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CASE REPORT

Confluent and reticulated papillomatosis of Gougerot - Carteuad: case report

Papilomatose confluente e reticulada de Gougerot - Carteuad: relato de caso

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ABSTRACT

Introduction: Confluent and reticulated papillomatosis is a rare dermatological disorder of the epidermis, with a benign course and uncertain etiopathogenesis, characterized by the slow and progressive appearance of brownish or pink papules with a peripheral reticulate pattern, preferentially located at the center of the trunk, armpits and neck. It affects all races, genders and age groups, with a predilection for adolescents and young adults with higher phototypes. **Case report:** Two years ago, a 14-year-old male patient presented the appearance of speckled papules, with a rough surface, brownish, confluent, periaxillary, and a peripheral reticulate pattern. There was no response to treatment with the use of urea cream, topical antifungal agents and rubbing alcohol on the lesions. Direct mycological examination was negative. Fasting blood glucose, glycosylated hemoglobin, fasting insulin, and HOMA insulin resistance index were normal. The histopathological examination showed hyperkeratosis, papillomatosis, thinning of the granulosa and focal interpapillary acanthosis, confirming the clinical diagnosis of Confluent and Reticulated Papillomatosis. Treatment with 100 mg/day doxycycline associated with 0.1% adapalene at night for 60 days was established, with excellent improvement of the condition. **Conclusion:** Confluent and reticulated papillomatosis is a rare dermatosis with a benign course that must be recognized early in order to avoid misdiagnosis and inaccurate therapy, in addition to high costs for patients.

Keywords: Dermatosis; Papillomatosis; Hyperpigmentation; Keratosis.

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RESUMO

Introdução: A papilomatose confluente e reticulada é uma afecção dermatológica rara da epiderme, de curso benigno e etiopatogenia incerta, caracterizada pelo aparecimento lento e progressivo de pápulas acastanhadas ou róseas com padrão reticulado periférico localizadas preferencialmente no centro do tronco, axilas e pescoço. Acomete todas as raças, gêneros e faixas etárias, com predileção por adolescentes e adultos jovens com fototipos mais altos. **Relato de caso:** Paciente 14 anos, masculino, há dois anos iniciou quadro com surgimento de pápulas pontilhadas, de superfície áspera, acastanhadas, confluentes, periaxilares com padrão reticulado periférico. Não houve resposta ao tratamento com uso de creme de ureia, aos antifúngicos tópicos e à fricção de álcool nas lesões. O exame micológico direto foi negativo. Glicemia de jejum, hemoglobina glicosilada, insulina de jejum, índice de resistência insulínica HOMA estavam normais. O exame histopatológico evidenciou hiperceratose, papilomatose, adelgaçamento da granulosa e acantose focal interpapilar confirmando o diagnóstico clínico de Papilomatose Confluente e Reticulada, foi instituído tratamento com doxicicilina 100 mg/dia associado a adapaleno 0,1% à noite por 60 dias com excelente melhora do quadro. **Conclusão:** A papilomatose confluente e reticulada é uma dermatose rara, de curso benigno, que deve ser reconhecida precocemente, de forma a evitar diagnósticos e terapêutica equivocados, além de custos dispendiosos para os pacientes.

Palavras-chave: Dermatose; Papilomatose; Hiperpigmentação; Ceratose.

Introduction

Described in 1927 by the French dermatologists Gougerot and Carteaud, Confluent and Reticulated Papillomatosis (CRP)[1] is a rare dermatological disease of the epidermis characterized clinically by the slow and progressive appearance of brownish or rose papules with a peripheral reticulated pattern preferably located at the center of the trunk (mainly intermammary, interscapular and epigastric regions), underarms and neck. The mucous membranes, palms and plants are usually spared. Most of the lesions are asymptomatic; however, thin flaking and pruritus may be present in some cases[2,3].

