Facial Granular Parakeratosis: The First Report of Combined Eccrine and Follicular Involvement

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ABSTRACT:
Granular parakeratosis is a rare cutaneous disorder that typically affects intertriginous areas such as the axillae and groin. Non-intertriginous involvement is uncommon, and facial presentations are particularly rare. We report a case of a 40-year-old Thai woman with a two-year history of recurrent, pruritic, brownish, hyperkeratotic papules involving the face, neck, anterior chest, and upper back, without axillary or other typical flexural involvement. The lesions were aggravated by heat and sweating. Histopathological examination revealed focal compact parakeratosis with retained basophilic keratohyalin granules, extending into both the eccrine ostia and follicular infundibula, confirming the diagnosis of granular parakeratosis. Based on the available literature, this appears to be the first reported case of combined eccrine and follicular involvement. After a one-month follow-up, the patient showed marked improvement with topical corticosteroids and lifestyle modifications to reduce heat exposure.
Key words: granular parakeratosis, hyperkeratotic papules, keratinization

Introduction

Granular parakeratosis is an uncommon, acquired disorder of keratinization that typically presents as erythematous to brownish, scaly, hyperkeratotic papules coalescing into plaques[1,2]. Lesions may be bilateral or unilateral, and some patients experience pruritus. The condition predominantly affects intertriginous areas, most commonly the axillae, but involvement of non-intertriginous sites is rare.

Case report

A 40-year-old Thai woman presented with a two-year history of recurrent pruritic, hyperkeratotic brownish papules on the face, neck, and upper trunk. The initial episode began two years earlier, with small brownish papules localized to the neck. Lesions were aggravated by heat and sweating, resolved spontaneously within three months, and were not associated with new skincare products. Approximately six months before presentation, the rash recurred in summer with a more extensive distribution involving the face, neck, anterior chest, and upper back. The lesions were intensely pruritic. She was diagnosed with heat rash at a private clinic and prescribed an unknown topical medication without improvement.

Physical examination revealed multiple brownish, scaly to hyperkeratotic papules on the face, neck, upper chest, and upper back, sparing the axillae and inguinal areas. Oral mucosa and nails were normal (Figure 1). A skin biopsy from the posterior neck showed mild epidermal acanthosis with focal compact parakeratosis and retention of basophilic keratohyalin granules in the stratum corneum, extending into both the eccrine ostia and follicular infundibula, accompanied by a sparse superficial perivascular lymphocytic infiltrate. (Figure 2)

Based on clinical and histopathological findings, a diagnosis of granular parakeratosis was made. The patient was treated with topical triamcinolone acetonide 0.02% cream twice daily and advised to avoid heat and other potential triggers. At a one-month follow-up, clinical improvement was observed.

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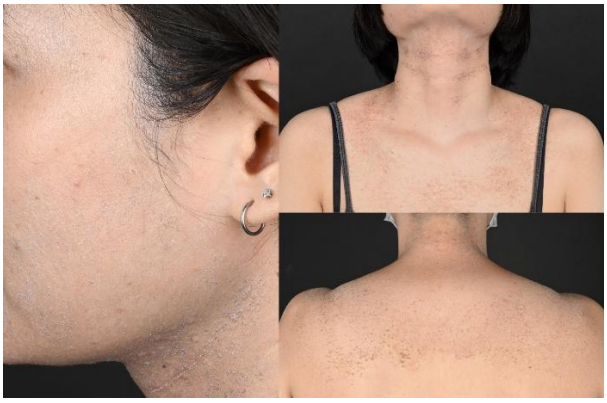

Figure 1 The clinical presentations showed multiple brownish, scaly to hyperkeratotic papules on the face, neck, upper chest, and upper back

