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Clinical case

Rapidly fatal anaplastic CD30+ primary cutaneous T-cell lymphoma: about a case

Lymphome cutané T primitif anaplasique CD30 + d'évolution agressive rapidement fatale : à propos d'un cas

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Résumé

Le lymphome cutané T primitif anaplasique CD30+ est un type de lymphome cutané rarement décrite dans la littérature et sa forme tumorale disséminée est exceptionnelle. Il s'agissait d'un homme 73 ans qui a présenté des tumeurs cutanées nodulaires dont la taille dépassait 5 cm, fermes, indolores, secondairement ulcérées, devenant ulcéro-bourgeonnantes à fond nécrotico-purulent très douloureuses. Ces lésions étaient localisées sur les membres inférieurs prédominant à droite. L'examen extra-dermatologique retrouvait : des macropolyadénopathies tumorales satellites droites, poplitées, inguinales, axillaires et cervicales. Le reste de l'examen physique était normal. L'histopathologie cutanée avec immunomarquage avais permis le diagnostic de lymphome T anaplasique CD30 positif. L'évolution était rapidement fatale malgré les cures de chimiothérapie. Le cas que nous rapportons illustre le problème diagnostic et thérapeutique des lymphomes T cutanés encore entier dans notre contexte d'exercice en Afrique par l'absence de l'immunothérapie et les effets secondaires de la chimiothérapie qui grève le pronostic du malade en dehors de l'agressivité de la maladie elle-même.

Mots-clés : lymphome cutanée T CD30+, lésions cutanées tumorales, rapport de cas.

Abstract

CD30+ anaplastic primary cutaneous T-cell lymphoma is a type of cutaneous lymphoma rarely described in the literature and has an exceptional disseminated tumor form. This was a 73-year-old man who presented with nodular skin tumors whose size exceeded 5 cm, firm, painless, secondarily ulcerated, becoming ulcerative budding with a necrotic-purulent background and very painful. These lesions were located on the lower limbs, predominant on the right. Extra-dermatological examination found: right satellite tumor macropolyadenopathy, popliteal, inguinal, axillary and cervical. The rest of the physical examination was normal. Immunostained skin histopathology led to the diagnosis of CD30-positive anaplastic T-cell lymphoma. The course was quickly fatal despite the chemotherapy treatments. The case we report illustrates the diagnostic and therapeutic problem of cutaneous T-cell lymphomas that are still intact in our context of practice in Africa by the absence of immunotherapy and the side effects of chemotherapy which weigh on the patient's prognosis

apart from the aggressiveness of the disease itself.
Keywords: CD30+ cutaneous T-cell lymphoma, tumor skin lesions, case report.

Introduction

CD30-positive primary cutaneous lymphomas are the most common group of primary cutaneous T-cell lymphoma after mycosis fungoides accounting for 30% of cases [1]. However, in their anaplastic forms, the aggressive multinodular clinical presentation is exceptionally described [2]. Thus, this clinical presentation led to confusion with other differential diagnoses, and mainly the systemic form of anaplastic lymphoma. The history of the disease and immunohistochemistry in particular, were an essential element of the diagnosis. Thus, through the case we report, we want to illustrate the problem of diagnosis and treatment in our African context linked to the inaccessibility of immunohistochemistry and targeted therapy which leads to the rapidly fatal evolution of our patients.

Clinical case

A 73-year-old man, known to be hypertensive, poorly monitored, with no particular pathological history, was seen for ulcerated tumor skin lesions that had been evolving for 3 months.

On admission, the general condition was altered, there was no fever, the pulse was normal, the blood pressure was normal. The dermatological examination found nodular tumors that were more than 5 cm in size, firm, painless, purplish with a beetroot skin appearance. These lesions were secondarily ulcerated, becoming ulcerative budding with a very painful necrotic-purulent background. The lower limbs were affected with a predominance on the right (Figure 1). Extradermatological examination recovered: right satellite tumor macropolyadenopathy, popliteal, inguinal, axillary and cervical. The rest of the physical examination was normal.

The diagnostic hypotheses mentioned were: cutaneous lymphoma, skin metastasis.

The cutaneous histopathology showed: an epidermis of normal thickness; the dermis hosted a dense atypical lymphoid infiltrate made up of a large number of atypical lymphocytes, with a hyperchromatic nucleus; it was associated with irregular lymphocytes of small size, of reactive appearance. The results of the immunohistochemical study were obtained after a 2-month follow-up with worsening of the disease in the absence of etiological therapy. Indeed, the unavailability and cost of immunohistochemistry in our context made this exploration inaccessible for our patient. The results showed that large lymphoid cells expressed the CD30 antigen intensely and homogeneously. Thus, in the face of this tumor strongly expressing the CD30 antigen, it was necessary to discuss at the immunohistochemical level certain differential diagnoses such as: CD30+ anaplastic cutaneous T-cell lymphoma, systemic anaplastic lymphoma, transformed mycosis fungoides (MF), B-cell lymphomas with T cell markers and CD30 expression such as plasmablastic lymphoma and primary effusion lymphoma. Thus, assays of additional immunohistochemical markers were performed and had shown: a T phenotype with CD2 expression and absence of significant expression of CD3, CD5, CD7, CD4 and CD8. There were rare CD8+ lymphocytes without antigenic loss. The granzyme B label was negative. The P80/ALK1 marking was negative. The search for EBV by in situ hybridization technique was negative. These results allowed us to rule out all differential diagnoses and retain the diagnosis of CD30+ anaplastic cutaneous T-cell lymphoma.

