

cytes and melanophages in the papillary dermis was also present (figure 1E).

With the diagnosis of PNGD associated with unclassifiable autoimmune disease, the treatment consisted of rest without medication and all symptoms, including PNGD, spontaneously resolved after one week. Five years after her first visit, the patient presented again with a fever of 39 °C and a rash after sun-exposure and overwork. She displayed similar clinical features to those present at her first visit, namely, a butterfly rash-like lesion and symmetric, erythematous maculopapules and plaques on the upper arms. A biopsy specimen from a lesion on the upper arm once again showed the diagnostic features of PNGD associated with moderate interface dermatitis. However, laboratory investigations revealed leucocytopenia ( $2.3 \times 10^3/\mu\text{L}$ ), positive antinuclear antibodies (1:160) with a speckled pattern, a decreased C4 of 7 mg/dL (normal 13-35), and the presence of anti-Smith (244 U/mL, normal 0-10) and anti RNP antibodies (169 U/mL, normal 0-10), which confirmed the diagnosis of SLE. A decreased C3 level was later seen during the follow-up. The fever and rash, including recurrent PNGD, did not respond to treatment with 20 mg/day of prednisolone, but resolved with 40 mg/day.

PNGD is a rare dermatological condition which shows various clinical features [1-3]. PNGD is characteristically associated with systemic diseases [1-3], particularly autoimmune conditions. There are a few reported cases of PNGD in patients with SLE [4-6], and PNGD is considered to be a non-specific cutaneous manifestation of SLE rather than an SLE-specific cutaneous manifestation (such as a butterfly rash). The butterfly rash-like lesion, which developed twice with PNGD in this patient, is interesting. Two possibilities have been considered, namely an SLE-specific butterfly rash associated with PNGD, or PNGD clinically manifesting as a butterfly rash-like lesion. As no skin biopsy was taken from the face lesion, it is impossible to distinguish between these two possibilities. It is interesting that the recurrent PNGD on the upper arm showed an association histopathologically with interface dermatitis, demonstrating that the lesions in this patient developed under combined (PNGD and SLE) pathogenic pathways. Although specimens of the recurrent PNGD were not available for immunofluorescence staining in this case, a previous case of PNGD associated with SLE demonstrated linear deposits of C3 at the basement membrane [4].

Whether the appearance of PSGD in an autoimmune disease is associated with a worse therapeutic response and/or a poor prognosis has not yet been clarified [4-6]. This case was notable for the presence and continued activity of an underlying disease, which was finally clearly identified as SLE. ■

**Acknowledgements.** Financial support: none. Conflict of interest: none.

<sup>1</sup>Dermatology, Department of Internal Medicine, Faculty of Medicine, Saga University, Nabeshima 5-1-1, Saga 849-8501, Japan

<sup>2</sup>Rheumatology, Department of Internal Medicine, Faculty of Medicine, Saga University, Saga, Japan  
<misago@post.saga-med.ac.jp>

Noriyuki MISAGO<sup>1</sup>  
Hisako INOUE<sup>1</sup>  
Takuya INOUE<sup>2</sup>  
Kohei NAGASAWA<sup>2</sup>  
Yutaka NARISAWA<sup>1</sup>

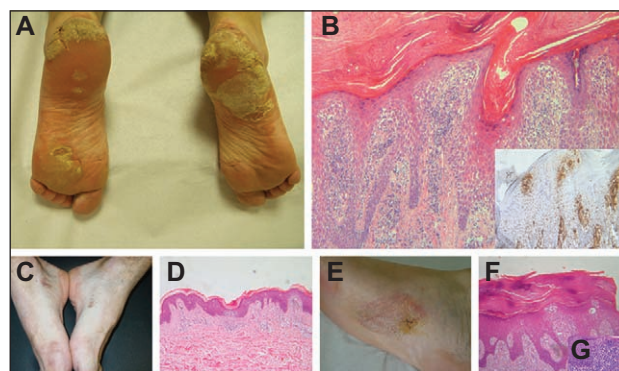
1. Chu P, Connolly MK, LeBoit PE. The histopathologic spectrum of palisaded neutrophilic and granulomatous dermatitis in patients with collagen vascular disease. *Arch Dermatol* 1994; 130: 1278-83.
2. Sanguenza OP, Caudell MD, Mengesha YM, Davis LS, Barnes CJ, Griffin JE, *et al.* Palisaded neutrophilic granulomatous dermatitis in rheumatoid arthritis. *J Am Acad Dermatol* 2002; 47: 251-7.
3. Hantash BM, Chiang D, Kohler S, Fiorentino D. Palisaded neutrophilic and granulomatous dermatitis associated with limited systemic sclerosis. *J Am Acad Dermatol* 2008; 58: 661-4.
4. Obermoser G, Zelger B, Zangerle R, Sepp N. Extravascular necrotizing palisaded granulomas as the presenting skin sign of systemic lupus erythematosus. *Br J Dermatol* 2002; 147: 371-4.
5. Germanas JP, Mehrabi D, Carder KR. Palisaded neutrophilic granulomatous dermatitis in a 12-year-old girl with systemic lupus erythematosus. *J Am Acad Dermatol* 2006; 55: S60-2.
6. Blaise S, Salameire D, Carpentier PH. Interstitial granulomatous dermatitis: a misdiagnosed cutaneous form of systemic lupus erythematosus? *Clin Exp Dermatol* 2008; 33: 712-4.