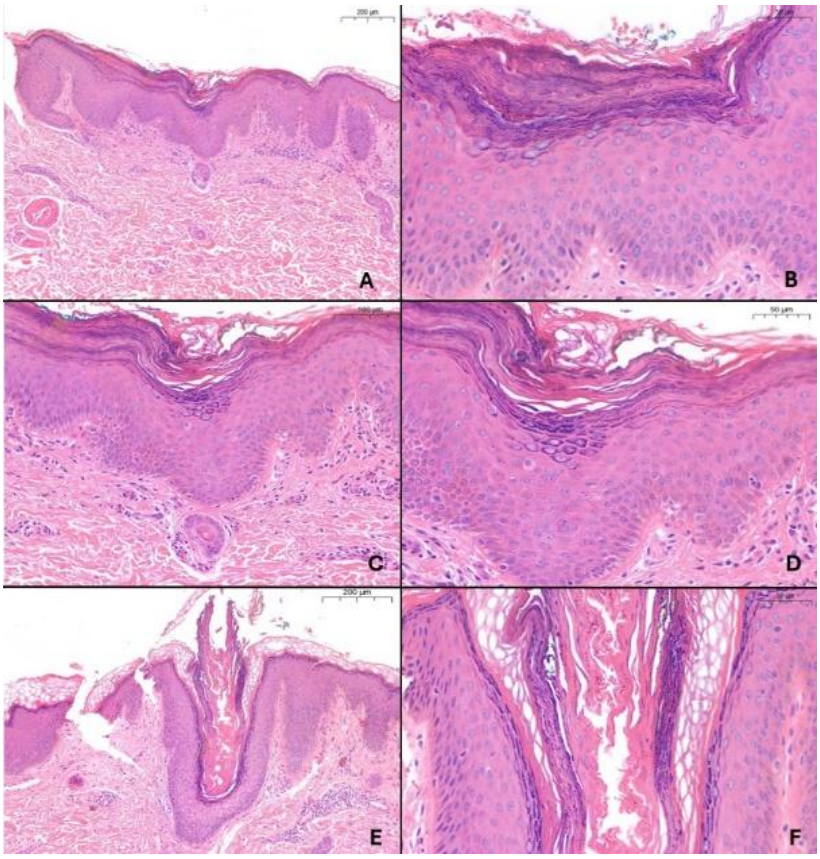

Figure 2 Histopathology demonstrates mild epidermal acanthosis with focal compact parakeratosis and retention of basophilic keratohyalin granules in the stratum corneum (A, B; H&E, ×100 and ×400). Granular parakeratosis extends into eccrine ostia (C, D; H&E, ×200 and ×400) and follicular infundibula (E, F; H&E, ×100 and ×400), accompanied by a sparse superficial perivascular lymphocytic infiltrate

Discussion

Granular parakeratosis is a rare keratinization disorder that can occur at any age and in both sexes, but it is more common in women.[1] It was first described by Northcutt et al.[3] in 1991 as "axillary granular parakeratosis." Subsequently, multiple cases involving other body sites beyond the axillae, such as the groin and abdomen, have been reported, leading to the adoption of the broader term "granular parakeratosis."

The exact pathogenesis of granular parakeratosis remains unclear, although aggravating factors have been identified in over half of reported cases.[1] The condition is thought to represent a contact reaction, either irritant or allergic, often triggered by topical products such as antiperspirants and deodorants[4]. This may lead to defective profilaggrin-to-filaggrin processing during keratinocyte cornification, resulting in impaired degradation of keratohyalin granules and abnormal keratin aggregation[5].

Intertriginous areas are the predominant sites of involvement, with the axillae being the most frequently affected, followed by the groin and infra-/intramammary regions. Non-intertriginous involvement is rare, occurring mostly in the anogenital region, torso, and thighs.[1] To date, only three cases in the English literature have reported facial lesions[6-8].

1). A 27-year-old woman from India presented with a unilateral cheek lesion, associated with repeated wiping in a hot environment. Histopathological examination revealed granular parakeratosis involving both the epidermis and follicular infundibulum.

2). A 7-year-old Chinese girl had lesions spreading from the neck to the face, particularly affecting the forehead and eyelids; no aggravating factors were identified, but the authors suggested a possible role of hot weather. Histopathology demonstrated granular parakeratosis confined to the eccrine ostia.