Its etiology is still uncertain; however it is believed that CRP may be caused by the alteration of the differentiation and maturation of keratinocytes or the abnormal epidermal reaction secondary to microrganisms[4]. These hypotheses are based on the findings of electron microscopy that show architectural alterations of the cells of the cornea layer in addition to the proliferation of lamellar granules in the keratinocytes of the granulosa[2,5], as well as the isolation of *Dietzia papillomatosis,* Gram positive aerobic actinomycete, in an immunocompetent patient with CRP[5]. Probable genetic predisposition has been investigated after reports of cases in the family[4,5].

This article describes a typical clinical case, with histopathological confirmation and excellent response to the proposed treatment and discusses the differential diagnoses drawing attention to this entity, sometimes underdiagnosed in clinical practice.

Case report

Two years ago, a 14-year-old male patient, phototype IV of Fitzpatrick, started presenting a condition with the appearance of slightly pruritic periaxillary brownish lesions with occasional flaking. Prior treatment was done with urea cream 10% and topical antifungals without improvement of the symptoms.

The physical exam showed the patient to be overweight (weight: 78 kg, height 166 cm, body mass index - BMI: 28.3), with dotted papules with a rough surface, brownish, confluent and periaxillary with a peripheral reticulated pattern. The axillary center bilaterally presents papillomatosis with a macerated aspect (Figure 1).

The following diagnostic hypotheses were suggested: pityriasis versicolor, terra firma-forme dermatosis, acanthosis nigricans and confluent and reticulated papillomatosis (CRP) for periaxillary lesions and, in the central region,

Figure 1. Dotted, confluent brownish papules in the center with peripheral reticulated pattern. A. Right periaxillary; B. Left periaxillary.

acanthosis nigricans. There was no response to alcohol friction 70% on the lesions. Direct mycological examination was negative. Fasting glycemia, glycosylated hemoglobin, fasting insulin and HOMA (*homeostasis model assessment*) insulin resistance index were within the reference values. The periaxillary histopathological exam revealed hyperkeratosis, papillomatosis, granulosal thinning and interpapillary focal acanthosis (Figure 2).

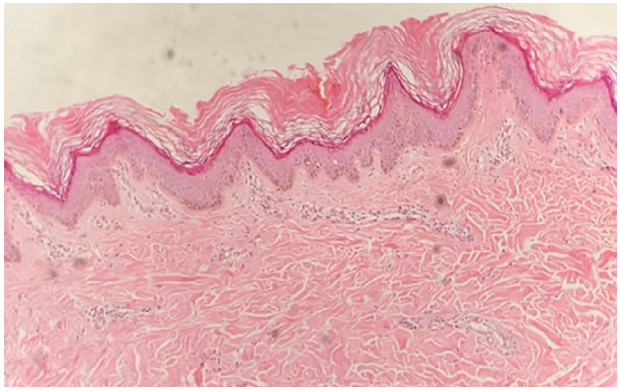

Figure 2. Histopathological examination: hyperkeratosis, papillomatosis, granulosa thinning and focal interpapillary acanthosis.

The clinic and the complementary exams confirmed the diagnosis of CRP in periaxillary lesions; treatment with oral doxycycline 100 mg/day, associated to adapalene 0,1% topical at night for 60 days was prescribed.

After the treatment, important regression of the lesions was observed -, except in the central area of acanthosis nigricans - which continued in the follow-up consultations (Figures 3 and 4).

Discussion

CRP is a dermatosis without systemic involvement that affects all races, genders and age groups, with a predilection for adolescents and young adults of the female sex from 10 to 35 years with higher phototypes[1,2], data compatible with the reported case.

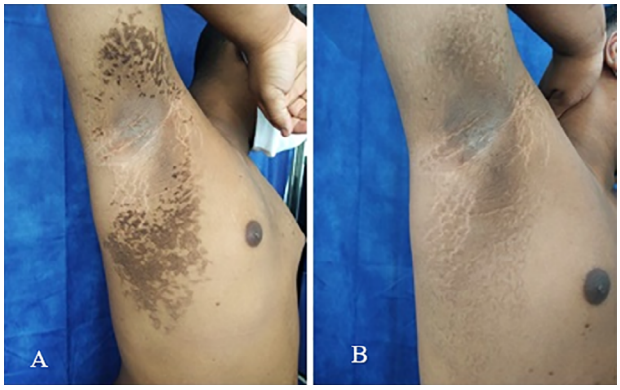

Figure 3. A. Right periaxillary before treatment; B. After treatment.

