Persistent Acantholytic Dermatosis: Sex-related Differences in Clinical Presentation?

MOURAD MOUKNI¹, SÉLIM ARACTINGL, RACHEL GROSSMAN¹, OLIVIER VEROLA², SERGE LETESSIER³, JEAN CIVATTE¹ and LOUIS DUBERTRET¹

¹Clinics for skin diseases and ²Dermato-Pathology Department, Hôpital Saint-Louis, Paris, France

We report the case of a 58-year-old man with a chronic papular eruption of 10 years’ duration. Histopathology revealed focal acantholytic dyskeratosis. This condition is thought to represent a distinct entity which has been reported under several names and frequently referred to as persistent acantholytic dermatosis. The relationship between this condition and transient acantholytic dermatosis (Grover's disease), emphasizing sex-related presentations, is discussed.

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Sélim Areacting, Clinique des maladies cutanées, Hôpital Saint-Louis, 1, Avenue Claude Vellefaux, 75010 Paris, France.

Transient acantholytic dermatosis is a primary acantholytic disease, first described by Grover in 1970 (1). Skin signs are predominantly self-limited papulo-vesicles which are present for a few months. The disease occurs more commonly in men past the age of 50. The etiology of the disease is still unknown, but most authors relate the onset of the eruption to sun exposure. Since 1970, there have been many reports of male patients with similar presentations; however, the disease persisted for several months. We report here the case of a patient with a persistent acantholytic dermatosis probably related to actinic damage. The disease responded well to treatment with acitretin. A review of the literature revealed a male predominance in the chronic forms of these acantholytic diseases.

CASE REPORT
A 58-year-old man presented with a recurrent eruption of asymptomatic erythematous papular lesions on the arms, forehead and upper trunk (Fig. 1). This condition had been present for the past 8 years. It had first appeared on the face, and the patient reported that exposure to sunlight had been an exacerbating factor. The patient was of fair complexion and had received considerable sun exposure during his service in the army. This exposure had resulted in extensive solar damage including actinic keratoses, multiple squamous and basal cell carcinoma lesions and Bowen's disease. A biopsy of persistent papulo-vascular lesions was performed. Histopathological examination revealed focal areas of acantholysis at different levels in the epidermis. Dyskeratotic cells could be seen in the mid-epidermis (Fig. 2). Direct immunofluorescence did not show immune complex or complement deposition. Results of routine laboratory studies were normal. Treatment with acitretin (0.5 mg/kg/day) was instituted. Six months after the treatment had begun, 89% of the lesions had disappeared.

DISCUSSION
Non-autoimmune and non-hereditary acantholytic dermatoses were described by Grover & Ackerman (1,2). They were characterized by pruritic papules and vesicles displaying the coexistence of both acantholytic and dyskeratotic cells at different levels of epidermis. These cases were transient with quick disappearance of lesions (1–3). Simon (4) presented in 1976 the case of a male patient, who had clinical and pathological features of Grover's disease yet with lesions persisting for 3 years. They called this condition "persistent acantholytic dermatosis". Since then, there have been other reports under the names of "chronic benign acantholytic dermatosis" (5) and "persistent acantholytic dermatosis" (6–8). All these patients were middle-aged men who developed multiple, chronic, pruritic papular lesions on the upper trunk (Table 1). Histopathological examination disclosed focal acantholysis with varying degrees of dyskeratosis and suprabasal clefting. In most of the cases, sun exposure was believed to initiate or

Fig. 1. Papular lesions of the trunk.

Fig. 2. Skin section showing presence of pseudo-acantholytic clefts through epidermis associated to dyskeratotic cells (HES ×200).

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exacerbate the eruption. The major distinguishing factor between these patients and those reported by Grover was the duration of the dermatosis. However, Chalet et al. (9) reviewed later “transient acantholytic dermatosis” in 54 patients. They noted that although the disease cleared within 3 months in the majority of patients, it persisted in some of them for up to 2 or 3 years.

