

## UV-TRIGGERED SYNDROME OF GOUGEROT-CARTEAUD - MECHANICAL DAMAGE OR IMMUNOLOGICAL REACTION

Sonya Marina, Kristina Semkova, Dimitrina Guleva, Jana Kazandjieva

*Clinic of Dermatology and Venereology, University Hospital Alexandrovska,  
Medical University of Sofia*

### ABSTRACT

Papillomatosis papulosa confluens reticulata et pigmentata is a rare dermatosis that presents clinically with pigmented, papillomatous, hyperkeratotic plaques with central confluence and reticulated peripheral pattern. The etiology remains unclear but current data points to a keratinization defect as the main pathological mechanism. There is one case report in the literature of a UV-induced Papillomatosis papulosa confluens reticulata et pigmentata. We present a 16-year old patient with clinical and histological diagnosis of confluent and reticulated papillomatosis. The characteristic lesions involved sun-exposed areas and developed after intense sun exposure. As an etiologic factor, ultraviolet light may lead to mechanical epidermal damage and further keratinization modification. On the other hand, in the setting of genetically predetermined resistance to UV-immunosuppression mechanisms, the newly formed epidermal antigens could potentially trigger an immune reaction resulting in skin eruption.

**Key words:** papillomatosis, ultraviolet light, keratinization, minocycline

Papillomatosis papulosa confluens reticulata et pigmentata (PPCRP) (Morbus Gougerot-Carteaud, Syndroma Gougerot-Carteaud) was first described in the literature in 1927. It is a rare dermatosis with unknown etiology and very characteristic clinical presentation. The disease occurs predominantly in young adults (17-21 years old on the average), with no racial or sexual predilection. Clinically, it presents with hyperpigmented, papillomatous, confluent, scaly patches and plaques, involving the neck, axillae and upper trunk in a rhomboid pattern. Gougerot and Carteaud proposed a set of diagnostic criteria which are still in use today, though new criteria have been proposed recently. The classical criteria

include the presence of scaly, brown macules, patches and papules that appear reticulated or papillomatous; negative fungal staining; and classical histology with papillomatous and hyperkeratotic epidermis and scarce perivascular lymphocytic infiltrate (9). In 2006 Davis et al, proposed new criteria derived from a large retrospective study of patients with PPCRP. These are 1. Typical clinical presentation; 2. Involvement of the upper trunk and neck; 3. Fungal staining of scales negative for fungus; 4. No response to antifungal treatment; 5. Excellent response to minocycline (5).

PPCRP is an asymptomatic skin disease without systemic involvement. Nevertheless, the rash is cosmetically unacceptable which warrants active treatment. Several treatment modalities have been tried and greatest success was reported with oral minocycline (7,22,25), oral and topical retinoids (13,17,26,27) and topical calcipotriol (1, 2). Isolated case reports of good treatment outcomes include other antibiotics (6), antifungals (3) and local mupirocin (8).

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**Address for correspondence:**

Kristina Semkova, MD, MSc  
Clinic of Dermatology and Venereology,  
University Hospital Alexandrovska,  
Medical University of Sofia  
1 G. Sofiiski Str., Sofia 1431, Bulgaria  
e-mail: kristina\_semkova@yahoo.com

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## CLINICAL CASE

A 15-year old girl was admitted to our clinic with a rash on the abdomen and back. The rash occurred two weeks after excessive sun exposure at the seaside. The initial changes were pigmented macules that later on transformed into greyish, flat, confluent papules. The skin looked 'dirty'. There was no family or personal history of similar disorders, nor personal history of concurrent diseases, endocrine disorders or intake of any medicinal products.

Physical examination revealed small grayish-brown, flat, confluent hyperkeratotic papules. The lesions were distributed over photoexposed areas of the body, mainly the abdomen and back, sparing the skin that was under the swimming suit. (Fig. 1, Fig. 2). Laboratory investigations did not show any pathology. Fungal staining was negative. On histological examination the epidermis was



Fig. 1. Papillomatous, hyperpigmented, velvety plaques involving the abdomen



Fig. 2. Papillomatous, hyperpigmented, velvety plaques involving the back and sparing the area under the swimming suit straps

papillomatous, hyperkeratotic and atrophic with vacuolar degeneration and increased melanin in the basal layer. Scarce perivascular lymphocytic infiltrate was observed in the dermis.

