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SHORT REPORT

Position Statement: Linear prurigo is a subtype of chronic prurigo

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Abstract

Background Chronic prurigo (CPG) is a distinct disease characterized by chronic pruritus, history and/or signs of prolonged scratching and multiple pruriginous lesions. It may present with various clinical manifestations, including papules, nodules, plaques or umbilicated lesions. Some patients with chronic pruritus show pruriginous linear and scaring scratch lesions (LSSL) and it is unclear whether these lesions belong to the spectrum of CPG.

Objective To achieve a consensus on the classification of pruriginous LSSL and establish criteria to differentiate them from similar appearing conditions of different nature.

Methods Members of the Task Force Pruritus (TFP) of the European Academy of Dermatology and Venereology participated in the consensus conference, discussing representative clinical cases. Using the Delphi method, consensus was reached when ≥75% of members agreed on a statement.

Results Twenty-one members of the TFP with voting rights participated in the meeting. It was consented that LSSL occurs due to chronic pruritus and prolonged scratching, and share common pathophysiological mechanisms with CPG. LSSL were thus considered as belonging to the spectrum of CPG and the term 'linear prurigo' was chosen to describe this manifestation.

Conclusion Considering linear prurigo as belonging to the spectrum of CPG has important clinical implications, since both the diagnostic and therapeutic approach of these patients should be performed as recommended for CPG. Importantly, linear prurigo should be differentiated from self-inflicted skin lesions as factitious disorders or skin picking syndromes. In the latter, artificial manipulation rather than pruritus itself leads to the development of cutaneous lesions, which can show clinical similarities to linear prurigo.

[†]Further members of the Task Force Pruritus group are listed in Appendix.
All authors and further members of the Task Force Pruritus group have equally contributed.

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Conflicts of interest

MPP, CZ, TN, AWME, SB, SG, JAH, JL, FJL, TL, MM, LM, KN, AR, GS, HS, MS, JW, EW and SST declare that they have no conflict of interests. MG declares no conflict of interests for this manuscript but has participated in advisory boards from Novartis and Sanofi-Genzyme. JCS is a consultant and advisor for AbbVie, Celgene, Dignity Sciences, Leo Pharma, Novartis, Pierre-Fabre, Menlo Therapeutics, Sienna Pharmaceuticals and Sandoz, an investigator for AbbVie, Actelion, Amgen, GSK, Janssen, Merck, Novartis, Regereron, Takeda, Trevi and a speaker for AbbVie, Actavis, Astellas, Janssen, Leo Pharma, Novartis, SunFarm, Sandoz, Eli Lilly.

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Introduction

The terminology of chronic prurigo (CPG) conditions has been confusing over the years with a number of prurigo-associated terms without a clear definition.[1] Addressing this issue, the Task Force Pruritus (TFP) of the European Academy of Dermatology and Venereology achieved a consensus on the nomenclature and classification of CPG.[2] CPG was considered a distinct disease characterized by chronic pruritus, history and/or signs of prolonged scratching and multiple localized or generalized pruriginous lesions.[2] Of note, CPG may present with different clinical manifestations, including papules, nodules, plaques or umbilicated lesions.[2,3]

In some instances, patients with chronic pruritus present with pruriginous but linear arranged and/or scarring scratch lesions (LSSL; Fig. 1) due to scratching. One open point of discussion is how to categorize LSSL, whether they belong to the spectrum of CPG and whether these lesions can be differentiated from self-inflicted skin lesions (SISL). This article provides an expert consensus on the classification and diagnostic criteria of LSSL and their demarcation to other similar appearing conditions of different nature.

Material and methods

All members of the TFP were invited for a consensus conference taking place in Brussels, Belgium on 9 April 2018, to discuss whether LSSL should be considered as part of the spectrum of CPG and how the terminology could be adopted. First, representative clinical cases were presented and discussed among the group, including possible differential diagnoses. According to the Delphi method,[4] each member voted anonymously choosing either 'I agree' or 'I do not agree' to pre-selected questions using a televoting system. There were no abstentions. Consensus was reached when 75% or more participants agreed on a statement.

