Our Dermatology Online

Case Report

Pagetoid reticulosis Woringer–Kolopp type: A report of two cases at a third-level center

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ABSTRACT

Pagetoid reticulosis is a clinical and histological variant of mycosis fungoides that shows epidermotropism in histology and a classic clinic with acral disease. Herein, we present two clinical cases, an 84-year-old male and a nineteen-year-old male with clinical, immunophenotype, topography, and genotype compatible with mycosis fungoides-variety pagetoid reticulosis, both showing excellent responses to radiation therapy and in clinical remission. Despite the low frequency of this type of mycosis fungoides, we should keep it in mind in our differential diagnoses since, despite its benign course, it requires different treatment options and long-term follow-up.

Key words: Pagetoid reticulosis, Woringer–Kolopp disease, Cutaneous T-cell lymphoma, Radiotherapy

INTRODUCTION

Primary cutaneous lymphomas are a heterogeneous group of diseases of the B and T lymphoid lineage, defined as non-Hodgkin lymphomas of primary involvement to the skin without presenting manifestations in another organ at the time of diagnosis. It is reported that 75% to 80% correspond to cutaneous lymphomas of T cells. Mycosis fungoides (MF) represent 50% of all primary cutaneous lymphomas [1]. Three variants of MF have been recognized according to the 2018 classification of the World Health Organization-European Organization for Cancer Research and Treatment (WHO-EORTC): folliculotropic MF, pagetoid reticulosis, and granulomatous slack skin. Each has its clinical behavior, histological characteristics, and prognostics [2].

Pagetoid reticulosis, first described in 1939 by Woringer and Kolopp, was recognized as an entity that presents histological similarities to epidermotropic lymphoid cells with intraepidermic adenocarcinomatous cells of Paget's disease of the nipple. [3] Pagetoid reticulosis Woringer–Kolop type (localized variant), other than the disseminated type or Ketron–Goodman type, is characterized by intraepidermal proliferation of neoplastic CD3+, CD4+, CD8- or CD3+, CD4+, and CD8+, with a frequent expression of CD30+, and clinically expressed as keratotic or psoriasiform-looking solitary patches or plaques that usually affect the limbs, of slow progression, without an aggressive course. There are no reports at the time of extracutaneous involvement or deaths related to the disease [1,4].

Herein, we present two cases diagnosed at a third-level care center in Mexico City.

CASE REPORTS

Case 1

An 84-year-old male presented to the dermatology service with erythematous plaques in the interdigital

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folds of the left foot with severe, thick, adhered scaling of eight months of evolution and onychomycosis. He was first diagnosed with tinea pedis and onychomycosis and was treated with oral terbinafine and clioquinol for three months. There was an improvement in most lesions and onychomycosis. Still, a plaque persisted between the fourth and fifth fingers. The plaque had indurated edges and an infiltrated appearance with thick scales adhered in the periphery (Figs. 1a and 1b). On dermatoscopy, there were red and white areas without a structure (Fig. 1c). Dermatosis was painful, with limited walking. A biopsy of the persistent plaque was taken, with the clinical diagnosis of suspected epidermoid carcinoma.

In histology, a lichenoid infiltrate with lymphocyte epidermotropism was found, with immunohistochemical CD3 (+), CD4 (+), CD5 (-), CD7 (-), and CD8 (-), and monoclonality was demonstrated for the T lymphocyte receptor (TCR) (Figs. 2a – 2e). Extension studies were performed, in which extracutaneous activity was not evident (Figs. 3a and 3b). The patient received 8GY single-dose radiation therapy for the treatment of the interdigital lesion. There was, at two weeks, interdigital erythema and scaling with painless and gradual improvement. The patient did not have evidence of recurrence after eighteen months of follow-up.

Case 2

A nineteen-year-old male presented with hypopigmented lesions in the thenar eminence and dorsal area of the hand after ten years of evolution, with a gradual progression to form psoriasiform plaques and achromic spots in the interdigital region. An external approach was taken, and a biopsy was performed, which was

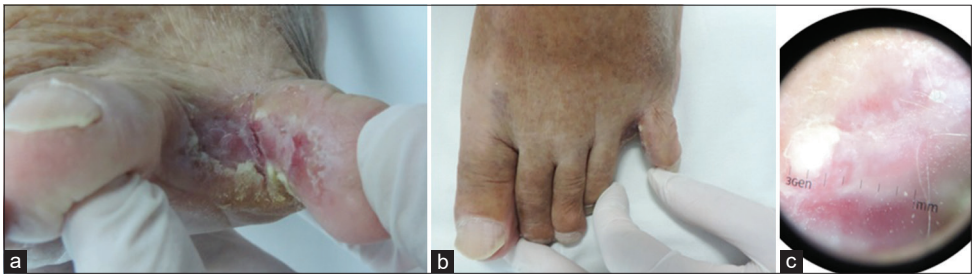


Figure 1: (a-b) Plaque with indurated edges and an infiltrated appearance with thick scales adhered to the periphery. (c) Dermatoscopy revealing red and white areas without a structure.