doi:10.1684/ejd.2010.0822

## Mycosis fungoides plantaris

Mycosis fungoides palmaris en plantaris (MFPP) is a variant of mycosis fungoides (MF) that presents primarily on the palms and soles [1], and it has been infrequently described. Herein we report three new cases of MF plantaris, each with a different clinical form.

Case 1. A 63 year-old female presented a 1 year history of multiple hyperkeratotic and fissured plantar plaques (figure 1A). She was treated for plantaris hyperkeratosis resembling keratoderma with topical corticosteroids and keratolytics, and oral acitretin, with partial response. One 4 mm punch biopsy specimen was obtained from



**Figure 1.** A) Soles showing multiple nonerythematous, hyperkeratotic and fissured plantar plaques. B) The skin biopsy shows psoriasiform epidermal hyperplasia and atypical lymphocytes with cerebriform nuclei. The immunohistochemical study showed a marked predominance of CD4. C) Soles showing some brownish and purple plaques, a little desquamative. D) The skin biopsy shows a psoriasiform epidermal hyperplasia and focal epidermotropism of cerebriform lymphocytes and Pautrier's microabscesses. E) The right sole shows a solitary eczematous plantar plaque. F) The biopsy specimen from the lesion shows psoriasiform epidermal hyperplasia, a bandlike infiltrate containing numerous cerebriform lymphocytes in the papillary dermis, and a single Pautrier's microabscess, which is showed in G), with higher magnification.

the right foot, and it showed psoriasiform epidermal hyperplasia and atypical lymphocytes with cerebriform nuclei (figure 1B). The immunohistochemical study showed positivity for CD4 (> 70%).

Case 2. A 25-year-old male presented some brownish and purple plaques, a little desquamative, on both feet (figure 1C). A punch biopsy from the right foot was diagnostic for MF plantaris. The skin biopsy showed a psoriasiform epidermal hyperplasia and focal epidermotropism of cerebriform lymphocytes and Pautrier's micro-abscesses in some sections (figure 1D). Immunohistochemical findings showed the infiltrate to be composed mostly of CD4.

Case 3. A 43-year-old female presented a 2-year history of an eczematous plantar plaque. It was pruritic and had not responded to emollients and topical keratolytics (figure 1E). A biopsy specimen from the lesion showed psoriasiform epidermal hyperplasia, a band-like infiltrate containing numerous cerebriform lymphocytes in the papillary dermis, and a single Pautrier's micro-abscess (figures 1F, G). The immunohistochemical study showed a marked predominance of CD4.

In all three cases a diagnosis of MF plantaris was established. No other cutaneous involvement was observed, besides the plantar lesions. In addition, extension studies were negative in all patients. The first case is currently receiving treatment with re-PUVA (acitretin 25 milligrams per day, and topical 8-metoxypsoralen and UVA), with a poor response after three months of treatment. The second case presented clinical remission after two months of treatment with topical clobetasol propionate 0.05% and oral acitretin 25 mg/day. The third case also presented a complete response with topical clobetasol propionate 0.05% after six months of treatment. The last two cases have had a follow-up period of almost 4 years.