Results

Twenty-one members of the TFP with voting rights participated in the meeting. Voting results are shown in Table 1. TFP members agreed that chronic pruritus underlies LSSL, and that LSSL occur due to prolonged scratching, and may appear in localized areas or occur generalized over the whole integument, sharing thus characteristics with papules, nodules, plaques or umbilicated lesions belonging to the spectrum of CPG. In fact, TFP members consented that LSSL show common pathophysiological mechanisms to CPG and may overlap with CPG lesions in the same individual. As a result, LSSL were considered as belonging to the spectrum of CPG by a vast majority of the participants (90%), being a further subtype of this condition (Fig. 2). The term 'linear prurigo' was chosen to describe this manifestation. Finally, TFP members agreed that linear prurigo should be differentiated from skin picking syndromes, in which artificial manipulation, rather than pruritus itself and subsequent scratching, leads to the development of the lesions.

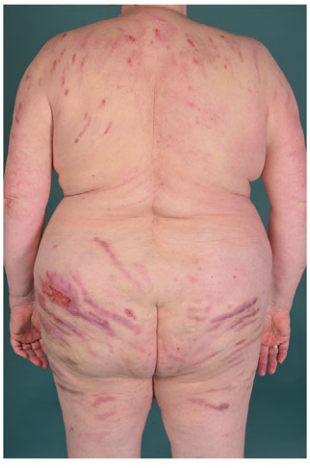

Figure 1 Patient with representative linear prurigo lesions.

Table 1 Results of the Delphi process on the terminology of linear prurigo

Question	Voting results
Q1: Do you agree that chronic pruritus underlies linear scarring scratch lesions?	**I agree: 90%** I do not agree: 10%
Q2: Do you agree that linear scarring scratch lesions occur due to prolonged scratching?	**I agree: 90%** I do not agree: 10%
Q3: Do you agree that linear scarring scratch lesions can be localized or generalized?	**I agree: 100%** I do not agree: 0%
Q4: Do you agree that linear scarring scratch lesions and chronic prurigo may share similar pathophysiological mechanisms?	**I agree: 90%** I do not agree: 10%
Q5: Do you agree that linear scarring scratch lesions and other subtypes of chronic prurigo may overlap in the same patient?	**I agree: 95%** I do not agree: 5%
Q6: Do you agree that linear scarring scratch lesions due to chronic pruritus belong to the spectrum of chronic prurigo?	**I agree: 90%** I do not agree: 10%
Q7: Do you agree with the term Linear Prurigo?	**I agree: 81%** I do not agree: 19%
Q8: Do you agree that linear prurigo should be classified as the fifth subtype of chronic prurigo.	**I agree: 81%** I do not agree: 19%
Q9: Do you agree that skin picking syndromes and linear prurigo are different entities, since the underlying pathophysiological mechanisms are different?	**I agree: 76%** I do not agree: 24%

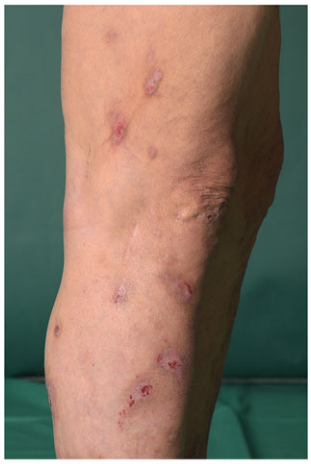

Figure 2 Linear prurigo: a subtype of chronic prurigo. Chronic prurigo may present with various clinical manifestations, including papular, nodular, plaque-type, umbilicated and linear lesions. These share common core symptoms and associated criteria and belong thus to the spectrum of chronic prurigo. A representative chronic prurigo lesion of nodular type is shown.

