

Confluent and Reticulated Papillomatosis (Gougerot-Carteaud Syndrome) in Two Brothers

Confluent and reticulated papillomatosis (CRP) is a rare dermatosis of unknown origin characterized by hyperpigmented, confluent papules (1). CRP was first described in 1927 by Gougerot and Carteaud as "papillomatose pigmentée innominée" (2). It has been described as a relatively rare dermatosis manifesting as persistent papules that are confluent in the center and reticulated at the periphery (3). Lesions usually appear as small erythematous papules that evolve into hyperkeratotic verrucous plaques. Although the disease presents with characteristic cutaneous signs, it is difficult for physicians to diagnose (4,5). The differential diagnosis of CRP includes tinea versicolor, Darier's disease, acanthosis nigricans, prurigo pigmentosa, Dowling-Degos disease, and reticulate hyperpigmented eruptions (6). Sometimes, the accompanying positive family history, considered together with characteristic clinical findings, may point to the correct diagnosis (6). We report on the cases of two brothers with CRP that suggest genetic predisposition in family members and a possibility of bacterial etiology.



Figure 1. Older brother before treatment.

Two brothers, aged 19 and 16, were admitted to our outpatient service with maculopapular eruptions on the chests. The lesions had started two years ago in the older brother and two months ago in the younger brother. Both of them had been treated before for seborrheic dermatitis, but neither of them had any improvement with these treatment regimens. They did not have any other family members affected with similar lesions. In dermatological examination, both brothers had milimetric, light to dark brown colored, maculopapular lesions which were coalescing at the center and reticulating at the periphery, creating an inverted triangular shape on their chest skin (Figures 1,2). In laboratory examination, the results of a complete blood count and routine biochemistry were within normal limits. No fungal elements were seen in native microscopic examination.

Skin punch biopsies were performed, and pre-diagnoses of confluent and reticulated papillomatosis and acanthosis nigricans was established. In histopathological examinations, slight acanthosis, minimal papillomatosis, and superficial perivascular



Figure 2. Younger brother before treatment.

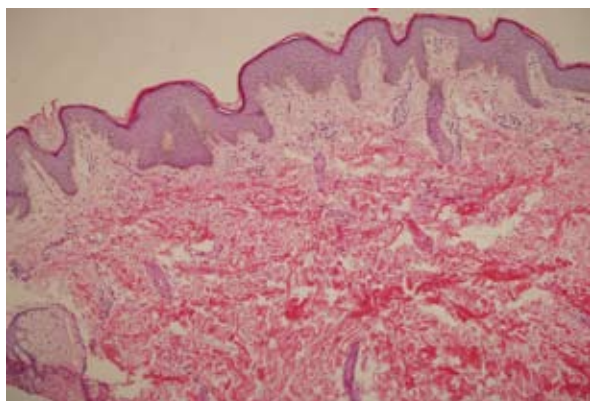


Figure 3. Histopathology of the older brother's lesions; slight acanthosis, minimal papillomatosis, and minimal superficial perivascular lymphocytic infiltration (H&E; x 100).

dermatitis were observed (Figures 3,4). Both of the brothers were treated with doxycyclin 100 mg/day PO, and topical tretinoin cream. After three months of treatment, lesions improved, becoming smaller in size and lighter in color (Figures 5,6). Oral doxycyclin, topical calcipotriol ointment, and topical emollients were also used in the follow up period. The patients are still being followed up every 3 months, with minimal clinical findings.

The etiology of confluent and reticulated papillomatosis (CRP) is not clear (2). The majority of cases are sporadic but familial cases were also reported. There are several hypotheses about the pathogenesis of the disease. The first suggests that the disease is an abnormal host response to *Pityrosporum orbiculare* or follicular bacteria (4,5). *Malassezia spp.* yeast and the Dietzia strain of *Actinomyces* infections are also considered in pathogenesis (2,4). The second hypothesis is a possible disorder of keratinization. Presence of amyloid in the lesions led some authors to classify the disease as a variant of skin amyloidosis. It was also defined as pseudoacanthosis nigricans (2). Other hypotheses include photosensitivity, endocrine abnormalities, and a hereditary predisposition (5).

The conditions considered in the differential diagnosis of CRP include tinea versicolor, Darier's disease, acanthosis nigricans, prurigo pigmentosa, Dowling-Degos disease, and reticulate hyperpigmented eruptions (1,6). Histological evidence of papillomatosis, resulting from the upward projection of finger-like dermal papillae which are covered by thinned epidermis specific to acanthosis nigricans, was not clearly present. However, slight acanthosis and minimal papillomatosis can be observed. No hyperkeratotic plugs of Darier's disease could be determined. Native preparation and mycological culture did not present any fungal elements.

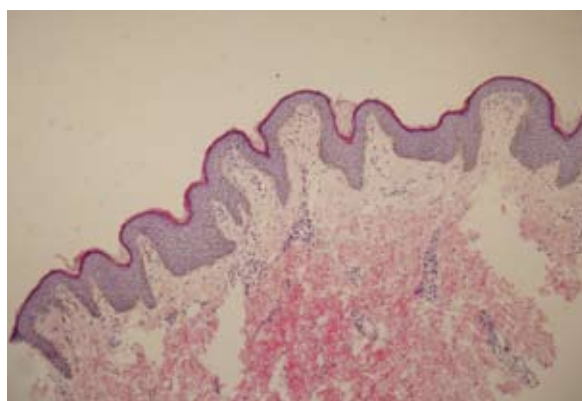


Figure 4. Histopathology of the younger brother's lesions is similar to that of the older brother: slight acanthosis, minimal papillomatosis, and minimal superficial perivascular lymphocytic infiltration (H&E; x 100).

Histopathologically, CRP shows slight hyperkeratosis, papillomatosis, and focal acanthosis (1). The presence of papillomatosis has been viewed as highly characteristic and important for making the diagnosis of CRP. Although the histological findings of CRP are characteristic, they are not diagnostic. Confluent and reticulated papillomatosis with no papillomatosis found histopathologically was also reported (7). While papillomatosis is usually found in fully developed lesions, it may not be seen or may be subtle in early or late lesions (2).

Treatment alternatives for CRP are variable and not standardized (1). Topical treatment agents that are generally used are calcipotriol, miconazole, tazarotene, 5-fluorouracil, salicylic acid, topical steroids, and topical tretinoin (1,5,8). Lesions may recur in a few months. Systemic antibiotics like minocycline, fusidic acid, azithromycin, clarithromycin, doxycyclin and erythromycin were also used.^{5,9} It is believed that these agents influence CRP through their anti-inflammatory,



Figure 5. Older brother after treatment.



Figure 6. Younger brother after treatment.

rather than antibacterial action, since no bacterial trigger has ever been identified in CRP lesions (10). Oral retinoids, radiotherapy, cryotherapy, and dermabrasion were also reported in the literature with variable efficacy results (5). Topical and systemic antifungal drugs were reported to be ineffective (7). Minocycline was reported as the treatment of choice in treatment of CRP (4). Since minocycline is not available in Turkey, we prescribed oral doxycyclin in our cases.

Despite the fact that the histopathological criteria were not fully compatible, these two brothers were diagnosed with CRP because of typical localization, clinical presentation, and good response to doxycyclin therapy. These cases are important reminders on the possibility of a genetic predisposition for CRP.

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