Recalcitrant acquired palmoplantar keratoderma: Think about Mycosis fungoides

Kératodermie palmoplantaire acquise récalcitrante : penser au Mycosis fungoïde

Mycosis fungoides (MF) is the most common type of cutaneous T-cell lymphoma. It has an indolent course and a wide spectrum of clinicopathological manifestations. Acquired palmoplantar keratoderma (PPK) is a rare manifestation of MF [1], which may be misdiagnosed as common benign dermatoses like palmoplantar psoriasis or eczema.

We report four cases (3 women and 1 man) who presented an acquired palmoplantar keratoderma initially diagnosed and treated suboptimally for various palmoplantar dermatitis until histopathologic and immunohistochemical findings revealed Mycosis fungoides. The average age of our patients was 62 years. The mean onset before diagnosis was 2 years. The KPP was the first manifestation of MF in the four cases. All patients had initially limited skin involvement without evidence of extracutaneous involvement (Table 1) (figures 1-5).

Discussion

Acquired PPK is an uncommon feature of MF, with the exception of erythrodermic MF and Sezary syndrome. Involvement limited to, primarily or predominantly affecting the palmoplantar surfaces is rare and was reported only in 0.6% of MF cases [1]. Indeed, Resnik et al. found in their collection of 722 MF cases only four cases restricted to the acral region and coined the term MF palmaris et plantaris (MFPP) [1]. Since this publication, cases of acquired PPK as the first or only sign of MF have been infrequently reported mostly in case report format with short-term follow-up. MFPP affects twice more men than females at an average age of 52 years [2]. The clinical appearance is usually unspecific. MFPP mimics palmoplantar psoriasis, dermatophytosis, secondary syphilis, hand eczema, hyperkeratotic lichen planus, contact dermatitis and verrucae. It is quite difficult to diagnose MFPP just by its clinical features, but it may be diagnosed with histopathologic findings. The typical lesion of MF shows a band-like infiltrate of lymphocytes in the upper dermis, epidermotropism in the epidermis with the presence of atypical T-cells ranging in size from small to normal with cerebriform nuclei and Pautrier micro-abscesses. Typical immunohistochemical (IHC) staining assesses for T-cell antigen expression by evaluating CD2, CD3, CD5, and CD7. Often, the malignant lymphocytes are CD3+CD4+ and CD8- [3]. T-cell receptor (TCR) gene rearrangement analysis on lesional skin using polymerase chain reaction (PCR) may be helpful as an adjunct to the histopathologic features of early MF [2,3]. Major therapeutic options include topical steroids, psoralen-UVA phototherapy and systemic alitretinoin, among others [4]. The course of this MF subtype is usually indolent. The disease remains confined to the initial area of involvement in most cases but extension to the limbs and trunk can occur, although no extracutaneous involvement has been reported. In our cases, Mycosis fungoides was not initially considered and the PPK was suboptimally managed as palmoplantar eczema (first and second patients) or as palmoplantar psoriasis (third and fourth patient), but in front of no improvement, palmoplantar biopsies were realized and the diagnosis of MF was made.

Conclusion

Mycosis fungoides palmaris et plantaris is a rare expression of MF primarily or predominantly affecting the palmoplantar surfaces. Its diagnosis may not initially be considered due to its infrequency and unspecific clinical appearance, which lead to misdiagnosed especially with benign dermatoses. Therefore, in front of recalcitrant acquired palmoplantar dermatoses, it is important to practice a biopsy to establish the diagnosis.
<table>
<thead>
<tr>
<th>Cases No.</th>
<th>Age (Years)</th>
<th>Sex</th>
<th>Duration of disease (Years)</th>
<th>Sites</th>
<th>Clinical features</th>
<th>Histological features</th>
<th>Immunohistochemical findings</th>
<th>Extent evaluation</th>
<th>Treatments</th>
<th>Response</th>
<th>Recurrence</th>
<th>Follow up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>54</td>
<td>F</td>
<td>2</td>
<td>Palms + soles</td>
<td>Erythema and eczematos plaque-like lesions with scales, hyperkeratosis and fissures</td>
<td>Acanthosis and spongiosis epiderma</td>
<td>+++ +++ -</td>
<td>No visceral or lymph node invasion</td>
<td>Topical corticosteroids + MTX (15 mg/w) + local puvarterapy</td>
<td>CR</td>
<td>NR</td>
<td>Lost sight (after 4 months)</td>
</tr>
<tr>
<td>2</td>
<td>67</td>
<td>F</td>
<td>3</td>
<td>Palms + soles; Arms + legs</td>
<td>Hyperkeratotic and fissured plaques generalized pruritus, papular lesions with excoriations in arms and legs, xerosis and lichenification</td>
<td>An infiltrate of atypical lymphocytes with epidermotropism and Pautrier's microabscesses nonspecific epidermic hyperplasia</td>
<td>+++ NS -</td>
<td>No visceral or lymph node invasion</td>
<td>Topical corticosteroids + local Puvarterapy</td>
<td>CR</td>
<td>1 year later: Generalized cutaneous lesions with lymph node invasion (N3)</td>
<td>Referred to hematology department to receive a polychemotherapy</td>
</tr>
<tr>
<td>3</td>
<td>68</td>
<td>F</td>
<td>2</td>
<td>Palms + soles; Nails; Legs</td>
<td>Pruritic scaly erythematous and fissured plaques Nail dystrophy Few nummular scaly erythematous patches</td>
<td>Polychemotherapy Focal epidermotropism + Pautrier's microabscesses + lichenoid dermal infiltrate</td>
<td>+++ NS +</td>
<td>No visceral or lymph node invasion</td>
<td>Topical corticosteroids + MTX (25 mg/w) + local UVbalneotherapy</td>
<td>CR</td>
<td>NR</td>
<td>2 years</td>
</tr>
<tr>
<td>4</td>
<td>59</td>
<td>M</td>
<td>1</td>
<td>Palms + soles</td>
<td>Fissural scaly diffuse keratoderma</td>
<td>Lichenoid infiltrate with epidermotropism</td>
<td>+++ +++ -</td>
<td>No visceral or lymph node invasion</td>
<td>Topical corticosteroids + MTX (20 mg/w) + local UVbalneotherapy</td>
<td>PR</td>
<td>NR</td>
<td>Lost sight (after 5 months)</td>
</tr>
</tbody>
</table>

F: female; M: male; NS: not specified; MTX: methotrexate; CR: complete regression (> 50%); PR: partial regression (< 50%); NR: no recurrence.

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**Figure 1**
Case 1: erythema and eczematous plaque-like lesions with scales, hyperkeratosis and fissures: a, b: before treatment; c, d: after 4 month of treatment

**Figure 2**
Case 2: hyperkeratotic and fissured plaques in the palms and soles
Case 3: fissural palmoplantar keratoderma with nail dystrophy and few nummular scaly erythematous patches in the legs.

Case 3: a: histology from a plantar skin biopsy showing epidermotropism of cerebriform lymphocytes and lichenoid dermal infiltrate; b: immunohistochemical findings: strong positivity of CD3+.

Case 3: improvement after 10 months of treatment.
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Disclosure of interest: the authors declare that they have no competing interest.

References


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