Case for diagnosis*

Caso para diagnóstico

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CASE REPORT

A 53-year-old Caucasian male was referred to our Department due to an asymptomatic erythematous plaque on the left buttock evolving for 5 months and with progressive increase. On physical examination, an erythematous-brownish plaque was observed with a raised erythematous-red border and areas of erosion (Figure 1). There were no other signs or symptoms. A biopsy showed hyperkeratosis, acanthosis and spongiosis, as well as epidermotropism of isolated and grouped large-sized atypical lymphocytes with pale halos, also present in the upper dermis. Immunohistochemistry revealed positive staining of these cells for CD3 and CD8 (Figure 2) without staining for CD4, CD30 and CD56, consistent with the diagnosis of pagetoid reticulosis (PR) – Woringer-Kolopp disease (WKD). The microbiological examinations of the skin were negative. Three weeks later the patient developed several nummular erythematous lesions with a scaling border scattered on the legs (Figure 3). We repeated the biopsy, which revealed identical findings plus positive staining for CD45 and a proliferative index of 90% (Ki67). Subsequently, these lesions increased in size, assuming the shape of the original plaque and a diagnosis of Ketron-Goodman disease (KGD) was established. The patient started treatment once daily with betamethasone dipropionate ointment and, simultaneously, was referred to the haematological outpatient department to exclude systemic involvement. All tests were normal, including the peripheral blood immunophenotyping, the bone marrow biopsy and the thoraco-abdomino-pelvic computed tomography scans. After 4 months of therapy with topical betamethasone dipropionate, all the lesions cleared, with the exception of a single lesion on the left foot, which was submitted to radiotherapy and resolved in 4 months. In a follow-up over 2 years the patient has remained free of signs and symptoms.

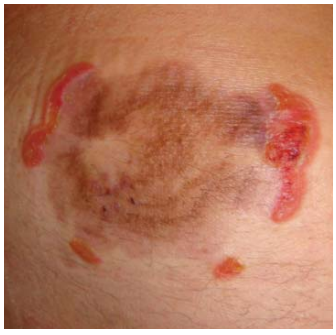

FIGURE 1 : Erythematous brownish plaque surrounded by a raised erythematous red border with some erosions

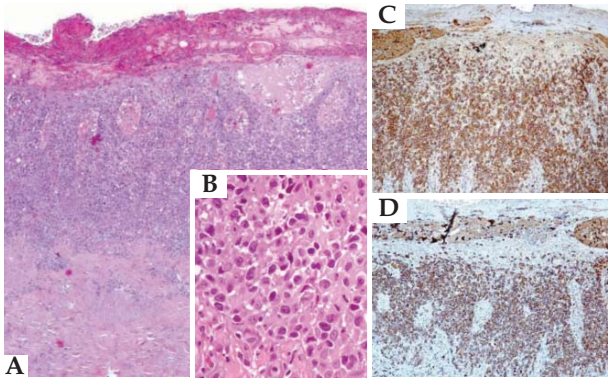

FIGURE 2 : A pathological infiltration of the epidermis by large-sized cells, single or arranged in clusters and dermal fibrosis containing these cells on H&E 10x **(a)**. Atypical large cells displaying vacuolated, abundant cytoplasm and big, hyperchromatic, pleomorphic nuclei on H&E 40x **(b)**. Immunohistochemistry with positive staining for CD8 **(c)** and CD3 **(d)**

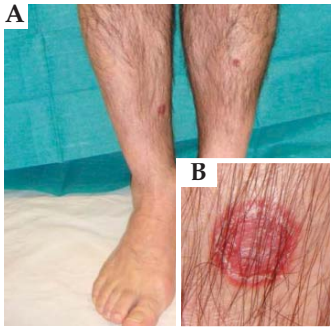

FIGURE 3 : Nummular erythematous lesions scattered on the legs **(a)** in close detail displaying a scaling border **(b)**

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DISCUSSION

Pagetoid reticulosis is a low-grade rare malignant variant of mycosis fungoides (MF) characterized by the presence of localized patches or plaques, but usually presents as a solitary lesion that is commonly localized on the extremities, with an intraepidermal proliferation of neoplastic T cells that may have either a $CD4^+$,$CD8^-$ or a $CD4^-$,$CD8^+$phenotype.[1] KGD was initially described as a disseminated type of PR.[2] However, according to the most recent classification consensus for cutaneous lymphomas, KGD would currently be more likely to be classified as aggressive epidermotropic CD8 T-cutaneous lymphoma, cutaneous gamma/delta T-lymphoma, or tumor-stage MF, depending on the clinical-histological picture.[1] The term PR should only be used for the localized type – WKD.[1] Our case raises some issues in the light of recent classification consensus.[1] Our patient was initially diagnosed with localized PR – WKD, but then developed widespread lesions on the legs which, together with the histopathology and immunophenotype ($CD45^+$, $CD56^-$) displaying an high proliferative index, led to a reclassification for KGD.[3] According to the classification consensus, this would be in line with a diagnosis of aggressive CD8 T-cutaneous lymphoma.[1] Nevertheless there were no signs of aggressive clinical behavior or necrotic keratinocytes in histopathology, which provided us with the motive to continue with a low-grade treatment regimen. The preferred mode of treatment for PR is radiotherapy and, in some cases, topical treatment with nitrogen mustard or steroids.[1] Surgical excision was used in the past for few small lesions, but the excellent response to topical and/or radiation therapy rendered surgical intervention obsolete.[4,5] Localized PR has good prognosis without extracutaneous dissemination or disease-related deaths.[1] In contrast, the few published cases of KGD report fatal outcomes, systemic involvement and interferon therapy, with an 8-month follow-up without relapse.[5,6] In our case, a follow-up of 2 years without recurrences or systemic involvement favors an indolent form of cutaneous T-lymphoma. Long-term observation is however necessary, considering that even if the cutaneous lesions clear up this may not mean resolution of the disease, since the disease may recur as long as 10 years after therapy.[5] ❑

Abstract: Ketron-Goodman disease was formerly considered a disseminated type of pagetoid reticulosis. However, according to the new classification consensus, it should be regarded as aggressive epidermotropic CD8 T-cutaneous lymphoma, cutaneous gamma/delta T-lymphoma, or tumor-stage mycosis fungoides, depending on the clinical-histological picture. This case highlights a rare and challenging presentation of Ketron-Goodman disease with an indolent presentation and evolution and good response to a low-grade treatment regimen, not fitting well into the new classification criteria.
Keywords: Lymphoma, T-Cell, cutaneous; Mycosis fungoides; Pagetoid reticulosis

Resumo: A doença de Ketron-Goodman foi inicialmente considerada uma forma disseminada de reticulose pagetóide. Mas, de acordo com o atual sistema de classificação e dependendo do quadro clínico-patológico deve ser antes vista como um linfoma T CD8 agressivo epidermotrópico, linfoma T gama/delta ou micose fungóide, estadio tumoral. Pretendemos realçar esta doença rara que pode suscitar dúvidas no diagnóstico. Neste caso, a apresentação e evolução foram indolentes com boa resposta a um tratamento pouco agressivo, não se enquadrando bem nas novas propostas de classificação da doença.
Palavras-chave: Linfoma cutâneo de células T; Micose fungóide; Reticulose pagetoide

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