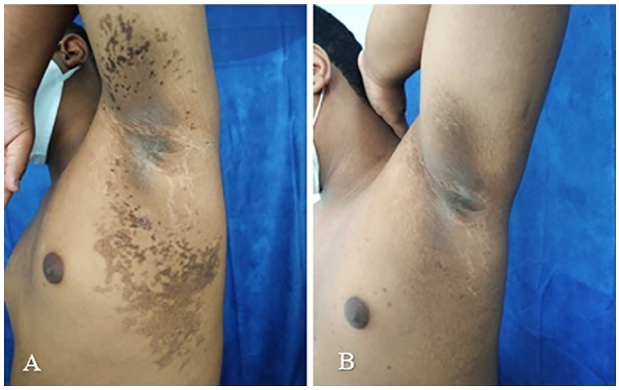

Figure 4. A. Left periaxillary before treatment; B. After treatment.

The diagnostic criteria for CRP were proposed by Davis et al. (2006)[6] based on a retrospective study in 39 patients: (a) clinical finding of brownish macules/plaques, some reticulated, and papillomatosis; (b) located in the upper portion of the trunk and neck; (c) negative mycological examination; (d) absence of response to antifungals; (e) and excellent response to minocycline and/or other antibiotics[6].

The histopathological study, although not specific, may present hyperkeratosis, more often orthokeratotic, with irregular papillomatosis, focal acanthosis and thinning of the granulous layer. Hyperpigmentation in the basal layer, with no change in the number of melanocytes, can also be observed[7].

The main differential diagnosis should be made with the pityriasis versicolor, whose identification of the fungus or its hyphae by preparation with KOH or biopsy confirms the disease. Cutaneous lesions located in intertriginous areas may be confused with acanthosis nigricans, but CRP extrapolates to areas classically affected by acanthosis nigricans for generalized trunk lesions, and it is also distinguished by the dotted feature of their papules; however, they can often coexist in the same patient[2,8].

The neglecta dermatosis and the terra firme-forme dermatosis are rare conditions, clinically similar to CRP. The first is due to the progressive accumulation of sebum, sweat, keratin, cellular debris and exogenous impurities, located on

the trunk, shoulders and neck, which can be removed with water and soap or alcohol 70%[9-11]. The second is a disorder of the maturation of keratinocytes that, despite normal sanitization, leads to the retention of keratin, sebum, sweat and melanin preferably on the neck, face and trunk, and is not removed by soap and water, but has an excellent response to rubbing with 70% alcohol-soaked cotton. The simple removal of verrucous lesions with alcohol 70% allows the distinction between these clinical conditions and CRP[2,12].

Other differential diagnoses possible with CRP are cutaneous amyloidosis, whose histopathological aspects are different from CRP; and Darier disease whose papular lesions have a follicular location and typical regional distribution[6,8].

In the therapeutic approach, topical agents (ceratolytics, vitamin A and D derivatives, antimicrobials) and systemic agents (antibiotics and retinoids) are described; however, although several of these have been used with relative success, there is no standard treatment[4]. More recent reports support the efficacy of minocycline 100mg/day and even other cyclins, during a period ranging from 8 to 10 weeks, without recurrence after 12 months, allowing to verify that this alternative is safe and effective[8,13,14]. Oral isotretinoin at a dose of 1 to 2mg/kg/day for 8 weeks is reported as a therapeutic possibility in refractory cases[2,15].

In the present case, there was a favorable and sustained response for a period of one year with the use of doxycycline 100 mg/day twice a day associated with Adapalene 0.1% at night for 60 days.

Conclusion

The CRP is a rare dermatosis with a benign course and uncertain etiopathogenesis, which arises in adolescence and should be recognized early in order to avoid misdiagnosis and inaccurate therapy, in addition to high costs for patients.

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