3). A 52-year-old Thai man developed follicular granular parakeratosis on the face after using a topical anti-melasma cream. Histopathological findings showed granular parakeratosis limited to the follicular infundibulum.

We herein report a fourth case, involving a 40-year-old Thai woman with hyperkeratotic papules involving the face, neck, upper chest, and upper back, aggravated by sweating.

Interestingly, none of the reported facial cases, including ours, demonstrated concomitant axillary involvement, most involved only the head and neck region. Our patient's lesions showed a slightly more extensive distribution, affecting the head, neck, upper chest, and back, while still sparing the axillae. This may reflect differences in underlying pathogenesis between sites. In the axillae, granular parakeratosis is commonly associated with irritant exposure from deodorants, antiperspirants, and occlusion.[4] In contrast, all four reported facial cases originated from Asia, where hot and humid climates are common. Three of these cases, including ours, reported recurrent flares during summer, coinciding with episodes of excessive sweating, supporting the hypothesis that heat and perspiration may play a key role in the pathogenesis of facial lesions.

The diagnosis relies on both clinical and histopathological examination. Clinically, lesions present as erythematous to brown hyperkeratotic papules and plaques, which may appear as discrete, confluent, or reticulated lesions and are often accompanied by satellite papules[4]. Characteristic histopathological features supporting the diagnosis include hyperkeratosis, compact parakeratosis, and retention of basophilic keratohyalin granules within the stratum corneum[2]. Subsequent reports have demonstrated a wide spectrum of clinical and histopathological findings. In 2021, Chirasuthat et al. proposed a classification system comprising five types: intertriginous

(axillary), eccrine ostium, follicular, acanthoma, and incidental types[8]. The intertriginous form typically presents with lesions in flexural areas and shows parakeratotic corneocytes containing keratohyalin granules localized to the epidermal surface, sometimes extending into adjacent hair follicles. The eccrine type presents with spiny keratotic papules, mainly on the neck, with one reported facial case (the second facial case in our discussion)[7,9,10]. Lesions are often aggravated by sweating, and histology shows changes confined to the stratum corneum of the eccrine ostia. The follicular type, presenting as follicular papules, was described in one facial case (the third facial case in our review) and one truncal case, both showing changes limited to the follicular infundibulum[8,11]. Notably, the first facial case showed typical epidermal alterations extending into the follicular epithelium, resembling the intertriginous type but without clinical involvement of flexural areas.

Our patient presented with lesions on the face, neck, and upper trunk, aggravated by sweating, which were clinically consistent with the eccrine ostium type. However, histopathological examination in our case revealed compact parakeratosis and retention of basophilic keratohyalin granules within the stratum corneum, involving the eccrine ostium but not predominantly localized to it, with additional involvement of the follicular epithelium. To the best of our knowledge, this is the first reported case of concurrent granular parakeratosis involving both eccrine and follicular structures. We hypothesize that this presentation may represent either a newly recognized subtype of appendageal involvement or a previously underrecognized feature. Further studies with a larger number of patients are warranted to confirm this hypothesis and to better delineate the spectrum of appendageal involvement in granular parakeratosis.

Treatment involves identifying and discontinuing the precipitating factor[1]. In some cases, spontaneous resolution has been reported. Management options include topical emollients, corticosteroids, salicylic acid, and vitamin D analogs, which may be used alone or in combination[12]. Recurrence may occur, particularly after re-exposure to triggering factors.

Conclusion

Facial involvement in granular parakeratosis is extremely rare, with only a few cases reported in the literature. Our case demonstrates atypical locations involving the face, neck, upper chest, and back, without axillary involvement or known topical triggers. These findings suggest that heat and sweating may play an important role in the pathogenesis of facial lesions. Histopathological examination showed extensive involvement of the normal epidermis, as well as the eccrine ostia and hair follicles. Awareness of this uncommon presentation is essential to avoid misdiagnosis and to guide appropriate management.

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