The patient was diagnosed with Papillomatosis papulosa confluens reticulata et pigmentata (Morbus Gougerot-Carteaud) on the ground of the typical clinical presentation and the histological examination.

Treatment was initiated with 0.05% retinoic acid cream and 5% salicylic ointment. Full resolution of the lesions was observed after 1.5 months.

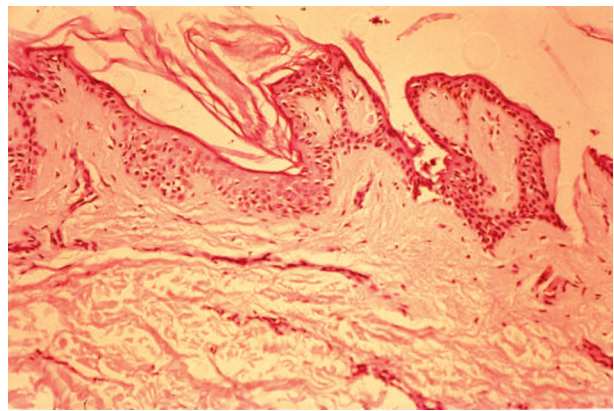


Fig. 3. Histological examination. H&E x100. Epidermal atrophy with hyperkeratosis, papillomatosis, vacuolar degeneration and sparse infiltrate in the upper dermis

## DISCUSSION

Papillomatosis papulosa confluens reticulata et pigmentata was described first by H. Gougerot and A. Carteaud in 1927. Initially they named it papillomatose pigmentée innommée, and later on renamed it to papillomatose pigmentée confluente et réticulée (10, 21). The etiology of the disease remains unknown. A reaction to *Pityrosporum ovale* was long thought to be the main triggering factor, but in 75% of patients the KOH staining for fungus is negative and the antifungal treatment is not effective (5). Other suspected etiological factors include endocrine disturbances (17), impaired keratinization, bacterial infection with a newly found *Actinomyces* strain – *Dietzia X* (20), as well as an association with the cutaneous form of amyloidosis (11). Exposure to UV light could trigger or exacerbate the disease (28). Famil-

ial forms are only rarely described (14,15), but they might be due to defects in keratinization.

G. Miescher was the first to propose the idea of impaired keratinization as the pathophysiological mechanism of PPCRP (19). This idea was further supported by electron microscopy and immunohistochemical analysis. Electron microscopy shows an increased number of lamellar granules in stratum granulosum, transit cells between stratum granulosum and stratum corneum and increase in the number of melanosomes in stratum corneum (16, 18). Immunohistochemical studies show increased expression of markers for keratinocyte differentiation and proliferation – involucrin, keratin 16 and Ki-67 protein (15). Additional evidence to this hypothesis is the remarkable therapeutic effect of retinoids and vitamin D analogues whose main mechanism of action is regulation of cellular differentiation and inhibition of keratinocyte proliferation.

Ultraviolet light causes multiple variable changes in skin structures and cellular elements. In the epidermis the damage is presented by epidermal hyperplasia of stratum corneum with increased proliferation of keratinocytes, increased melanogenesis, defective barrier function and keratinocyte DNA alterations (4). Immunohistochemically these changes translate into amplified expression of K6, K16, K17 and Ki-67 (27). Besides the cellular morphological changes, UV light causes local and generalized immunosuppression. This immunosuppression prevents an immunological reaction to the newly formed antigens resulting from the keratinocyte DNA injury. It is now known that the genetically determined resistance to UV-light-induced immunosuppression is the pathogenetic reason for the development of polymorphic light eruption (29).

Ultraviolet light could trigger the syndrome of Gougerot – Carteaud through several pathogenetic mechanisms. UV-induced changes and PPCRP have similar immunohistochemical and histological characteristics, both being characterized by enhanced proliferation and impaired keratinization. On the background of certain genetic predisposition UV changes may present clinically as PPCRP. On the other hand, similar to polymorphic light eruption, in our patient and in the other few UV-sensitive cases reported in the literature (20), the mechanism might

be a genetically pre-determined resistance to the immunosuppressive action of UVB resulting in the characteristic epidermal reaction towards the newly formed antigens. This hypothesis is supported by the successful treatment with minocycline and other antibiotics from the group of tetracyclines, that are known to have an immunomodulatory effect besides their antibacterial activity. (24)

PPCRP is a benign dermatosis but its clinical presentation is cosmetically unacceptable and embarrasses the patients. Understanding the etiology and pathogenesis would help to specify the most optimal treatment.

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