was made and a chemotherapy was indicated for this patient (Figures 1 & 2).



Figure 1: Infiltrated plate in the left cheek with cuculiform depression.

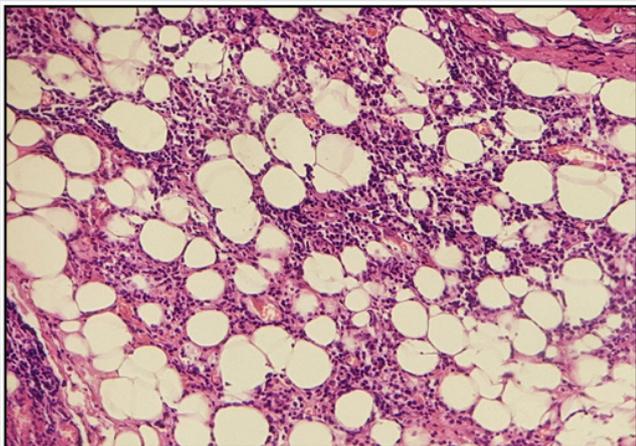


Figure 2: Skin biopsy showing a lymphoid infiltrate dissociating adipocyte, made of atypical cells with round nucleus and delicate chromatin (HE x 10).

Discussion

SPTCL is a uncommon cutaneous T-cell lymphoma, which perform 1% of all non-Hodgkin's lymphomas. it was first reported by Gonzalez in 1991 [2], then described by the World Health Organization as a distinguish entity in 2001[3]. It affects the young adult; the median age of the diagnosis is 39 years. Clinically, SPTCL is characterized by indolent evolutionary lesions, Plaque type, painless subcutaneous nodules or necrotic ulcers, sometimes associated with mucosal involvement. Generally, the lesions are localized at the extremities and the trunk, the localization on the cheek is extremely rare. The differential diagnosis in this localization arises at the beginning with the benign panniculitis, the eczema and the cellulite and especially with the lupus panniculitis at a late stage. Our patient had an atypical clinical manifestation with cuculiform scars suggestive of lupus panniculitis, for which she had an assessment of lupus, but it was normal.

There are two types of SPTCL [4]: an $\alpha\beta$ (SPTCL-AB) and a $\gamma\delta$ subtype (SPTCL-GD).1 SPTL-ABs generally have a CD4-/CD81/

CD56- T-cell phenotype and a favorable prognosis, whereas SPTL-GDs typically have a CD4- /CD8- T-cell phenotype with frequent coexpression of CD56, a higher association with hemophagocytic syndrome, and a poor prognosis. The diagnosis of SPTCL is based on clinical, histological and immunohistochemical criteria. The treatment is essentially based on chemotherapy with cyclophosphamide, doxorubicin, vincristine and prednisolone, with an overall remission rate of 50% [5]. In some cases, an autologous bone marrow transplant may be indicated. The overall survival rate of the Tcell receptor at 5 years exceeds 80%.

Conclusion

In conclusion, we reported a case of SPTL with a very rare localization in the cheeks, which almost always poses the problem of differential diagnosis with autoimmune diseases, and specifically lupus panniculitis in our case. Hence the interest of a good interrogation and a complete clinical examination in front of a subcutaneous nodules of the cheeks.

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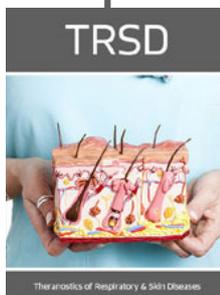
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