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Gougerot—Carteaud confluent and reticulated papillomatosis

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Gougerot—Carteaud confluent and reticulated papillomatosis is a dermatosis due to a genetically determined keratinization defect. It is a rare disease with typical histopathologic findings of hyperkeratosis, papillomatosis and modest epidermic acanthosis. We report two patients suffering from confluent and reticulated papillomatosis, presented with extensive lesions of symmetrically disposed plaques formed by the confluence of grayish-brown rough papules, distributed on the skin of the trunk and upper extremities. In both cases the histological examination was very important for establishing the proper diagnosis. We began treatment with topical calcipotriol and obtained remission that persisted during the control visits.

Key words: confluent and reticulated papillomatosis, Gougerot—Carteaud syndrome, histopathology, hyperkeratosis, therapy.

Introduction

Gougerot—Carteaud confluent and reticulated papillomatosis (CRP) is a rare disease due to a genetically determined keratinization disorder. There are reports of familial cases where two or more members of the same family were affected [7], but sporadic cases are more frequently described. Female are more frequently affected with a peak incidence during the pubertal period.

CRP lesions are usually persistent, asymptomatic, verrucous papules with a tendency to coalesce. Previously they were interpreted as pseudo acanthosis nigricans, but now it is a separate nosological entity with not well defined etiopathogenesis and treatment [4]. Histopathologic examination of the skin efflorescence is characteristic and shows hyperkeratosis, papillomatosis and modest epidermic acanthosis. Therapeutic opportunities include systemic antibiotics, retinoids, topical tazarotene, tacalcitol, or calcipotriol. Dermoabrasion, UVA phototherapy and laser treatment are also proposed [2, 5, 9].

We describe two cases of this rare disease CRP where the microscopic findings played a very important role for establishing the diagnosis.

Materials and Methods

A 16-year-old white female presented with a ten months history of a skin eruption of papules with a rough verrucous surface. They were localized on the nuchal region and forearms. They had no enlarged in size but had a tendency to coalesce. A slight scratch with fingernails produced a fine powdery material.

The other case was an 11-year-old white female presented plaques symmetrically disposed on the skin of the abdomen and upper extremities. The lesions consisted of grayish-brown rough papules, measuring some millimeters in diameter and had first appeared one year before. She had neither pruritus nor ache. The patient had no other compliances except the bad aesthetic view. She was treated with emollients and cold cream but only improvement of the skin mildness was achieved.

Laboratory investigations including complete blood counts, erythrocyte sedimentation rate (ESR), blood sugar, hepatic and renal function tests, serum electrolyte levels, lipid analysis and urinalysis, were performed. Skin biopsies of the papules were obtained. They were fixed in 10% neutral-buffered formaldehyde solution, processed routinely, and stained with hematoxylin-eosin.

Results

We formulated the diagnoses of CRP in both cases on the basis of the clinical picture, the histological findings, evolution of the disease and the age of the patients. The physiologic and pathologic anamnesis gave negative results and laboratory investigations did not reveal any alterations worthy of note. Wood's lamp observation showed no fluorescence. The histological examination evidenced the following alterations of the epidermis: hyperkeratosis, papillomatosis, mild acanthosis and the absence of melanocyte hyperplasia. Periodic acid- Schiff reaction did not evidence the presence of hyphal cells or mycetes (Fig. 1). The second case showed mild hyperkeratosis, mild acanthosis and almost an absence of papillomatosis, no spongio-

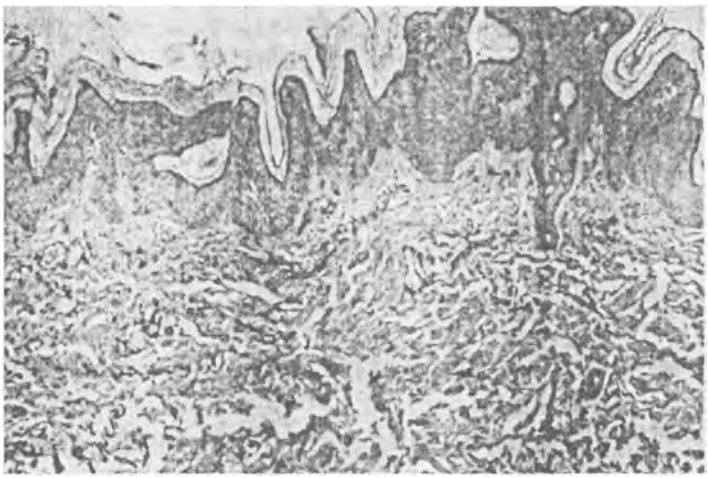

Fig. 1. Hyperkeratosis, papillomatosis, modest acantosis and absence of melanocyte hyperplasia. Hematoxylin-eosin stain × 100

