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CASE REPORT

Granular Parakeratosis

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Abstract. Granular parakeratosis is a rare entity that results from an acquired disorder of keratinization. Clinically presents as dark erythematous plaques, occasionally pruritic, that usually involve the axilla and other intertriginous areas. The pathology is characteristic and consists of thickening of the stratum corneum with compact parakeratosis and retention of keratohyaline granules, whereas the stratum granulosum is preserved. The etiology is unknown although some factors such as irritating physical or chemical agents have been implicated. Treatment response is variable. We report a new case in a 50-year-old woman with brownish and hyperkeratotic plaques on both axillae, of two years duration, with a compatible pathology that showed a favorable response to tacalcitol.

Key words: granular, parakeratosis, tacalcitol.

PARAQUERATOSIS GRANULAR

Resumen. La paraqueratosis granular es una entidad poco frecuente que se caracteriza por ser una alteración adquirida de la queratinización. Clínicamente se presenta como placas eritematoparduscas, ocasionalmente pru-riginosas, que clásicamente aparecen en la axila y áreas intertriginosas. La histología es característica, donde se observa un engrosamiento de la capa córnea con una paraqueratosis compacta y persistencia de gránulos de que-ratohialina, mientras que el estrato granuloso se encuentra preservado. La etiología es desconocida, se postula la acción de factores irritantes físicos o químicos. La respuesta al tratamiento es variable. Presentamos un nuevo caso en una mujer de 50 años, con placas marronáceas abollonadas e hiperqueratósicas en ambas axilas, de dos años de evolución y con histología compatible, que presentó una buena respuesta al tratamiento con tacalcitol.

Palabras clave: paraqueratosis, granular, tacalcitol.

Introduction

Granular parakeratosis is a rare entity with characteristic histologic signs and is due to an abnormality in the keratinization process that leads to a marked thickening of the stratum corneum. The etiology is unknown but because irritants have been implicated, the condition could be considered as a protection mechanism of the epidermis. It typically presents in the axillae in association with the use of deodorants or perfumes. Cases have also been recorded in other intertriginous areas as well as in the abdomen, buttocks, and knees.

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We report a new case of axillary granular parakeratosis in a 50-year-old woman, with no known irritant factor and a good response to treatment with tacalcitol.

Case Report

A 50-year-old woman presented with a brownish, occasionally pruritic, plaque in the left axilla that had grown slowly over the previous 2 years. The patient reported that a similar asymptomatic lesion had begun to grow in the right axilla 3 months previously. The lesion was not related to the use of topical agents in the affected area. The patient had received treatment with corticosteroids and the condition had not improved.

Examination revealed a 6 cm × 1.5 cm brownish plaque in the left axilla with a granular, hyperkeratotic surface. The plaque had well-defined borders, with little infiltration and no flaking or vesiculation (Figure 1). The right axilla presented small confluent papules forming a smaller plaque

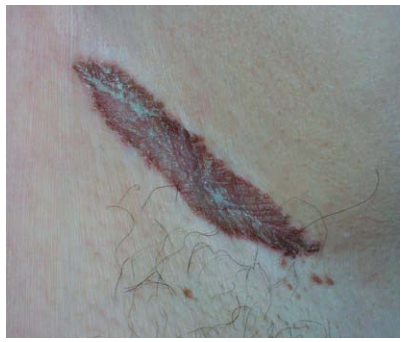

Figure 1. Brownish hyperkeratotic plaque in the left axilla.

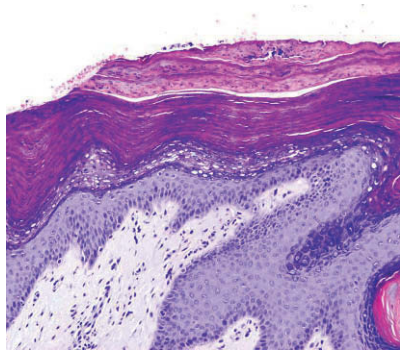

Figure 2. Acanthotic epidermis with a marked parakeratotic stratum corneum and retention of keratohyaline granules in the stratum spinosum. The papillary dermis shows a discrete perivascular lymphocytic infiltrate.

of similar characteristics. There were no other cutaneous or mucosal lesions.