Discussion

Patients with chronic pruritus presenting with LSSL are rare, but experience a highly onerous pruritus and associated burdens, including a substantial impairment of their quality of life. LSSL are considered to belong to the spectrum of CPG, since patients with LSSL share pathophysiological and clinical similarities to CPG patients. As in other subtypes of CPG, in linear prurigo an ongoing itch-scratch cycle and sensitization mechanisms are believed to play a central role leading to perpetuation of the condition.[5] As for the clinical presentation, linear prurigo patients suffer from chronic pruritus, present with pruriginous lesions in similar distribution to CPG and are affected by the same associated factors as in CPG (e.g. continued pruritus of high intensity, accompanying paresthesias and alloknesis, sleep impairment, concomitant anxiety and depression and overall impairment of the quality of life).[2] Moreover, CPG nodules or papules may coexist with linear prurigo lesions in the same patient, but linear prurigo lesions may be the single lesional type in a patient. Linear prurigo seems to result from a different scratching pattern when compared to other subtypes of CPG. However, it remains unclear which factors contribute to the discrepant scratching behaviours.

The underlying pruritus in linear prurigo may have various possible etiologies, including psychological origins. Therefore, self-inflicted skin lesions (SISL) represent an important differential diagnosis of linear prurigo (Fig. 3). As defined by the European Society for Dermatology and Psychiatry (ESDaP), the term SISL should be applied to skin lesions, which are the consequence of pathological skin manipulation due to underlying psychological factors. SISL are further categorized into those in which the person denies his/her behaviour and those in which the behaviour is not denied, but is compulsive or impulsive.[6] Patients with linear prurigo normally do not deny their scratching (as in malingering or factitious disorders). In linear prurigo, the scratching behaviour is usually not compulsive or impulsive, in most patients it is a reaction to chronic pruritus, while in skin picking and related SISL the respective behaviour is executed without severe pruritus.[6,7] Therefore, underlying pruritus is essential in the pathophysiology of linear prurigo. Of course in some patients with pruritus (and also with linear prurigo), the scratching can develop into an obsessive, compulsive or impulsive behaviour,

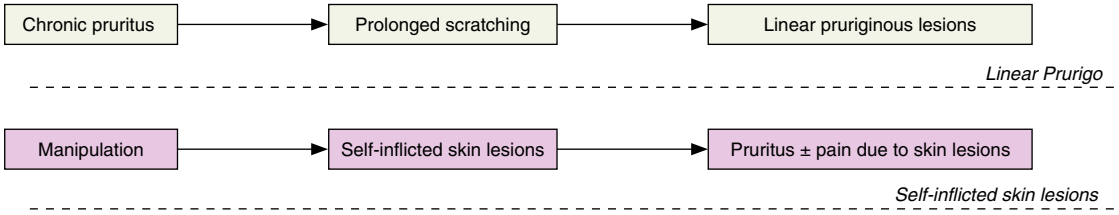

Figure 3 Demarcation of linear prurigo from self-inflicted skin lesions. Self-inflicted skin lesions and linear prurigo are different entities, which need to be differentiated. In linear prurigo, chronic pruritus induces a prolonged scratching behaviour, which eventually leads to the development of the typical linear lesions, while in self-inflicted skin lesions manipulation without underlying pruritus induces the occurrence of factitious lesions, which may themselves be itchy and/or painful.

which they regularly execute even when no itch is present. In this subgroup of patients with linear prurigo, the additional diagnosis of obsessive-compulsive or impulsive disorder may be justified. Also the differential diagnosis to factitious disorders can be very difficult, if patients pretend to suffer from pruritus.

Since psychiatric comorbidity such as anxiety and depression may coexist in linear prurigo patients, the differentiation from SISL may be difficult. A detailed medical history, physical examination and the assessment of eventual coexisting mood disorders are necessary to allow such distinction. Experts of the TFP considered that SISL often have a different clinical appearance in comparison with linear prurigo, as they are often not symmetrically distributed and not as homogenous in appearance as linear prurigo lesions.

Categorizing linear prurigo within the spectrum of CPG has important clinical implications. Both the diagnostic workup of these patients and the therapeutic measures to be initiated should be performed as recommended for CPG.[8] Additionally, patients with linear prurigo should be able to qualify for randomized clinical trials including CPG patients, enabling the access of these patients to possibly more promising agents in the treatment of this refractory condition.

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