Our patient is very similar to previous case reports (7, 8). The presence of extensive solar damage and the frequent occurrence of skin malignancies suggest that chronic actinic damage may be an important precipitating factor. It is different from Darier’s disease, particularly because cleavage and dyskeratosis had a superficial pattern and filiform papillomatosis was not observed. Finally, the absence of epidermal dysplasia and the eruptive nature of the lesions differentiate them from Darier-like actinic keratosis (7). Retinoids seemed to improve this condition remarkably and to prevent the occurrence of further lesions in our patient.

Review of the literature showed that nearly all disseminated persistent acantholytic dermatoses reported until now occurred in men (Table I). There have been several reports of persistent papular eruption with acantholysis and dyskeratosis in female patients; however, these were always restricted to the vulvo-crural region (Table II). Authors first questioned whether these cases could be a distinct entity (10). However, the report by Van Joost et al. (13) of the case of a female patient with a persistent acantholytic eruption on the chest and the vulva led these authors to consider vulvar lesions similar to previously reported cases in male patients.

The predominance of lesions in sun-exposed areas in men may be due in part to the fact that men work more frequently out-of-doors and consequently are more susceptible to solar-induced skin damage. Clinically similar lesions with similar histopathology have been reported in female patients in non-sun-exposed areas. Environmental and possibly genetic factors may ultimately be responsible for the sex-related differences in clinical presentation.

### Table I. Persistent acantholytic dermatosis in male patients

<table>
<thead>
<tr>
<th>Classification</th>
<th>No. of patients</th>
<th>Age</th>
<th>Duration</th>
<th>Predominant location</th>
<th>Long UV exposure</th>
</tr>
</thead>
<tbody>
<tr>
<td>Heaphy et al. (3)</td>
<td>7</td>
<td>41-63</td>
<td>Persistent</td>
<td>Trunk</td>
<td>+</td>
</tr>
<tr>
<td>Simon et al. (4)</td>
<td>1</td>
<td>82</td>
<td>Persistent</td>
<td>Trunk</td>
<td>-</td>
</tr>
<tr>
<td>Yosmik et al. (5)</td>
<td>1</td>
<td>55</td>
<td>Persistent</td>
<td>Trunk</td>
<td>-</td>
</tr>
<tr>
<td>Rocha et al. (6)</td>
<td>1</td>
<td>43</td>
<td>Persistent</td>
<td>Trunk</td>
<td>-</td>
</tr>
<tr>
<td>Fawcett et al. (7)</td>
<td>10</td>
<td>38-65</td>
<td>Persistent</td>
<td>Trunk</td>
<td>+</td>
</tr>
<tr>
<td>Ott et al. (8)</td>
<td>1</td>
<td>72</td>
<td>Persistent</td>
<td>Trunk</td>
<td>+</td>
</tr>
<tr>
<td>Van der Putte et al. (14)</td>
<td>1</td>
<td>34</td>
<td>6 years</td>
<td>Penis</td>
<td>-</td>
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</tbody>
</table>

### Table II. Persistent vulvo-crural acantholytic dermatosis in female patients

<table>
<thead>
<tr>
<th>Classification</th>
<th>No. of patients</th>
<th>Age (months)</th>
<th>Duration (months)</th>
<th>Other localization</th>
</tr>
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<tr>
<td>Chorzeck et al. (10)</td>
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<td>72</td>
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<td>Coppola et al. (11)</td>
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<td>44</td>
<td>36</td>
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<tr>
<td>Cooper et al. (12)</td>
<td>6</td>
<td>28-83</td>
<td>12-36</td>
<td>No</td>
</tr>
<tr>
<td>Van Joost Th et al. (13)</td>
<td>1</td>
<td>35</td>
<td>36</td>
<td>Yes</td>
</tr>
</tbody>
</table>

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REFERENCES