Flexural scaly papules and plaques

A 16-year-old female presented with slightly erythematous scaly papules involving both the axillae since the past 3 months. The lesions were episodically pruritic precipitated by sweating on exertion or warm weather. In addition, she had similar lesions involving both the cubital fossae and groins. There was no personal or family history of atopy. The lesions were predominantly discrete papules but had coalesced at places to form plaques with fine scaling [Figure 1, black circle]. The lesions partially remitted with topical agents (self-medicated by the patient) but never completely healed. Rest of the cutaneous examination was unremarkable. A potassium hydroxide preparation of the scales did not show any fungal elements. A discretely located scaly papule was biopsied for histopathological analysis [Figures 2 and 3].

What is Your Diagnosis?

Figure 1: Erythematous papules involving the left axilla with fine adherent scaling

Figure 2: Compact parakeratosis, epidermal hyperplasia, and dermal perivascular infiltrate is seen at lower magnification. Note the prominent granular layer (H and E, ×100)

Figure 3: Higher magnification of the epidermis shows compact parakeratotic stratum corneum with retention of keratohyalin granules within the cells and a prominent stratum granulosum (H and E, ×400)
**Answer**
Granular parakeratosis.

**Discussion**
Granular parakeratosis (intertriginous granular parakeratosis, zombie patch) is an uncommon acquired pruritic disorder of the flexures. It was first described by Northcutt et al. in 1991 as “axillary granular parakeratosis.” It affects all ages and both sexes, although a slight female preponderance is noted. Involvement of other flexures and nonflexural areas has also been described. Typical lesions are itchy and erythematous to brownish keratotic scaly papules that may coalesce into verrucous plaques. The exact etiopathogenesis of granular parakeratosis is unclear. The disorder is possibly an irritant response to antiperspirants, deodorants, and excessive use of soaps and creams. However, the current consensus is that such agents aggravate but do not induce granular parakeratosis. Recent studies have assigned this entity to the group of acquired keratinization disorders as a defect in the processing of profilaggrin impairing the normal cornification process demonstrated previously. Hoste et al. observed that mice deficient in caspase-14 (a vital enzyme necessary for the formation of an adequately functional epidermal barrier) were more prone to develop parakeratosis than control mice. Exploring this phenomenon in humans may help unravel the exact etiopathogenesis of granular parakeratosis. Co-occurrence of dermatophytosis and granular parakeratosis that responded effectively to antifungal treatment has been reported, suggesting the possible role of fungi in the disease. Hence, histopathological analysis should also include periodic acid Schiff and Giemsa staining to examine fungal elements.

**Table 1: Differential diagnoses of granular parakeratosis**

| Differential diagnoses | Differentiating clinical features |
|------------------------|----------------------------------|
| Acanthosis nigricans    | Characteristic hyperpigmented and velvety plaques with warty projections. Generally asymptomatic, along with involvement of other areas such as the neck aids in differentiation |
| Hailey-Hailey disease   | Moist warty plaques and erosions with surrounding erythema involving the flexures bilaterally and symmetrically. Involvement is generally limited to flexures |
| Darier’s disease        | Characteristic greasy, “dirty-warty papules” which may coalesce to form plaques in the flexures. The typical morphology and other pathognomonic features such as cobble-stoning of the palate, palmar pits, and the distal “V-shaped” nicking of the nail plate helps in differentiation |
| Pemphigus vegetans      | Moist vegetative lesions often studded with pustules and erosions as opposed to verrucous, keratotic plaques of granular parakeratosis |
| Inverse psoriasis       | Moist pruritic erosions with central fissuring at the depth of the flexure and peripheral collarette of fine scaling |
| Seborrheic dermatitis   | Similar to and occasionally indistinguishable from inverse psoriasis. Yellowish greasy scales and perifollicular erythema when present are helpful diagnostic pointers |
| Dermatophytosis         | Pathognomonic annular lesions with central clearing and a peripheral advancing erythematous edge which may exhibit tiny vesicles or pustules |
| Pityriasis rosea        | Annular erythematous lesions with central clearing and a distinctive peripheral collarette of scaling that typically trails the advancing edge |

Histopathology shows compact parakeratosis, psoriasiform epidermal hyperplasia, and the pathognomonic retention of keratohyaline granules within areas of parakeratosis with persistent granular layer. The process may also involve follicular and eccrine ostia.

Treatment options include topical steroids, topical calcipotriol, topical calcineurin inhibitors, and ammonium lactate. Oral isotretinoin is useful in refractory cases. Spontaneous resolution, as well as relapse after treatment are not uncommon. An infantile form, attributed to diaper wearing, has been described which may present in one of the two forms – linear bilateral keratotic plaques in the inguinal folds and erythematous geographical plaques undermine pressure points. Our patient was treated satisfactorily with topical tacrolimus 0.1% ointment. Granular parakeratosis resembles a host of other flexural dermatoses [Table 1]; hence, Ding et al. suggested reappraisal of this disorder merely as a reaction pattern rather than a distinct condition.

**Declaration of patient consent**
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

**Conflicts of interest**
There are no conflicts of interest.

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