MFPP is infrequent, about 0.6% of all MF [2]. Clinically, MFPP mimics many other dermatoses. Cutaneous lesions may present as multiple or single lesions, scaly and erythematous patches, hyperkeratotic and dyshidrotic plaques [3, 4]. A wide clinical spectrum is the reason why diagnosis of this entity is often delayed. When the differential diagnosis is difficult, T cell-receptor (TCR) gene rearrangement study is one of the most useful methods for diagnosing MF [1]. In addition, clinical and histological findings should be considered.

It is important to distinguish it from palmo-plantar involvement in the course of a generalized MF, which is not uncommon. The histopathological findings in a skin biopsy are indistinguishable between these two entities. In single lesions, the most important differential diagnosis is Woringer-Kolopp disease, which is characterized by the presence of one or several scaly patches and plaques with acral distribution. It is a benign T-cell lymphoproliferative process with certain histological and phenotypical similarities both to early epidermotropic mycosis fungoides-type cutaneous T-cell lymphoma and other T-cell lymphoproliferations.

In MFPP, various therapeutic modalities have been described with PUVA [5], narrow band UVB [5], excision of solitary lesions [5], high potency topical corticoids and intralesional corticoids, methotrexate [5], bexarotene [5], radiotherapy [6], and electron beam therapy [6], CO2 laser and topical nitrogen mustard [5].

This form of MF typically has a long benign course [4], no case of extracutaneous spread has been described [5].

A skin biopsy should be included in the study of all recalcitrant palmo-plantar dermatoses, and MFPP considered in the differential diagnosis. ■

**Acknowledgements.** Financial support: none. Conflict of interest: none.

Dept of Dermatology, Dermatology  
Univesitary Hospital Sagrat cor,  
Viladomat street 288,  
08029 Barcelona, Spain  
<40871gmb@comb.cat>

**Gemma  
MÁRQUEZ BALBÁS  
Angeles SOLA CASAS  
Maribel  
IGLESIAS SANCHO  
Manuel  
SÁNCHEZ-REGAÑA  
Loida  
GALVANY ROSSELL  
Elisabet  
DILMÉ CARRERAS  
Joaquim SOLA  
ORTIGOSA  
Pablo UMBERT  
MILLET**

1. Kim ST, Jeon YS, Sim HJ, *et al.* Clinicopathologic features and T-cell receptor gene rearrangement findings of mycosis fungoides palmaris et plantaris. *J Am Acad Dermatol* 2006; 54: 466-71.
2. M. Fernández-Guarino R. Carrillo-Gijón M. Fernández-Lorente C. García-Millán E. Muñoz-Zato y P. Jaén-Olasolo. Placa eritematoescamosa plantar. *Actas Dermosifiliogr* 2007; 98: 207-8.
3. Masmoudi S. Bouassida, A. Khabir, M. Loukil, J. Wechsler, M. Bagot, T. Boudawara, H. Turki, A. Zahaf. Plaques érythémato-squameuses palmo-plantaires. *Ann Dermatol Venereol* 2006; 133: 715-6.
4. Spieth K, Grundmann-Kollmann M, Runne U, *et al.* Mycosis fungoides-type cutaneous T cell lymphoma of the hands and soles: a variant causing delay in diagnosis and adequate treatment of patients with palmoplantar eczema. *Dermatology* 2002; 205: 239-44.
5. Laguna C, Pérez-Farriols A, Martín B. Alegre. V. Placas hiperqueratóticas palmoplantares. *Actas Dermosifiliogr* 2007; 98: 565-6.
6. Resnik KS, Kantor GR, Lessin SR, *et al.* Mycosis fungoides palmaris et plantaris. *Arch Dermatol* 1995; 131: 1052-6.

doi:10.1684/ejd.2010.0830

## Primary cutaneous extranodal NK/T-cell lymphoma, nasal type, in an adolescent

A 15-year-old girl was admitted to our hospital in April 2002, with a nearly two year history of repeated intense skin reactions to insect and mosquito-bites. Each rash began with asymptomatic edematous erythemas on the face and lower extremities, some lesions became bullae and sometimes evolved into painful ulcers. The lesions self-cured after 4-5 days with pigmentation and scars remaining. No fever, malaise, or other systemic symptoms were noticed. At the time of the first admission, physical examination showed marked swelling on the right face and upper lip, multiple edematous erythemas, pigmentation and scars scattered over the face, trunk and extremities. A central-crusted nodule and centrifuged enlarged edematous erythemas with tiny scaling on the upper extremities were also noticed (figures 1A, B). Enlarged lymph nodes were not present on palpation. Routine lab-