The extension work-up first included a complete physical examination which was normal apart from right inguinal, axillary and supraclavicular lymphadenopathy of tumor appearance. Then, a thoraco-abdomino-pelvic CT scan showed pulmonary nodules strongly suspected of secondary location. In the blood, the blood count was normal, the retroviral serology was negative. The tumor was classified as T3N2M1.

COP cytoreductive chemotherapy (Cyclophosphamide 600mg/m2 IV on D1, Oncovin 1.4mg/m2 on IV on D1, Prednisone 40mg/m2 on D1 to D5) was initiated, after a multidisciplinary consultation meeting (SmPC) with the haematologists and oncologists. An extension of the lesions to the chest was noted 15 days after the chemotherapy course with an alteration in the general condition. Then, after a PCR, the patient was put on gemcitabine at a dose of 800mg/m² 3 months after the first course of COP. The death occurred 7 days later in a shock picture with multi-organ failure despite all resuscitation measures.

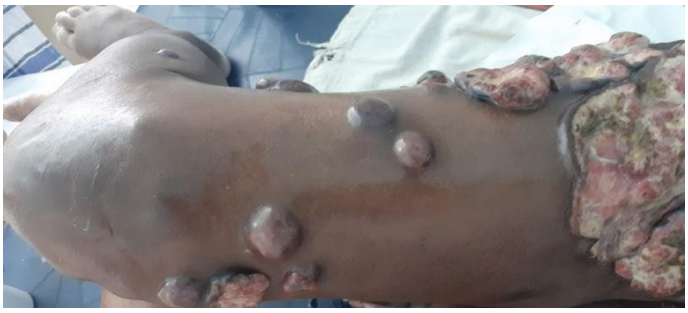

Figure 1: Lower limb nodular purplish skin lesions of the lower limb progressing to ulcerative budding lesions in primary cutaneous anaplastic lymphoma.

Discussion

We reported one case of CD30-positive anaplastic T-cell lymphoma with a rapidly fatal course. CD30-positive primary cutaneous T-cell lymphomas (PCLs) CD30 are the most common group of primary cutaneous T-cell lymphoma after mycosis fungoides accounting for 30% of cases [1]. These cutaneous lymphomas have usually been classified on the basis of their clinical presentation as lymphomatoid papulosis, primary cutaneous anaplastic large cell lymphoma and intermediate forms. Anaplastic CD30+ LCTP is, by definition, an anaplastic large T-cell neoplasia, pleomorphic CD30+.

This aggressive multinodular diffuse ulcerated clinical form is not usual. Indeed, primary cutaneous anaplastic lymphomas are characterized by nodules or solitary or localized tumors, papules, ulcerations. A rapid and aggressive course is rarely described and is a factor in a poor prognosis [2]. The extracutaneous lymph node and pulmonary involvement found in our patient is described in 10% of cases [2,3,4]

Because of the aggressiveness and systemic involvement, the difficulty lay first in the differential diagnosis with systemic anaplastic lymphoma. This diagnosis was ruled out by the history of the disease, which did not find chronic lymph node involvement preceding the skin lesions [5] and by immunohistochemistry which found a loss of broad antigenic expression including CD3 and CD5 [6]. Transformed mycosis fungoides (MF) has been ruled out due to the absence of a previous history of MF [7]. The negativity of EBV testing by hybridization technique ruled out the hypothesis of B-cell lymphomas with T-cell markers and CD30 expression, such as plasmablastic lymphoma or primary effusion lymphoma. [8]

The etiological factors found in our patient were the male sex and the age of over 60 years [8]. Other etiological factors described in the literature are secondary immunosuppression, HIV infection, and organ transplant recipients. [9,10]. The rapidly fatal evolution of our patient illustrates the problem first of all diagnosis with a delay in the immunohistochemical examination, the only way to make a positive diagnosis of these types of lymphoma, then a therapeutic problem of cutaneous T-cell lymphomas which still remains unsolved in our context of practice in the absence of immunotherapy with the side effects of chemotherapy which affects the patient's prognosis apart from the aggressiveness of the disease itself.

Conclusion

Our case illustrates the problem of delayed diagnosis and treatment of T-cell cutaneous lymphomas, which still remains unresolved in our African practice context

due to the inaccessibility of immunohistochemistry and targeted immunotherapy. The rapidly fatal course we observed may be the only possible course for the majority of patients with cutaneous T-cell lymphoma in our countries. This tumor aggressiveness could be increased by chemotherapy. This deficit could be improved by the development of international collaborations for better patient care.

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