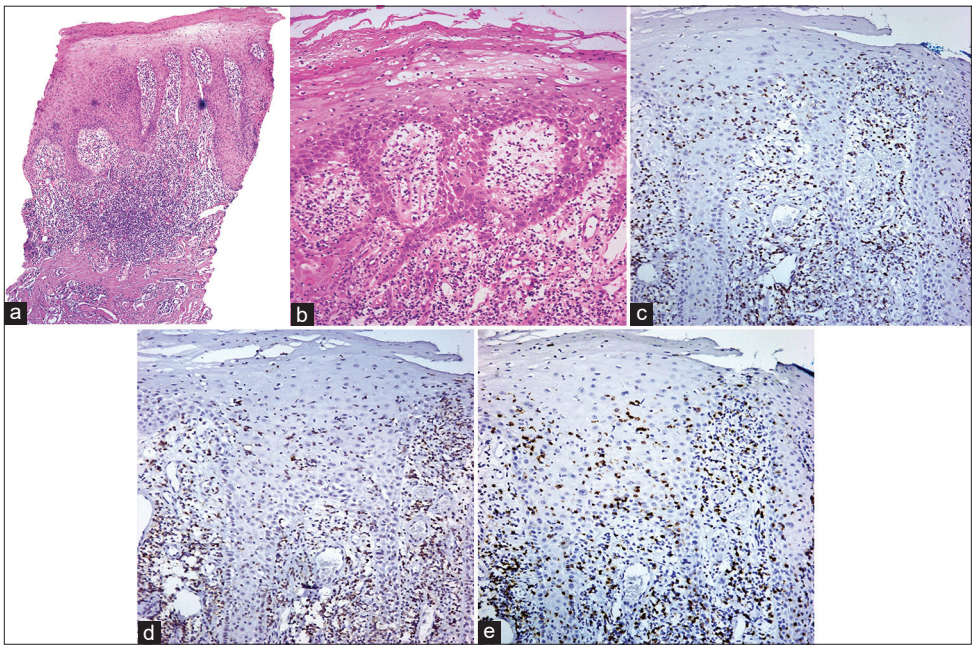
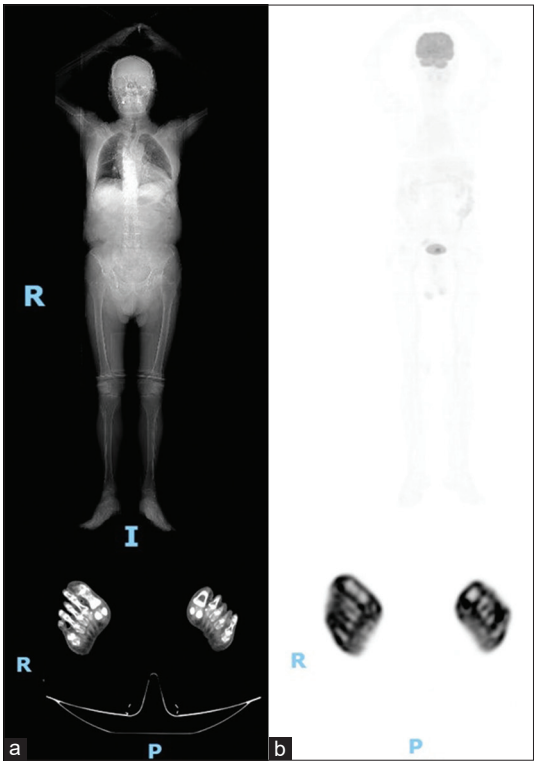


Figure 2: (a-b) Histological sections (H&E; 2x) showing a pattern of pseudoepitheliomatous hyperplasia associated with parakeratosis and vacuolar degeneration of the basal layer with necrotic keratinocytes. Lymphocytic infiltrate with a band arrangement accompanied by epidermotropism. Infiltrate consisting of irregular lymphocytes of small to medium nuclei, hyperchromatic and cerebriform. (c-e) Immunohistochemistry technique showing an immunophenotype with a predominance of positive CD8 lymphocytes (CD3; 10x).

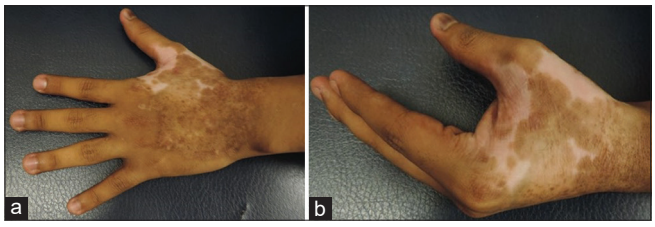
compatible with mycosis fungoides-variety pagetoid reticulosis CD3 (+), CD4 (+), and CD8 (-). In his first evaluation at our dermatology department, he presented dermatosis located on the left hand with erythematous and squamous plaques of psoriasiform appearance and perilesional achromic areas (Figs. 4a and 4b). The patient did not have extracutaneous activity. Regarding the treatment, surface radiation with electron beam therapy was performed. He received 12Gy in six fractions, presenting mild erythema after the sessions. The patient did not have evidence of recurrence after twenty-four months of follow-up.

DISCUSSION

Mycosis fungoides-variety pagetoid reticulosis has a benign behavior, with a high rate of healing and an adequate response to surgery and/or radiation therapy, yet it is advisable to follow these individuals since recurrences have been reported.



Figures 3: (a-b) Positron emission tomography without evidence of extracutaneous activity.



Figures 4: (a-b) Erythematous and squamous plaques of psoriasiform appearance and perilesional achromic areas.

Clinically, it behaves as a single asymptomatic lesion, distal to the extremities, with slow growth and well-defined edges, psoriasiform appearance, or keratose plaques [5,6]. Due to its indolent course and its unspecific clinical features, it may remain undiagnosed for years. Currently, some dermoscopy characteristics have been described and the main features have included dotted/glomerular vessels on a homogeneous pink background and white scales [7]. In a recent systematic review of 84 studies and 143 patients, 78.2% had a complete remission and 9.4% recurred [8].

For its diagnosis, it requires the sum of the clinical, histopathological, immunophenotypic, and genotypic correlation (demonstrating TCR monoclonality) [3]. More studies are needed to determine the prognosis and effective treatments for this disease [8].

CONCLUSION

Despite the low frequency of this type of mycosis fungoides, we should keep it in mind in our differential diagnoses since, despite its benign course, it presents different treatment options and requires long-term follow-up.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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