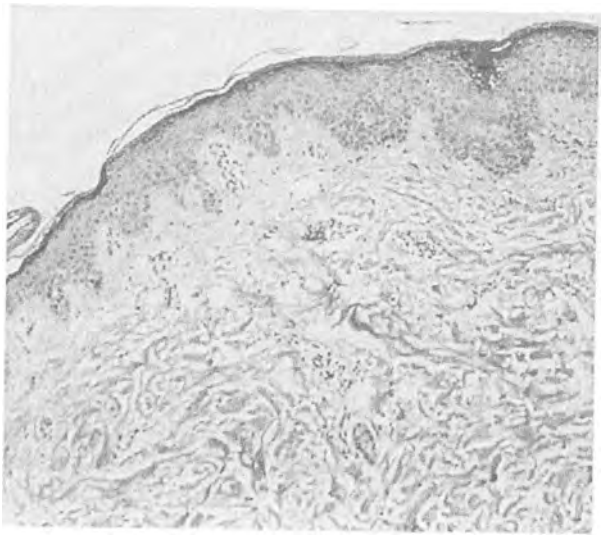

Fig. 2. Mild orthohyperkeratosis, acanthosis and almost absence of papillomatosis. Hematoxylin-eosin stain × 100

sis or significant inflammatory infiltrate (Fig. 2). These findings were very important to make the proper diagnosis as there were some differential diagnoses including acanthosis nigricans, Darier's disease, follicular hyperkeratosis, pityriasis versicolor and epidermodysplasia verruciformis. We began treatment with topical calcipotriol 0,005% applied twice daily for a two months period. On control visits performed at 1 and at 2 months no relapses were evident and a complete resolution was reached.

Discussion

Gougerot—Carteaud CRP is a rare dermatosis, which has a unique clinical feature and shows hyperkeratosis and papillomatosis histologically. This disease was first described by Gougerot and Carteaud in 1927 [6]. It is characterized by persistent, asymptomatic, verrucous papules that have a tendency to coalesce. The intermammary region is ussualy affected first with subsequent spread to the breast and abdomen. The interscapular area, neck and axilla also may be involved. The eruption often begins during puberty and more commonly affects women. Endocrine-metabolic alterations, an anomalous reaction of the host to colonization by the Pityrosporum orbiculare, a keratinization disorder either genetically determined or induced by toxic substances produced by unidentified agents, an early form of cutaneous amyloidosis or an anomalous epidermic response to UV rays with consequent aberrant keratinization, have been reported in an attempt to explain the pathogenesis of CRP [1].

Both evidence of papillomatosis clinical and histological has been pointed as important to achieve the diagnosis of CRP. These findings are presented in our first patient, the 16-year-old one. Almost the lack of papillomatosis in biopsy specimen from the lesions of the skin of our 11-year-old patient is interesting and unexpected.

We think that the reason for this is that this girl clinically had not verrucous surface of the eruption.

In our cases, the distribution of the lesions and histopathologic findings are consistent with those of CRP. One of the most important sign for the disease in histological examination is hyperkeratosis. In a recent publication an electronic microscopic study showed a marked increase in the number of lamellar granules in the granular layer [8]. This finding is interesting to speculate the pathophysiology of hyperkeratosis of CRP. Lamellar granules can mediate cell cohesion of stratum corneum by realizing contents and lipids. The hydrolytic contents of the granules are, in part, responsible for their reorganization and subsequent assembly in the intercorneocyte spaces to form the intercellular lamellae in the stratum corneum.

A treatment of choice for CRP does not exist. We treated our patients with calcipotriol, which is an analogous synthetic of 1,25 dihydroxyvitamin D3, a potent regulator of the cellular differentiation and an inhibitor of keratinocytes proliferation. Its mechanism is based on the regulation of an anomalous keratin expression. In CRP it leads the keratinocytes towards a more normal maturation and differentiation, reducing the expression of keratins of high molecular weight that are normally anomalous [3].

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