A biopsy of the lesion in the left axilla showed an acanthotic epidermis with a marked parakeratotic stratum corneum and retention of keratohyalin granules in the stratum spinosum. The papillary dermis showed a discrete perivascular lymphocytic infiltration. These results were consistent with a diagnosis of granular parakeratosis (Figure 2).

Treatment was prescribed with tacalcitol twice daily for 3 months and the lesions improved.

Discussion

Granular parakeratosis is a rare entity caused by abnormal keratinocyte maturation and was first described in 1991 by Northcutt et al,[1] who presented 4 cases in the axillae. It has since been reported in other areas including the groin, submammary region, vulvar and perianal area, abdomen, buttocks, and knees.[2] Landes, in 1981, and Jansen 10 years later described 2 cases of hyperkeratotic lesions on the face and scalp in 2 women with a history of excessive use of facial cosmetic products.[3] Histologic tests would probably have presented signs of granular parakeratosis. After Northcutt, Mehregan et al[4,5] and Metze and Rutten[6] reported new cases involving the intertriginous areas and abdomen.

The condition appears more frequently in women between 40 and 60 years of age and generally takes the form of a single plaque in the axilla, although cases have also been reported in children—frequently in the area of the diaper, groin, and abdomen.[3,7,8] There have been 31 reported cases up to 2003 and we did not find any in Spain. Those cases included patients between the ages of 9 months and 77 years in whom different sites were affected and a varying response to treatment was observed.[7]

In each of those cases, clinical and physical examination of the affected sites showed results that matched those of our patient: clearly defined, brownish, hyperkeratotic plaques with a cobblestone appearance that were occasionally friable and pruritic.

Northcutt described the histopathology findings as compact parakeratosis with thickening of the stratum corneum and maintenance of the stratum granulosum. The presence of keratohyalin granules in the stratum corneum is characteristic.[1,3,5,6] A certain degree of vascular proliferation and ectasia is observed in the dermis, as well as an inflammatory lymphocytic infiltrate containing CD4 cells.[3,6] Other disorders include psoriasiform acanthosis and papillomatosis along with vacuolization of keratinocytes. These findings are similar to the histologic abnormalities observed in our patient.

The etiology is unknown. Nearly all authors agree on the existence of an irritant factor[1-8] that causes an associated thickening of the epidermis as a protection mechanism.[5,7] This argument, however, has been weakened by the fact that the condition frequently manifests unilaterally, and in many patients, it has not been possible to identify irritant factors (Mehregan et al,[4] Webster et al[9]). There must be other factors involved, including some individual predisposition. This was the case with our patient.

Regardless of the cause, keratinocyte maturation is altered on transition from the stratum granulosum to the stratum corneum. Profilaggrin in the keratohyalin granules is not processed into filaggrin monomers, which act as an adhesive matrix for keratin filaments. This leads to a failure in the breakdown of the keratohyalin granules, which are then retained in the stratum corneum.[1-3,7,9] Because filaggrin is only one component of these granules, however, this sequence is purely theoretical and abnormalities in other elements may explain the histologic findings.[1]

Differential diagnosis should be performed with entities such as Hailey-Hailey disease, pemphigus vegetans, acanthosis nigricans, seborrheic keratosis, Darier disease, inverse psoriasis, dermatophytosis, contact dermatitis, and pigmented and hyperkeratotic napkin dermatitis (PHND). These diseases also show intense hyperkeratosis with orthokeratosis and parakeratosis but without retention of the keratohyalin granules. Granular parakeratosis may be considered as a variant of PHND.[8]

Response to treatment varies. Cases have been described of spontaneous remission and remission when use of the irritant factor is stopped together with changes in hygiene habits and even simply washing with cold water.[1,3,7] The defect lies in an abnormality of keratinization and there are cases that have been treated with oral isotretinoin or vitamin D derivatives (tacalcitol, calcipotriol) with complete remission of the lesions. [7,10,11] Other treatments that have been tested, with varying results, include antibiotics and antifungal agents,[1,4] topical corticosteroids,[1,4-7] ammonium lactate,[10] and cryotherapy.[1, 7]

Our patient received treatment with tacalcitol twice daily for 3 months and the lesions improved.

Conflicts of Interest
The authors declare no conflicts of interest.

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